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Neuro- Ophthalmology 2023

When Should I Worry?
Concerning Signs,
Symptoms, and Findings
in Neuro-Ophthalmology

Subspecialty Day | AAO 2023
San Francisco | Nov 3

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Neuro-Ophthalmology 2023

When Should I Worry? Concerning Signs, Symptoms, and Findings in Neuro-Ophthalmology

Program Directors

Peter A Quiros MD and Madhura A Tamhankar MD

In conjunction with the North American Neuro-Ophthalmology Society

Moscone Center
San Francisco, California
Friday, Nov. 3, 2023



Presented by:
The American Academy of Ophthalmology

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Neuro-Ophthalmology 2023

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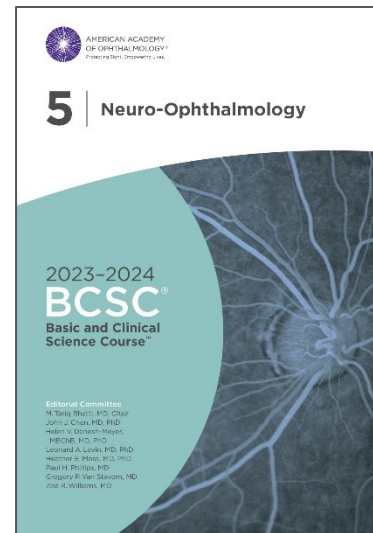


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On behalf of the American Academy of Ophthalmology and the North American Neuro-Ophthalmology Society (NANOS), it is our pleasure to welcome you to San Francisco and Neuro-Ophthalmology 2023:
When Should I Worry? Concerning Signs, Symptoms, and Findings in Neuro-Ophthalmology.



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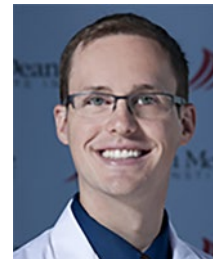
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Neuro-Ophthalmology 2023 Contents

	Program Planning Group	iii
	CME	viii
	Faculty Listing	x
	How to Use the Audience Interaction Application	xiv
	Program Schedule	xv
Section I:	What Meds Is My Patient On? Adverse Reactions of Systemic Medications	2, 18
	United for Sight: A Vision for Effective Advocacy	5
Section II:	Swollen Optic Nerve	7, 23
Section III:	Neuro-Ophthalmic Mimickers and Visual Disturbances	10, 28
Section IV:	Double Vision—What to Do?	13, 32
	Faculty Financial Disclosure	37
	Presenter Index	40

CME Credit

The Academy's CME Mission Statement

The purpose of the American Academy of Ophthalmology's Continuing Medical Education (CME) program is to present ophthalmologists with the highest quality lifelong learning opportunities that promote improvement and change in physician practices, performance, or competence, thus enabling such physicians to maintain or improve the competence and professional performance needed to provide the best possible eye care for their patients.

Neuro-Ophthalmology Subspecialty Day Meeting 2023 Learning Objectives

Upon completion of this activity, participants should be able to:

- Recognize neuro-ophthalmic side effects of systemic medications
- Differentiate the etiology of visual loss in those presenting with optic disc edema due to different causes
- Characterize symptoms such as photophobia, dysphotopsia, and hemeralopia as they can mimic neuro-ophthalmic disease
- Formulate hypotheses for patients who present with failure of visual improvement after cataract surgery that may harbor a potential neuro-ophthalmic condition
- Interpret crucial preoperative examination findings that can help diagnose such patients to avoid poor outcomes after cataract surgery

Neuro-Ophthalmology Subspecialty Day Meeting 2023 Target Audience

The intended audience for this program is comprehensive ophthalmologists.

Teaching at a Live Activity

Teaching an instruction course or delivering a scientific paper or poster is not an *AMA PRA Category 1 Credit*[™] activity and should not be included when calculating your total *AMA PRA Category 1 Credits*[™]. Presenters may claim *AMA PRA Category 1 Credits*[™] through the American Medical Association. To obtain an application form, please contact the AMA at www.ama-assn.org.

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Subspecialty Day 2023 CME Credit

The American Academy of Ophthalmology is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide CME for physicians.

Friday Subspecialty Day Activity: Glaucoma, Neuro-Ophthalmology, Ocular Oncology and Pathology, Refractive Surgery, and Retina (Day 1)

The Academy designates this Other (blended live and enduring material) activity for a maximum of 12 *AMA PRA Category 1 Credits*[™]. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

Saturday Subspecialty Day Activity: Cornea, Oculofacial Plastic Surgery, and Retina (Day 2)

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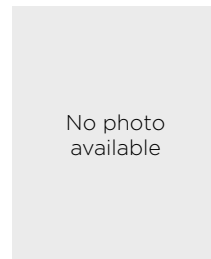
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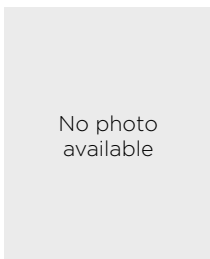
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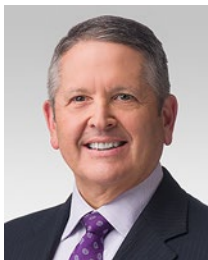
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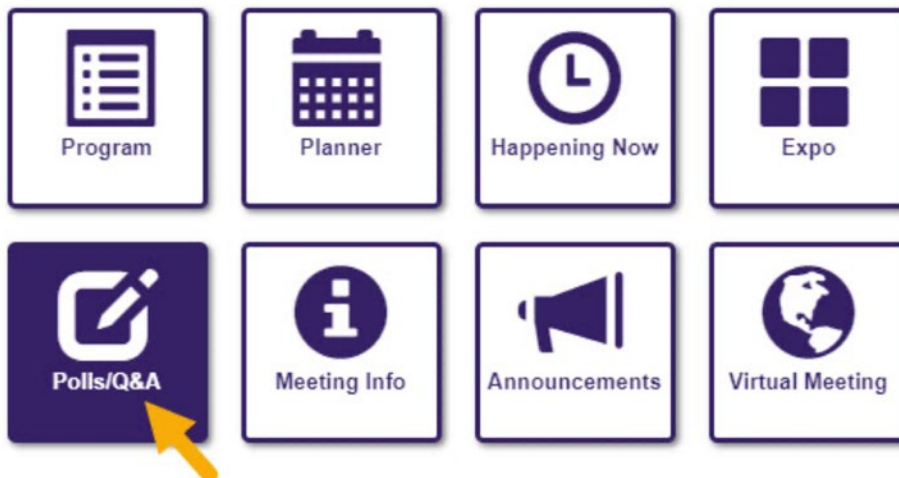
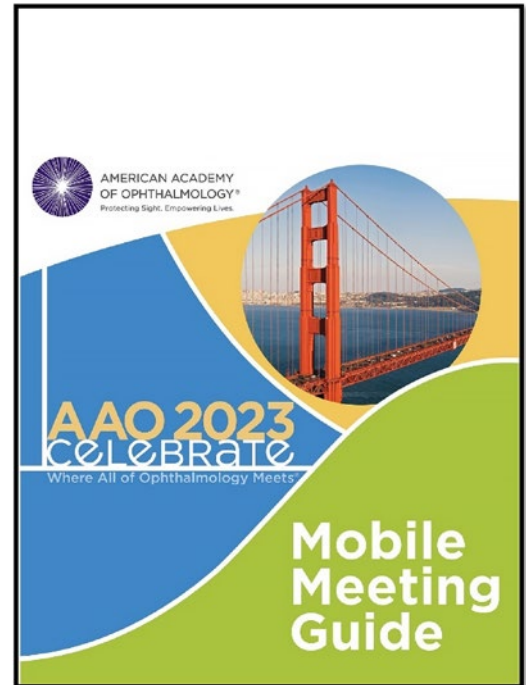


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- Choose “Ask a Question”
- Choose “Answer Poll”



Neuro-Ophthalmology Subspecialty Day 2023

When Should I Worry? Concerning Signs, Symptoms, and Findings in Neuro-Ophthalmology

7:00 AM CONTINENTAL BREAKFAST

8:00 AM Welcome and Introductions
Peter A Quiros MD
Madhura A Tamhankar MD

Section I: What Meds Is My Patient On? Adverse Reactions of Systemic Medications

Moderators: Courtney E Francis MD and Collin M McClelland MD

Panelists: Julie Falardeau MD, Mark L Moster MD, Howard D Pomeranz MD PhD, and Judith E Warner MD

8:02 AM	Audience Interaction	Peter A Quiros MD	
8:03 AM	"I Woke Up and Could Not See"	Michael S Lee MD	2, 18
8:23 AM	"I Have Melanoma, and I Am Seeing Double"	Dan R Gold DO	2, 18
8:43 AM	"I Am on Ethambutol—What Should I Look For?"	Nailyn Rasool MD	3, 19
9:03 AM	"I Cannot See, and Now I Cannot Hear"	Peter W MacIntosh MD	3, 20
9:23 AM	"I Have Blurred Vision and Swollen Nerves"	Amanda D Henderson MD	4, 22
9:43 AM	Summary	Madhura A Tamhankar MD	
9:45 AM	United for Sight: A Vision for Effective Advocacy	Prem S Subramanian MD PhD	5
9:50 AM	REFRESHMENT BREAK		

Section II: Swollen Optic Nerve

Moderators: John J Chen MD PhD and Kimberly K Gokoffski MD

Panelists: Steven L Galetta MD, Mark J Kupersmith MD, Nancy J Newman MD, and Jade S Schiffman MD

10:20 AM	Young Patient With Acute Vision Loss and Swollen Nerve	Neena Cherayil MD	7, 23
10:40 AM	Older Patient With Vision Loss and Swollen Nerve	Marc H Levin MD	7, 24
11:00 AM	65-Year-Old With Visual Loss and Swollen Nerve	Anita A Kohli MD	8, 24
11:20 AM	"I Have Eye Pain, and My Nerve Is Swollen"	Gregory P Van Stavern MD	8, 25
11:40 AM	"My Nerve Is Swollen, and I Cannot See"	Laura Bonelli MD	9, 27
12:00 PM	Summary	Peter A Quiros MD	
12:02 PM	LUNCH		

Section III: Neuro-Ophthalmic Mimickers and Visual Disturbances

Moderators: Chantal Boisvert MD and Eric L Berman MD

Panelists: Tomas S Aleman MD, Kathleen B Digre MD, Jeffrey G Odel MD, and Nicholas J Volpe MD

1:22 PM	"I Am Blinded by the Light"	Susan P Mollan MBChB	10, 28
1:42 PM	"I Have Normal Vision, but My Optic Nerves Are Pale"	Tatiana Bakaeva MD PhD	10, 28
2:02 PM	"My Vision Is Normal, but I Cannot See"	Rudrani Banik MD	10, 29
2:22 PM	"I Have a Visual Field Defect, but My Nerve Looks Normal"	M Tariq Bhatti MD	11, 30

2:42 PM	“I Have Light Sensitivity”	Samuel J Spiegel MD	12, 31
3:02 PM	Summary	Madhura A Tamhankar MD	
3:04 PM	REFRESHMENT BREAK		

Section IV: Double Vision—What to Do?

Moderators: Michael Dattilo MD and Valerie I Elmalem MD

Panelists: Jane C Edmond MD, Paul H Phillips MD, and Prem S Subramanian MD PhD

3:34 PM	Case of Acute Double Vision in a Younger Patient	Lauren C Ditta MD	13, 32
3:54 PM	Case of Acute Double Vision in an Older Patient	Crandall E Peeler MD	13, 33
4:14 PM	Cases of Esotropias With Sixth Nerve Palsies, Sagging Eye, Heavy Eye: When to Image?	Zoë R Williams MD	14, 34
4:34 PM	Surgical Considerations in Diplopia	Ore-Ofeoluwatomi O Adesina MD	15, 35
4:54 PM	Summary	Peter A Quiros MD	
4:56 PM	Closing Remarks	Peter A Quiros MD Madhura A Tamhankar MD	
5:00 PM	ADJOURN		

Case Presentations

Section I: What Meds Is My Patient On?

Adverse Reactions of Systemic Medications

“I Woke Up and Could Not See”

Michael S Lee MD

CASE PRESENTATION

History and Exam

An 84-year-old man underwent uncomplicated cataract surgery in the right eye (RE) in late April. He then had cataract surgery on his left eye (LE) in early May. He noted difficulty reading at his 1-week postop visit and was noted to have cystoid macular edema in the LE. He awoke in late June with slightly blurred vision in the LE and was noted to have optic disc edema in the LE at his 1-month postop visit.

His past medical history included gout, idiopathic cardiomyopathy, congestive heart failure, aortic regurgitation, aortic root dilatation, atrial fibrillation, hyperlipidemia, hypertension, prostate cancer, peripheral vascular disease, and erectile dysfunction. His medications were allopurinol, alendronate, amiodarone, atorvastatin, doxazosin, fluticasone, furosemide, metoprolol, and rivaroxaban. He denied symptoms of giant cell arteritis. He was a former smoker and drank about 1-2 alcoholic beverages several times a week. He was allergic to gabapentin, and his family history was noncontributory.

Examination on July 1 showed that his visual acuities were 20/40 RE and 20/50 LE. He had a subtle left afferent pupillary defect. He identified 11/11 Ishihara plates RE and 10/11 LE. His exam demonstrated corneal verticillate, both eyes, and his extraocular motility was normal. His cup-to-disc ratio was 0.05 RE and 0.0 LE. There was a normal optic disc RE and moderately severe disc edema LE, with no hemorrhages, lipid, or cotton wool spots. Visual field testing showed an arcuate visual field defect RE and a subtle central scotoma and inferior constriction LE. OCT showed a lamellar macular hole RE and significant macular edema LE, without vitreopapillary or vitreomacular traction in either eye.

Clinical Course

The patient's lab testing showed normal ESR and CRP. Cardiology agreed to stop the amiodarone. He returned 7 weeks later with only mild improvement of the optic disc edema and significant improvement in the macular edema LE. The macular hole was moderately improved in the RE. He returned 3.5 months after his initial visit, and there was mild persistent optic disc edema LE and the macular hole was closed RE. He returned 1 year later, and the disc edema was resolved LE. The visual fields remained abnormal in each eye, and the RE was deemed testing artifact. The retinal nerve fiber layer remained stable in the RE and became thinned in the LE.

“I Have Melanoma, and I Am Seeing Double”

Dan R Gold DO

CASE PRESENTATION

History and Exam

A 73-year-old man presented with 2 weeks of new binocular horizontal diplopia, which occurred suddenly and then gradually worsened. He experienced occasional headaches but denied vision loss, eye pain, or any limb weakness or numbness.

Examination demonstrated normal visual acuity OU and pupils that were briskly reactive to light, equal, without relative afferent pupillary defect. Confrontation visual fields were normal OU. There were bilateral mild abduction deficits, -0.5 OU. He had a comitant 6 PD esotropia. IOPs were normal at 14 OD, 16 OS, and anterior segment and dilated fundus examinations were unremarkable.

Past medical history was significant for hypertension and hyperlipidemia, with a recent diagnosis of melanoma treated with pembrolizumab starting 2.5 months prior.

Clinical Course and Outcome

MRI of the brain and orbits with and without contrast was normal. Laboratory evaluation included acetylcholine receptor antibodies, muscle specific kinase antibodies, TSH, CBC, B12 and methylmalonic acid, SPEP, and ganglioside antibody panel (GM 1, GM 2, GD1a, GD1b, GQ1b), as well as a paraneoplastic antibody panel (which included amphiphysin, AGNA-1, ANNA-2, ANNA-2, ANNA-3, CRMP-5, neuronal K channel, P/Q Ca channel, PCA-a, PCA-2, PCA-Tr).

Examination 2 months after the onset of symptoms again showed normal afferent visual function without anisocoria, although the bilateral abduction deficits had progressed to -2 OU, with bilateral and symmetric -1 supraduction deficits and -0.5 adduction deficits. There was a comitant 14 PD esotropia and 4 PD left hypertropia. There was mild ptosis with fatigability and lid twitch OD.

At this time, additional diagnostic workup was initiated, including single-fiber EMG, which demonstrated an abnormality in orbicularis oculi neuromuscular transmission. Myasthenia gravis with ocular involvement was diagnosed, which was thought to represent an autoimmune adverse event of pembrolizumab, a monoclonal antibody that targets programmed death protein 1 (PD-1).

The pembrolizumab was stopped and prednisone 40 mg daily was started, with minimal improvement over weeks. Neuromuscular consultation resulted in the discontinuation of prednisone, and the initiation of intravenous immunoglobulin.

“I Am on Ethambutol— What Should I Look For?”

Nailyn Rasool MD

CASE PRESENTATION

History and Exam

A 32-year-old man (he/him) presented for progressive blurring of his vision. The patient reported that 6 weeks prior, he began noticing difficulty reading novels; this progressed to difficulty seeing his phone clearly and driving. He reported the decline in his vision was bilateral and painless and occurred insidiously. He denied having flashes, floaters, complete loss of vision, or double vision. He denied headaches, weakness, numbness, and difficulties with coordination and walking.

The patient was previously healthy but had been diagnosed with tuberculosis 7 months prior, when he presented to the emergency department with fevers, chills, weight loss, and an intractable cough. He had emigrated from Pakistan 1 year prior. The patient was placed on rifampin, isoniazid, pyridoxine, and ethambutol (25 mg/kg/day). He did not consume alcohol, cigarettes, marijuana, or other substances, and he lived alone.

On examination this patient's visual acuity was 20/400 OD and 20/300 OS. There was dyschromatopsia bilaterally, with 3/8 Ishihara plates correct OD and only the test plate correct OS. Pupils were equal, round, and reactive to light, without a relative afferent pupillary defect. Slit-lamp and dilated fundus examinations were unremarkable, including normal optic nerves, maculae, vessels, and periphery. Humphrey visual field testing demonstrated cecentral scotomas bilaterally. OCT showed normal retinal nerve fiber layer bilaterally compared to age-matched controls.

Clinical Course and Outcome

The patient underwent an MRI of his brain and orbits with and without contrast, which was unremarkable. He underwent extensive normal blood work, which was negative for syphilis, Lyme disease, bartonella, and HIV and had normal serum vitamin B12, folate levels and metabolite testing. QuantiFERON-TB Gold returned positive/abnormal.

He was diagnosed with ethambutol optic neuropathy and was started on oral B12, folate, copper, and zinc supplementation. Ethambutol was discontinued under the close guidance of an infectious disease specialist. Two weeks following, his vision improved to 20/200 OD and 20/100 OS and 4/8 Ishihara color plates bilaterally. At 6 weeks follow-up, his vision was 20/25 OD and 20/30 OS with 7/8 Ishihara color plates OD and 6/8 OS. His cecentral scotomas had decreased in size. His optic nerves demonstrated subtle temporal pallor.

“I Cannot See, and Now I Cannot Hear”

Peter W MacIntosh MD

CASE PRESENTATION

History and Exam

A 56-year-old man was referred with recent diagnosis of thyroid disease. Over the last few months, he had developed upper eyelid swelling OD>OS, binocular double vision, and left eye blurry vision.

His past medical history included diabetes mellitus, hypertension, hypercholesterolemia, and nonalcoholic steatohepatitis (NASH). His medications included methimazole, atorvastatin, rivaroxaban, metoprolol, and sitagliptin. He had no history of hearing problems or exposure to ototoxic medications or loud noises.

On examination, his BCVA was 20/40 pinhole 20/25 in the right eye and 20/20 in the left. His external exam was significant for upper and lower eyelid edema with retraction and conjunctival injection but no corneal erosions OU. He had no relative afferent pupillary defect, his IOPs were in the normal range, and his Ishihara plates were full OU. His exophthalmometry measurements were 24 mm OU. His motility measurements are shown in Figure 1.

His posterior exam was normal without disc edema or pallor. His initial clinical activity score (CAS) was 4/7.

	-2			-4	
-1	OD	-1	-1	OS	0
	-4			-1	

Figure 1

Clinical Course and Outcome

This patient presented with several months of symptoms consistent with active thyroid eye disease, and his CAS was 4. His Humphrey visual field testing showed a few scattered changes superiorly in the left eye, and contrast-enhanced MRI of the orbits showed enlargement of several extraocular muscles, including bilateral inferior rectus, medial rectus, and superior rectus muscles, with some apical crowding OD>OS. The patient was offered teprotumumab at 10 mg/kg at the first infusion, followed by 20 mg/kg for the remaining 7 infusions, given at 3-week intervals. His baseline hearing testing was normal.

While awaiting approval for teprotumumab, his CAS worsened to 5/10 due to worsening motility.

He completed 5 rounds of teprotumumab, with improvement in double vision, acuity, proptosis, and CAS. However, he noted he was developing bothersome hearing problems, with a sense of his ears being plugged and difficulty hearing. He also developed elevated blood sugars. He was referred for repeat audiogram testing, which confirmed moderately severe sensori-

neural hearing loss in both ears. His PCP adjusted his diabetic medications.

Multidisciplinary discussion with audiology, endocrinology, and oculoplastics came to the decision to stop teprotumumab to avoid further hearing loss and diabetic complications. Two months after cessation of teprotumumab, the patient's HbA1c was improving and his CAS was 1/10. He reported no improvement in hearing. A follow-up audiogram is currently pending, and bilateral sound amplification devices were recommended by the audiologist.

"I Have Blurred Vision and Swollen Nerves"

Amanda D Henderson MD

CASE PRESENTATION

History and Exam

A 26-year-old woman presented for evaluation of transient visual obscurations. She reported constant poor quality of vision in both eyes for 1 month, with associated dimming and tunneling of her vision that occurred when turning her head in either direction or when quickly changing position. She also had constant, dull headaches, which were worse on lying down, and intermittent pulsatile tinnitus. She did not have diplopia.

She had been in her normal state of health until about 6 months prior, when she developed pulsatile tinnitus and positional dizziness. Neuroimaging at that time demonstrated a large posterior fossa cyst that was associated with significant mass effect on her cerebellum and resultant Chiari malformation. Four months prior, she underwent fenestration of the cyst with neurosurgery. She did well in the immediate postoperative period and was discharged home on postoperative day 2. However, 8 days later, she re-presented with wound drainage and underwent wound revision surgery. No cerebrospinal fluid (CSF) leak was identified, but cultures grew *Staphylococcus pseudintermedius*. Therefore, she was evaluated by the infectious disease team and was started on antibiotic treatment with vancomycin. A PICC line was placed, and she continued IV vancomycin treatment for 6 weeks. During that time, additional sensitivity analysis was performed, and sensitivity of the organisms to minocycline was demonstrated. Therefore, her PICC line was removed, and oral treatment with minocycline was initiated, with a plan for at least 6 months of treatment. MRI completed 2 months postoperatively showed postoperative changes from the arachnoid cyst fenestration, with a small residual cystic fluid collection.

On examination, visual acuity was 20/20 in the right eye and 20/20 in the left eye. There was no relative afferent pupillary defect. Extraocular motility was full. IOP was 23 mmHg in the right eye and 17 mmHg in the left eye. She correctly identified 13/13 Ishihara color plates with the right and left eyes. Anterior segment examination was unremarkable, and fundus examination demonstrated bilateral Frisen grade 4 optic disc swelling. Humphrey visual field 24-2 showed a few nasal missed spots in both eyes, with an enlarged blind spot in the left eye. OCT of the retinal nerve fiber layer demonstrated thickening in both eyes, with average values of 338 microns in the right eye and 399 in the left eye. Her weight was 127 pounds, with body mass index of 21.

Clinical Course and Outcome

Due to the severity of the patient's optic nerve swelling, acetazolamide 1 gram twice daily was initiated immediately, with titration to 1500 mg twice daily, with resultant improvement in optic nerve swelling. An updated MRI showed postoperative changes without any concern for obstructive cause for her optic disc swelling.

At this point, she had been on minocycline treatment for 3 months. Due to concern that minocycline was the cause for or a contributing factor to the optic nerve swelling, cessation of minocycline was trialed after approval by infectious disease team.

Ten days after holding the minocycline, she underwent lumbar puncture under fluoroscopic guidance, with an opening pressure of 24 cm H₂O (holding her acetazolamide for 3 days prior). CSF studies were unremarkable, including negative cultures. She followed up 3 weeks later, at which time her papilledema continued to improve, off acetazolamide and minocycline. By 2 months later (4 months after initial presentation to neuro-ophthalmology), her papilledema had completely resolved, with marked improvement in her visual symptoms. Visual acuity remained 20/20 in the right and left eyes, and Humphrey visual fields were unremarkable aside from slight blind spot enlargement in the left eye.

United for Sight: A Vision for Effective Advocacy

Neuro-Ophthalmology Subspecialty Day 2023

Prem S Subramanian MD PhD

Action Requested: *Donate to strengthen ophthalmology's legislative voice and protect patients and your profession*

Please respond to your Academy colleagues and join the community that advocates for ophthalmology: OPHTHPAC, the Surgical Scope Fund, and your State Eye PAC. Ensure you and your patients are heard by our nation's lawmakers by giving to each of these funds.

Where and How to Contribute

During AAO 2023 in San Francisco, please contribute to OPHTHPAC® and Surgical Scope Fund at one of our two convention center booths or [online](#). You may also donate via phone to both funds by sending two texts:

- Text **MDEYE** to **41444** for OPHTHPAC
- Text **GIVESSF** to same number (**41444**) for the Surgical Scope Fund

We also encourage you to support our congressional champions by making a personal investment via OPHTHPAC Direct, a unique and award-winning program that lets *you decide* who receives your political support.

Surgical Scope Fund contributions are *completely confidential* and may be made with corporate checks or credit cards. PAC contributions may be subject to reporting requirements.

Why Should You Contribute?

Member support of the Academy's advocacy funds—OPHTHPAC and the Surgical Scope Fund—powers our advocacy efforts at the federal and state levels. When you give to OPHTHPAC, you give ophthalmology a voice on Capitol Hill on critical issues like Medicare payment, optometry's scope expansion efforts in the VA, and prior authorization and step therapy burdens. When you give to the Surgical Scope Fund, you're funding our efforts to fight dangerous optometric surgery initiatives at the state level, whenever and wherever they arise. And finally, when you give to your state Eye PAC, you help elect officials in your state who will support the interests of you and your patients. Giving to *each* of these three funds is essential to helping protect sight and empower lives.

Protecting quality patient eye care and high surgical standards is a "must" for everybody. Our mission of "protecting sight and empowering lives" requires robust funding of both OPHTHPAC and the Surgical Scope Fund. Each of us has a responsibility to ensure that these funds are strong so that ophthalmology continues to thrive and patients receive optimal care.

OPHTHPAC for Federal Advocacy

OPHTHPAC is the Academy's award-winning, non-partisan political action committee representing ophthalmology on Capitol Hill. OPHTHPAC works to build invaluable relationships with our federal lawmakers to garner their support on issues such as:

- **Improving** the Medicare payment system, so ophthalmologists are fairly compensated for their services, and working to prevent impending payment cuts of 3.36% scheduled to take effect in 2024
- **Securing** payment equity for postoperative visits, which will increase global surgical payments
- **Stopping** optometry from obtaining surgical laser privileges in the veterans' health-care system
- **Increasing** patient access to treatment and care by reducing prior authorization and step therapy burdens

Academy member support of OPHTHPAC makes all this possible. Your support provides OPHTHPAC with the resources needed to engage and educate Congress on our issues, helping advance ophthalmology's federal priorities. Your support also ensures that we have a voice in helping shape the policies and regulations governing the care we provide. Academy member support of OPHTHPAC is the driving factor behind our advocacy push, and we ask that you get engaged to help strengthen our efforts and make sure that the ophthalmology specialty has a seat at the table for the critical decisions being made that affect our ability to care for our patients.

At the Academy's annual Mid-Year Forum, the Academy and the North American Neuro-Ophthalmology Society (NANOS) ensure a strong presence of neuro-ophthalmologists to support ophthalmology's priorities. As part of this year's meeting, NANOS supported participation of fellowship trainees via the Academy's Advocacy Ambassador Program. During Congressional Advocacy Day, they visited Members of Congress and their key health care staff to discuss ophthalmology priorities. NANOS remains a crucial partner with the Academy in its ongoing federal and state advocacy initiatives.

Surgical Scope Fund (SSF) for State Advocacy

The Surgical Scope Fund works in partnership with state ophthalmic societies to protect patient safety from dangerous optometric surgery proposals through advocacy. The Fund's mission is to ensure surgery by surgeons, and since its inception, it has helped 43 state/territorial ophthalmology societies reject optometric scope-of-practice expansions into surgery.

Support for the Surgical Scope Fund from ophthalmic interest societies like the North American Neuro-Ophthalmology Society makes our advocacy efforts possible. These efforts include research, lobbyists, political organization, polling,

Surgical Scope Fund	OPHTHPAC®	State EyePAC
To protect patient safety by defeating optometric surgical scope-of-practice initiatives that threaten quality surgical care	Working across the political spectrum to advance ophthalmology and protect its members and patients at the federal level Support for candidates for U.S. Congress	Support for candidates for state House, Senate and governor
Political grassroots activities, government relations, PR and media campaigns No funds may be used for campaign contributions or PACs.	Campaign contributions, legislative education	Campaign contributions, legislative education
Contributions: Unlimited Individual, practice, corporate, and organization	Contributions: Personal contributions are limited to \$5,000. Corporate contributions are confidential.	Contribution limits vary based on state regulations.
Contributions are 100% confidential.	Personal contributions of \$199 or less and all corporate contributions are confidential. Personal contributions of \$200 and above are public record.	Contributions are on the public record depending upon state statutes.

advertising, social media, digital communications, and grassroots mobilization. However, the number of states facing aggressive optometric surgery legislation each year has grown exponentially. And with organized optometry's vast wealth of resources, these advocacy initiatives are becoming more intense—and more expensive. That's why ophthalmologists must join together and donate to the Surgical Scope Fund to fight for patient safety.

The Academy's Secretariat for State Affairs thanks NANOS for its past support of the Surgical Scope Fund and looks forward to its 2023 contribution. The North American Neuro-Ophthalmology Society's support for the Surgical Scope Fund is essential to fighting for patient safety and quality eye care!

State Eye PAC

The presence of a strong state Eye PAC providing financial support for campaign contributions and legislative education to elect ophthalmology-friendly candidates to the state legislature is critical as scope of practice battles and many regulatory issues are all fought on the state level.

Support Your Colleagues Who Are Working on Your Behalf

Two Academy committees made up of your ophthalmology colleagues are working hard on your behalf. The OPHTHPAC Committee continues to identify Congressional Advocates in each state to maintain close relationships with federal legislators to advance ophthalmology and patient causes. The Surgical Scope Fund Committee is raising funds used to protect Surgery by Surgeons during scope battles at the state level.

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Section II: Swollen Optic Nerve

Young Patient With Acute Vision Loss and Swollen Nerve

Neena R Cherayil MD

CASE PRESENTATION

History and Exam

A 35-year-old Black woman presented with right eye pain and vision loss. Ten days prior to presentation, she noted throbbing right eye pain that progressed in intensity over several days and became worse with eye movements. Two days prior to evaluation, she awoke with dim vision centrally in the right eye, “like a smudge,” and presented to an optometrist, where visual acuity was 20/40 OD with noted red desaturation and a right relative afferent pupillary defect (RAPD) but intact visual field to confrontation. Fundus examination revealed right optic disc swelling.

Her past medical history was notable for well-controlled hypertension, uterine fibroids, and benign glomus tympanicum tumor that was fully resected. She worked from home in human resources and did not use tobacco or drugs or drink alcohol. Family history was noncontributory.

A month prior to the onset of symptoms, she had received the Pfizer COVID-19 booster shot and an influenza vaccine. She denied any sick contacts, animal exposures, or recent travel. She denied a history of headache, prior episodes of vision loss, pain on eye movements, diplopia, vertigo, dysarthria, dysphagia, weakness, numbness, paresthesias, difficulty with balance or walking, or bowel/bladder disturbances.

On examination 1 day after the optometry visit, visual acuity was 20/200 in the right eye, with 1.5/11 Ishihara color plates and right RAPD. Humphrey 24-2 SITA Fast noted diffuse suppression OD and normal OS. Slit-lamp examination was notable for normal anterior chamber examination, without cell or flare. Dilated fundus examination revealed 270-degree right optic disc swelling without disc hemorrhage or pallor. Afferent and efferent examination of the left eye was normal.

Clinical Course and Outcome

The patient refused presentation to the ER and high-dose steroids due to concerns around an ongoing surge in COVID cases. Urgent contrast MRI of the brain and orbits completed the following day revealed asymmetric enlargement and T2 hyperintensity of the right optic nerve, with associated enhancement of the entire prechiasmatic optic nerve and nerve sheath extending to the insertion on the globe. There was no abnormal signal or enhancement of the optic chiasm. There were scattered, non-specific 1-2 millimeter areas of subtle T2 hyperintensities in the periventricular white matter without enhancement. Her vision on the day of MRI had reduced to count fingers at 1 foot in the right eye, with slightly worsening optic disc swelling.

She was treated with 1000 mg of IV methylprednisolone for 5 days with significant improvement in right eye pain and visual acuity to 20/50 with 2.5/11 Ishihara color plates; she was started on oral prednisone 70 mg daily. MRI of the cervical and thoracic spinal cord was unremarkable. Serum cell-based assay of anti-aquaporin 4 antibodies for neuromyelitis optica (NMO) was negative, but anti-myelin oligodendrocyte glycoprotein (MOG) antibodies returned highly positive, with a titer of 1:1000, confirming the diagnosis of MOG antibody-associated disease (MOGAD).

She was treated with a 3-day course of IV immunoglobulin (IVIG), and her vision improved to 20/20 with 9/11 Ishihara color plates with resolving optic disc swelling. Oral prednisone was tapered by 10 mg per week until off. She is continued on monthly maintenance IVIG.

Older Patient With Vision Loss and Swollen Nerve

Marc H Levin MD

CASE PRESENTATION

History and Exam

A 70-year-old male with uncontrolled hypertension, coronary artery disease and congestive heart failure presented with 1 month of insidious onset of intermittent photopsias. BCVAs were 20/20-1 and 20/50-1, right and left eyes, respectively. He was found to have a relative left afferent pupillary defect with unilateral inferior left optic nerve head swelling with nerve fiber layer hemorrhage. Nonarteritic anterior ischemic optic neuropathy was felt most likely given a crowded contralateral right optic disc and vascular risk factors. Two months after symptom onset, there was resolving disc edema and adjacent nasal macular atrophy and fine lipid deposits, which along with cecentral scotoma raised suspicion for neuroretinitis. Serologies for infectious causes were ordered but not drawn until 4 months after symptom onset. At that time, visual acuity was 20/80 OS and there was persistent inferior disc edema along with evolving temporal disc pallor.

Clinical Course and Outcome

Serum RPR test returned as positive (1:256), whereas *Bartonella*, *Borrelia*, and *Toxoplasma* testing was negative, and the patient was admitted for further workup of and treatment for neurosyphilis. He denied a history of rash or genital lesions, and human immune deficiency virus (HIV) testing was negative. Serum fluorescent treponemal antibody-absorption (FTA-ABS) test was positive. Lumbar puncture (LP) showed 5 white blood

cells (WBC)/cm³, 4 red blood cells (RBC)/cm³, 161 mg/dL protein, and 72 mg/dL glucose in cerebrospinal fluid (CSF). Venereal Disease Research Laboratory testing was positive (1:2). Contrast orbital MRI was notable for equivocal focal left optic nerve head enhancement and a contralateral right-sided T2 hyperintense intraconal orbital lesion between the inferior rectus and optic nerve, possibly a benign vascular lesion but suspicious for a necrotic granulomatous gumma. He was discharged to complete a 2-week course of intravenous (IV) penicillin G (4 million units every 4 hours) with recommendation to repeat lumbar puncture afterward as test of cure.

65-Year-Old With Vision Loss and Swollen Nerve

Anita A Kohli MD

CASE PRESENTATION

History and Exam

A 65-year-old man developed double vision, which occurred 1 week prior to presentation. Four days later, he developed blurred vision in the left eye, and the next morning, he woke up with complete vision loss of the left eye, which had not improved at the time of presentation. Vision in the right eye remained at baseline. He reported feeling well. However, on direct questioning, he reported mild fatigue, scalp tenderness, and right arm weakness for the last month. Two weeks prior, he noted that he had to take breaks while eating due to jaw pain. He denied snoring but had never been evaluated for sleep apnea.

His past medical history was notable for well-controlled hypertension, hyperlipidemia, and hypothyroidism, for which he was on stable doses of medications. He was a nonsmoker and drank wine occasionally. The ocular history was significant for early-stage primary open-angle glaucoma (on latanoprost [Xalatan]), and he had a normal eye examination a month prior.

On examination at the urgent visit, the BCVA was 20/20 OD and light perception OS, with loss of color vision OS and presence of a left afferent pupillary defect. Visual fields to confrontation were full OD and completely depressed OS. The IOP was 11 OU. He was pseudophakic in both eyes, and the posterior segment evaluation showed a normal right optic nerve, with cup-to-disc ratio of 0.45. The left optic nerve was pale and swollen, with a peripapillary cotton wool spot. The retinal vessels and peripheries were normal OU. Humphrey 24-2 SITA Standard visual field was normal in the right eye.

Clinical Course and Outcome

The patient was referred directly from the eye clinic to the emergency department for further evaluation, and high-dose IV steroids were started on admission. The ED physicians obtained a CT head and CTA head and neck, which revealed a lacunar infarct in the right internal capsule and focal dissection of the right internal carotid artery, with probable fibromuscular dysplasia. MRI brain without contrast showed the previously noted chronic lacunar infarct and diffuse parenchymal volume loss. MRA neck confirmed the focal dissection of the right midcervi-

cal internal carotid artery, fibromuscular dysplasia, and patent intracranial arteries. ESR was 61 mm/h, CRP was 115.1 mg/L, and platelets were 684/ μ L. ANCA, ANA, HIV, and RPR serologic evaluations were negative. A temporal artery biopsy was obtained within a week of starting steroids, which confirmed giant cell arteritis. He was seen by neurology and was started on aspirin for carotid dissection and stroke. Rheumatology evaluated the patient and recommended an oral prednisone taper and initiation of tocilizumab as an outpatient. The previously noted fatigue, scalp tenderness, right arm weakness, and jaw claudication resolved while on steroids.

One month after presentation, the vision was 20/20 OD and bare light perception OS. The right optic nerve remained stable, and the left optic nerve edema resolved, with persistent severe pallor.

"I Have Eye Pain, and My Nerve Is Swollen"

Gregory P Van Stavern MD

CASE PRESENTATION

History and Exam

A 62-year-old woman presented for evaluation of visual blurring from the right eye of 2 weeks' duration. She stated that her visual blurring got worse over the course of 2 weeks but since had plateaued. The left eye was unaffected. She denied diplopia, ptosis, facial numbness or paresthesias, jaw claudication, and scalp tenderness, as well as recent constitutional or systemic symptoms.

Her past medical history was significant only for well-controlled hypertension and mild hyperlipidemia, for which she was on medications. There was no significant tobacco or alcohol use history.

Ophthalmic examination revealed vision of 4/200 OD, 20/20 OS. Ishihara color plate testing showed no control plate OD and 14/14 plates OS. There was a right afferent pupillary defect. IOP was normal. External examination was normal without ptosis or proptosis. Extraocular motility showed mild limitation of ductions in all directions in the right eye and normal motility in the left eye. Alternate cover testing showed a small intermittent esotropia in primary gaze. The rest of her cranial nerve examination was unremarkable. Slit-lamp examination was remarkable only for mild nuclear sclerosis OU. Dilated fundus examination showed mild, diffuse optic disc swelling OD and normal disc OS with normal maculae, vessels, and periphery. Humphrey SITA Standard visual field with size V stimulus showed a dense central scotoma in the right eye and was normal for the left eye using the size III stimulus. OCT showed mild, diffuse retinal nerve fiber layer thickening OD, and normal OS. Macular OCT was normal. Ganglion cell complex showed mild focal superior thinning OD and was normal OS.

Clinical Course and Outcome

Patient was admitted for an expedited workup. MRI of brain and orbits with and without gadolinium showed unremarkable brain aside from nonspecific white matter changes. Orbital MRI showed mild enlargement and enhancement of the extraocular muscles, with perineural enhancement of the right optic nerve and enhancement of the orbital fat in the right orbital apex. These findings were suggestive of orbital inflammatory syndrome, either idiopathic or secondary to a systemic disease.

Laboratory workup showed normal ESR, CRP; negative RPR, HIV, ANCA, neuromyelitis optica, myelin oligodendrocyte glycoprotein; positive ANA; positive but nonspecific ENA. CT chest/abdomen/pelvis was normal, with no evidence of sarcoidosis or malignancy.

She was treated with high-dose intravenous corticosteroids and then placed on an oral prednisone taper. She had dramatic resolution of her ophthalmic symptoms, with return of vision, color, motility, and visual fields. However, she developed severe steroid-related adverse effects, including steroid-induced diabetes.

Follow-up orbital MRI showed resolution of the inflammatory changes noted previously.

“My Nerve Is Swollen, and I Cannot See”

Laura Bonelli MD

CASE PRESENTATION

History and Exam

A 55-year-old woman presented for evaluation of decreased vision and visual field loss in the left eye.

She first noticed changes in her vision approximately 6 months previously and described it as seeing a “blurred spot in the lower part of the visual field of the left eye.” Visual loss was gradual in onset and had slowly worsened over time. She denied pain with eye movements, headaches, and other associated symptoms. She had no significant ocular history except for myopia.

Past medical history was positive for hyperlipidemia treated with atorvastatin. She drank a glass of wine 3 times a week and did not smoke or use drugs.

On examination, BCVA was 20/20 OD and 20/50 OS. She correctly identified 14/14 Ishihara color plates OD and 1/14 correct OS. Right pupil was briskly reactive, and left pupil was sluggish with 3+ relative afferent pupillary defect (RAPD). Visual field to confrontation was full in the right eye and revealed an inferior defect in the left eye. Ocular motility was full, with no misalignment. There was no proptosis or eyelid malposition. Slit-lamp exam showed normal anterior segment, 1 + NS both eyes, and IOP measured 14 mmHg in each eye.

On dilated fundus exam, the right optic disc was normal and the left showed mildly pale edema. The remainder of the fundus exam was unremarkable bilaterally.

Humphrey 30-2 perimetry was normal for the right eye and showed an inferior paracentral scotoma involving fixation for the left eye. OCT peripapillary retinal nerve fiber layer average thickness measured 88 microns OD and 104 microns OS.

Clinical Course and Outcome

Contrast-enhanced MRI of the orbits with fat suppression showed circumferential thickening and enhancement along the left optic nerve sheath (tram-track sign) including the optic canal, suggestive of optic nerve sheath meningioma.

The patient was referred to radiation oncology for evaluation and treatment of presumptive optic nerve sheath meningioma. She underwent stereotactic fractionated radiotherapy in 28 sessions to a total of 50.4 Gy.

Following treatment, the patient’s visual acuity and visual field improved in the left eye but reduced color vision and RAPD remained unchanged. The left optic disc edema resolved, with atrophy developing over months.

Section III: Neuro-Ophthalmic Mimickers and Visual Disturbances

“I Am Blinded by the Light”

Susan P Mollan MB ChB

CASE PRESENTATION

History and Exam

A 57-year-old woman was referred to the neuro-ophthalmology clinic for excessive photophobia and medically unexplained visual loss. Following her worst ever migraine attack, she had suffered from persistent photophobia. She reported having to wear dark glasses due to intolerance of bright light. This difficulty of seeing clearly in the light had stopped her from driving and caused her problems using a computer screen at work. The past medical history included prior episodic migraine with aura, which had occurred following her menarche. She took high-dose ibuprofen as required for her episodic migraine and sertraline for depression. There was no family history of neurologic or ophthalmologic problems. She was a nonsmoker and consumed alcohol only on special occasions.

On examination, the BCVA was 20/60 in her right eye and 20/40 in her left eye. There was no pinhole improvement. The pupils were equal, round, and reactive to light, without a relative afferent pupillary defect. Color vision was tested with the Ishihara plates and was slow and reduced in both eyes to 13 plates out of 17. Visual field testing to confrontation was initially normal; however, Amsler grid testing revealed a small paracentral scotoma in the right eye. There was no evidence of blepharospasm. Slit-lamp examination revealed only mild uptake with lissamine green staining on the conjunctiva and an otherwise normal corneal examination. There was no anterior or posterior ocular inflammation, and the IOPs were 11 mmHg and 13 mmHg in the right and left eye, respectively. Dilated posterior segment evaluation was normal, with normal optic discs, maculae, vessels, and periphery. Cranial nerve examination was unremarkable, including ocular motility and corneal sensation. Humphrey 24-2 SITA Fast visual field was felt to be essentially normal in both eyes.

Clinical Course and Outcome

The past medical examinations and investigations were reviewed. These included documentation of fluctuating visual acuity measurements and normal OCT imaging of the maculae in the ophthalmology clinic. She had been reviewed by neurology, who had performed an electroencephalogram (EEG) and MRI brain, neither of which diagnosed any pathology.

At the neuro-ophthalmology review, fundus autofluorescence imaging with Optos was normal in both eyes. Patient was referred for electrodiagnostic testing, where her full-field electroretinogram (ERG) and visual evoked potentials were normal. The pattern ERG was reduced in the right eye.

“I Have Normal Vision, but My Optic Nerves Are Pale”

Tatiana Bakaeva MD PhD

CASE PRESENTATION

History and Exam

A 20-year-old female college student was referred to the neuro-ophthalmology clinic for incidental discovery of bilateral optic nerve pallor on a routine eye exam. Three years prior to her presentation she saw an optometrist for contact lens evaluation and was found to have mild pallor of both optic discs. Her vision was normal. She was referred to an outside hospital, where she underwent CT and MRI of the brain that were reportedly normal. One month prior to presentation she saw an outside ophthalmologist for a routine eye exam, was noted to have visual field defects in both eyes and was referred to the neuro-ophthalmology clinic.

Her past ocular and medical history were unremarkable. She occasionally drinks alcohol, does not smoke, and does not take any medications.

On examination, BCVA was 20/20 in each eye, and she had normal color vision and counted fingers in all quadrants with each eye. Pupils were equal, round, and reactive to light without relative afferent pupillary defect. IOP was normal in both eyes. External exam and anterior segment exam were normal.

Dilated funduscopy exam showed mild temporal pallor of both optic discs, more prominent on the left. Humphrey visual field testing showed nonspecific peripheral defects infratemporally in the right eye and inferior arcuate visual field defect in the left eye.

“My Vision Is Normal, but I Cannot See”

Rudrani Banik MD

CASE PRESENTATION

History and Exam

A 26-year-old male presents with blurry vision in both eyes, which he describes as “loss of sharpness.” The patient is a professional ice hockey player and reports that his visual symptoms developed immediately after sustaining a concussion with brief loss of consciousness. He now has difficulty seeing the puck on the ice, as it appears blurred and “not clear.” He also notes

trouble reading on screens and seeing at night, especially when driving. He also complains of severe light sensitivity, persistent headache, and new-onset floaters.

His past medical history is significant for multiple sports injury–related concussions in the past, with at least 5 prior episodes. None required hospitalization, though after his concussions he developed episodic migraine. His surgical and ocular histories are negative. He takes no medications and has no allergies. He reports social alcohol use and denies smoking or illicit drug use. His family history is significant for migraine in his mother and sister.

Examination shows uncorrected visual acuity of 20/20 in both eyes at both distance and near. Pupils are equal, with no anisocoria or relative afferent pupillary defect. Color vision is full in both eyes with HRR plates. Confrontational visual fields are full. Extraocular motility is full, and ocular alignment is normal via alternate cover testing. Slit-lamp exam is normal. Dilated fundus examination is normal, with sharp optic disc margins and normal cup-to-disc ratio of 0.3 OU. Macula, vessels, and periphery are normal. There is mild vitreous syneresis but no posterior vitreous detachment or other vitreous debris. Humphrey visual field showed mild generalized depression in both eyes with no focal defects. OCT retinal nerve fiber layer, ganglion cell analysis, and OCT macula are normal.

Clinical Course

The patient was initially seen by an optometrist, who did not find any abnormalities on exam. He was then referred to a comprehensive ophthalmologist, who also did not find any abnormalities. He was then seen by a retina specialist, who ordered fluorescein and indocyanine green angiography and microperimetry, all of which were normal. Electrophysiologic testing including electroretinogram (ERG), multifocal ERG, and visual evoked potential were performed and were normal.

The patient's symptoms progressively worsened with respect to his blurry vision, light sensitivity, and floaters. He now reports that he can no longer play hockey because "everything is blurred, and moving." He also sees afterimages of other players and trailing images of the puck. His neuro-ophthalmic exam remains normal, with acuity of 20/20 OU and normal retinal exam. The patient also has developed tingling of his right arm and leg and was seen by a neurologist, who found him to have a normal neurologic exam. MRI Brain with and without contrast was normal. He also complains of a high-pitched buzzing sound in his ears that is constant. ENT and audiology evaluations are negative.

"I Have a Visual Field Defect, but My Nerve Looks Normal"

M Tariq Bhatti MD

CASE PRESENTATION

History and Exam

A 74-year-old man was followed for 2 years by an ophthalmologist as a glaucoma suspect with asymmetric optic disc cupping (right greater left). At the initial visit, visual acuities were 20/20 OD and 20/25 OS. IOPs were 17 mmHg OU. Automated static perimetry revealed scattered defects OD and superior and inferior arcuate scotomas OS. Repeat automated perimetry at 8 months, 14 months, and 16.5 months later demonstrated possible progression of the visual field defects OS but with the appearance of the optic nerves, visual acuity, and IOP remaining stable.

The patient's only visual complaint was a long-standing problem with glare and difficulty with driving at night. He did not particularly notice any problems with the vision in the left eye and denied any systemic or neurological symptoms.

His medical history was notable for hypertension, hyperlipidemia, gout, osteoarthritis, nephrolithiasis, and multiple cutaneous basal cell carcinomas. He was taking aspirin, atenolol, lisinopril, atorvastatin, naproxen, and allopurinol.

His ocular history was notable for early cataracts OU and a tonic pupil OD dating back 12 years. He denied any family history of glaucoma or other eye disease.

Clinical Course and Outcome

Neuro-ophthalmic consultation was requested because of the persistent visual field defect OS. Visual acuity was 20/20 OD and 20/25 OS. IOPs were 12 mmHg OD and 13 mmHg OS. Color vision was intact OU. The right pupil was slightly larger than the left and poorly reactive to light but reactive to a near stimulus, consistent with a tonic pupil. There was no relative afferent pupillary defect. Slit-lamp biomicroscopy revealed 1+ nuclear sclerosis with anterior cortical spokes OU. The cranial nerve examination was normal. There was no scalp tenderness. Funduscopic examination revealed mild optic disc asymmetry with a cup-to-disc ratio of 0.5 OD and 0.3 OS. The neuroretinal rims were intact OU, with no notching or pallor. There were epiretinal membranes OU, and the retina appeared elevated along the superior and inferior arcades OS. Automated perimetry demonstrated an essentially full visual field OD and superior and inferior arcuate scotomas OS.

OCT was obtained, which showed thickening of the peripapillary retinal nerve fiber layer (pRNFL) of the superotemporal and inferotemporal quadrants in the left eye. The radial macular scans OS revealed significant traction at the vitreoretinal interface in the temporal peripapillary region and along the superotemporal and inferotemporal arcades, with schisis of the underlying retina. There was a complete vitreous detachment over the fovea, which was normal in contour except for a mild epiretinal membrane. The OCT findings were consistent with vitreomacular traction, prompting a referral to the retina service for further management.

"I Have Light Sensitivity"

Samuel Spiegel MD

CASE PRESENTATION

History and Exam

A 30-year-old male with past medical history of migraine presents for evaluation of sensitivity to light. He describes persistent light sensitivity that has progressed over the last 3 months and is now bothersome in any well-lit environment. He works as a computer programmer, and it is starting to affect his efficiency. He has adjusted his computer display brightness and is now working fully from home because the office lighting is extremely bothersome. When he does have to go into the office, he wears sunglasses because he isn't able to dim the lights like he does at home. Outside of work, he finds himself limiting participation in events and experiencing anticipatory stress due to fear of being exposed to triggering light conditions. When you ask about his migraine history, he notes they had been worse a few months ago but are improving. He takes ibuprofen, and his PCP restarted sumatriptan, which he hasn't used since college. Last month he had a few migraines, but they are improving.

On examination, BCVA is 20/20 with each eye. Pupils are equal, round, and reactive to light without afferent pupillary defect. IOP is 14 in each eye. Ocular motility is full. Anterior segment examination is remarkable only for moderately reduced tear film breakup time, with trace punctate epithelial erosions. Dilated posterior segment evaluation is normal, with sharp optic nerve margins, 0.3 cup-to-disc ratio, and healthy rim color in both eyes, and with normal maculae, vessels, and periphery. Cranial nerve examination is unremarkable. Humphrey 24-2 SITA Fast visual field is normal in both eyes, as are OCTs of the optic nerves and maculae.

Clinical Course and Outcome

You review your findings with the patient and recommend a multimodal treatment approach, including lubricant eye drops, use of FL-41 glasses, and establishing a gradual return to normal light conditions with minimization of sunglasses indoors. Additionally, you review nonpharmacologic lifestyle modifications for eye strain and headaches, such as breaks with prolonged screentime, adequate hydration, sleep hygiene, regular exercise, and keeping a healthy diet. You recommend that he consider seeing neurology for his migraine if this is contributing.

The patient likes the idea of special tinted lenses and agrees that he could work on lifestyle adjustments. He specifically mentions poor sleep and inability to exercise ever since his bad bicycling accident a few months ago. Between this and his light sensitivity, he feels stressed and cognitively fatigued. You provide reassurance that this is not uncommon, and that he should expect to make slow progress with the recommendations you have provided.

His PCP orders an MRI, which is normal, and refers him to neurology. His migraine medications are adjusted slightly, and his headaches have remained similar, occurring a few times per month.

At your 3-month follow-up encounter, he reports that his symptoms have gradually improved and although still present, they are much more manageable. He is back in the office, wearing FL-41 lenses, and able to go out with friends more often. He mentions he started seeing a therapist and is in a much better place mentally. He thinks he is headed in the right direction and expresses his gratitude. He feels his quality of life is much improved.

Section IV: Double Vision—What to Do?

Case of Acute Double Vision in a Younger Patient

Lauren C Ditta MD

CASE PRESENTATION

History and Exam

A 47-year-old man presented for evaluation of recent onset diplopia, worse in downgaze and lateral gaze. These symptoms were noticed one morning upon awakening 2 weeks prior to presentation. He described the images as being one on top of the other and slightly angled, and he felt his symptoms were stable since onset. He denied headaches, neck stiffness, associated pain, or blurred vision.

The patient was in excellent health, and he had no past medical history of significance. Family and social histories were also unremarkable. Laboratory work ordered by his PCP, whom he saw initially, yielded a positive Lyme titer by Western blot. The patient denied any history of tick bites, joint pain, or fevers; however, he did note recent fatigue. He also mentioned that as a child he was told that he had a “wandering eye.” He denied any prior patching or treatment for this.

On examination his visual acuity was 20/20 OU, with normal color and normally reactive pupils without an afferent pupillary defect. Visual fields by confrontation were full. He appeared to have a right head tilt. Ocular motility testing revealed full ocular ductions and versions. On prism alternate cover test, he had a 5 Δ left hypertropia (LHT) in primary gaze, which increased in right gaze (8 Δ LHT), downgaze (6 Δ LHT), and on left head tilt (6 Δ LHT). There was no obvious excyclo-torsion. He was able to fuse with 4 Δ of base-down (BD) prism over the left eye.

Clinical Course and Outcome

The clinical impression was that this patient had a left cranial nerve IV palsy. He was referred for MRI of the brain with and without contrast, which was unremarkable and showed no structural lesion in the brain. Lab work for thyroid and myasthenia gravis was negative. Given he was healthy, without a history of diabetes or hypertension, an ischemic event as causative etiology was felt to be less likely. It was difficult to relate his positive Lyme titer to his cranial nerve palsy, as he was otherwise asymptomatic. Infectious disease was consulted for treatment recommendations and felt that, given a negative serum immunoglobulin G (IgG), there was no concern for late or chronic Lyme disease. However, a conservative course of treatment with doxycycline 100 mg b.i.d. was recommended for the low positive IgM antibody (0.92), along with repeat serum antibody testing. The patient opted for conservative treatment for his diplopia, and prisms were placed in his reading glasses

(2 Δ base-up [BU] OD and 2 Δ BD OS) for diplopia in downgaze, which was causing him difficulty reading.

When the patient returned for re-evaluation, he continued to have a compensatory head posture, and he felt the prisms helped minimally. Clinical examination was stable. Repeat blood-work for Lyme was normal by enzyme-linked immunosorbent assay. He was referred to neurology and a second MRI was ordered with high-resolution imaging, including cranial nerve sequences. This revealed a focal area of enhancement along the left margin of the midbrain at the expected course of the fourth cranial nerve, felt to represent a schwannoma.

The patient was referred to neurosurgery and radiation oncology, and gamma knife radiosurgery was recommended. The rationale, risks, benefits, and alternatives of radiation therapy were explained to the patient, and he agreed to proceed with treatment. Four months after symptom onset, he received treatment for his (presumed) schwannoma of the left trochlear nerve with gamma knife radiosurgery (12.5 Gy). Following the procedure, his double vision resolved in primary gaze but persisted in right gaze and downgaze. He had a residual 2 Δ LHT in primary position, which increased in downgaze (5 Δ LHT). New glasses with prism were prescribed (1 Δ BU OD and 1 Δ BD OS). Over time, both the clinical examination and neuroimaging of the lesion have remained stable.

Case of Acute Double Vision in an Older Patient

Crandall E Peeler MD

CASE PRESENTATION

History and Exam

A 61-year-old man presented for evaluation of new-onset horizontal double vision beginning 3 days prior to presentation. He reported that the double vision worsened when looking to his right. He denied any preceding injury, illness, or associated symptoms, such as numbness or weakness. However, he did note intermittent right-sided tinnitus for 1 month and mild swelling and tenderness behind his right ear beginning 2 days prior to presentation.

His past medical history was significant for systemic hypertension, hyperlipidemia, and type 2 diabetes. His daily medications included atorvastatin 40 mg, hydrochlorothiazide 12.5 mg, and lisinopril 40 mg. His diabetes was diet controlled. His past ocular history included a branch retinal vein occlusion in the right eye 4 years prior and a right sixth nerve palsy 2 years prior with spontaneous resolution of diplopia after 10 weeks. He did not smoke or consume alcohol. He worked as an automobile mechanic.

On examination, BCVA was 20/20 in both eyes. Pupils were equal, round, and reactive to light without a relative afferent pupillary defect. IOP was 12 on the right and 14 on the left. Slit-lamp examination was normal other than early nuclear sclerosis. Dilated posterior segment evaluation demonstrated pigment mottling of the superior macula on the right but was otherwise unremarkable. Ocular motility examination revealed a -2 abduction deficit on the right with an esotropia of 25 prism diopters in right gaze, 10 prism diopters in primary gaze, and 2 prism diopters in left gaze.

Clinical Course and Outcome

An MRI of the brain with and without contrast showed complete opacification and enhancement of the right mastoid air cells and a portion of the middle ear cavity. The enhancing tissue in the mastoid air cells was abutting the right sigmoid sinus, likely representing bony erosion or dehiscence. Additionally, there was diffuse enhancement of the right tentorium, pachymeningeal enhancement overlying the right temporal, parietal, and occipital lobes, and leptomeningeal enhancement of the right cerebellum. A CT of the temporal bones also demonstrated an occlusive thrombus of the right transverse and sigmoid sinuses and jugular bulb.

The patient was admitted to the hospital for treatment of presumed infectious mastoiditis and received intravenous cefepime, vancomycin, and ampicillin/sulbactam. The following day he was taken to the operating room by otolaryngology for a right mastoidectomy. Intraoperatively, no purulent material was found. The entire mastoid was filled with a soft tissue mass with a flesh coloration and consistency, which was removed and sent for culture and pathology. Antibiotics were discontinued 2 days later with no growth on culture (including testing for acid-fast bacilli).

Pathology results from the biopsied mastoid tissue showed storiform fibrosis and a lymphoplasmacytic infiltration with an immunoglobulin G4 (IgG4) to IgG ratio of 48%. There was no evidence of lymphoma or plasma cell neoplasm by flow cytometry or immunohistochemistry. Serum levels of IgG4 subclass were elevated to 99.0 mg/dL (reference range: 4.0-86.0 mg/dL). He was discharged on a tapering dose of oral prednisone, and therapy with rituximab was initiated 2 weeks later. Six weeks after initial presentation, his ocular motility and alignment had returned to normal and his double vision resolved completely.

Cases of Esotropia With Sixth Nerve Palsies, Sagging Eye, Heavy Eye: When to Image

Zoë R Williams MD

CASE PRESENTATION

History and Exam

A 57-year-old woman presents with an 8-month history of binocular horizontal diplopia with progressively worsening diplopia. She denies associated weakness, numbness, or imbalance. Her diplopia is present for distance only and does not worsen in any particular field of gaze. She denies diurnal fluctuation of her diplopia. She was prescribed Fresnel prism (12 PD BO) 4 months ago. She denies any history of strabismus surgery or amblyopia. Past medical history is noncontributory. She has no history of cancer. She has been treated for idiopathic anterior uveitis in the right eye, with a negative workup. Her uveitis was treated with prednisone 60 mg daily for 6 weeks without improvement of her diplopia. Prior workup for her diplopia included negative laboratory screening for myasthenia gravis and giant cell arteritis and an unremarkable MRI brain without contrast.

On examination, BCVA was 20/20 in the right eye and 20/30 in the left eye. Refractive error was -5.25 sphere OD and -6.00 sphere OS. Pupils were equal, round, and reactive to light without a relative afferent pupillary defect. Color vision was full OU. Tangent screen was normal bilaterally. IOP was 15 in each eye. Ocular motility testing was full, with brisk saccades and smooth pursuit. Ocular alignment testing (with prismatic correction removed) showed a comitant esotropia of 20 PD for distance and an esotropia of 10 PD for near. External examination showed bilateral ptosis with marginal reflex distance-1 of 2 mm bilaterally with normal levator function. There was no increased ptosis with sustained upgaze or orbicularis oculi weakness. Exophthalmometry was 18 on the right and 19 on the left on a base of 97. Cranial nerve examination was unremarkable. Slit-lamp examination was remarkable only for 1+ punctate epithelial erosions in the left eye. Dilated posterior segment evaluation was normal, with sharp optic disc margins without pallor, 0.3 cup-to-disc ratio OU, and normal maculae, vessels, and periphery.

Clinical Course and Outcome

Patient's examination was suggestive of sagging eye syndrome, but given her relatively young age and her progressive diplopia over 8 months by history, a noncontrast MRI orbits was ordered. The lateral rectus-superior rectus bands demonstrated mild superotemporal bowing bilaterally. Mild irregularity of the left-sided lateral rectus-superior rectus band near the superior rectus attachment may be degenerative. However, the extraocular muscles, specifically the lateral rectus muscles, maintain normal size and positioning per radiology.

Her examination remained stable on follow-up 3 months later, and she elected to pursue strabismus surgery. Due to the lack of hypotropia despite asymmetric inferior displacement of the lateral rectus muscle pulley on MR image review,

only a small increase in target surgical dose was planned. She underwent a 4-mm bilateral medial rectus muscle recession. Six weeks postoperatively, she was orthophoric in all directions of gaze. At 1 year postoperatively, she remained asymptomatic but had a recurrent esotropia of 3 PD in right gaze, upgaze, and downgaze (3 PD esophoria in primary gaze).

Surgical Considerations in Diplopia

Double Vision Under Pressure

Ore-Ofeoluwatomi O Adesina MD

CASE PRESENTATION

History and Exam

A 63-year-old man with a past medical history of pre-diabetes developed horizontal binocular double vision, worse at distance than near, that progressively worsened for 1 year, ultimately requiring patching of the left eye to relieve his symptoms. He also developed associated headaches on the left side of his head and was seen by an ophthalmologist, who diagnosed a cranial nerve palsy. MRI brain with and without contrast demonstrated a left prepontine and cavernous sinus en plaque enhancing extra-axial mass, with encasement of the left cavernous carotid artery most consistent with a meningioma. On neuro-ophthalmic examination, his BCVA was 20/20 OD and 20/40 OS, with normal pupillary function and color vision. He had a 30 PD left esotropia (LET) and a small left hypertropia (LHT) in primary gaze, with an isolated complete -4 left abduction deficit. The rest of his examination was unremarkable with the exception of a left-sided choroidal nevus.

Clinical Course and Outcome

The patient was evaluated by neurosurgery and underwent uncomplicated radiosurgery. He was followed in the neuro-ophthalmology clinic, and 5 months after initial presentation had a 45-50 PD LET with a slightly worse abduction deficit. He was offered and received botulinum toxin injection to the left medial rectus (MR) to minimize contracture of the muscle, to be followed by strabismus correction when stable. Two months post-injection, his LET was 35 PD with a stable abduction deficit. His strabismus remained stable, and 9 months after receiving the injection, he underwent a modified Nishida procedure consisting of a 10-mm transposition of the superior rectus (SR) and inferior rectus muscles temporally at 12 mm from the limbus, equidistant to the lateral rectus, and a 6-mm recession of the left MR muscle. Postoperatively, his alignment improved, with 2-3 PD of LET and a stable small-angle LHT. He was essentially diplopia free, with mild diplopia at the end of the day and with fatigue. He was given glasses with 2 PD of base out prism with BCVA of 20/20 in each eye and was diplopia free. Serial imaging has revealed a stable meningioma.

Diagnosis and Teaching Points

Section I: What Meds Is My Patient On?

Adverse Reactions of Systemic Medications

“I Woke Up and Could Not See”

Michael S Lee MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Unilateral amiodarone-associated toxic optic neuropathy

Teaching Points

1. Amiodarone can cause a toxic optic neuropathy. Most cases of amiodarone toxicity show an insidious onset over the course of months, bilateral vision loss, and bilateral protracted optic disc edema for several months. After stopping amiodarone, the visual acuity and visual field deficits tend to stabilize.
2. There is debate about whether a unilateral optic neuropathy in a patient taking amiodarone is related to the medication or if the patient has incidental nonarteritic anterior ischemic optic neuropathy (NAION) instead. Reasonable cases have been reported in the literature of amiodarone causing unilateral optic disc edema, sequential optic disc edema, and retrobulbar optic neuropathy. In general, typical NAION will cause unilateral vision loss acutely or subacutely (days), and the optic disc edema often resolves in approximately 6 weeks. Patients who do not have those features are suspected of having amiodarone toxicity.
3. When a patient has unilateral or bilateral optic neuropathy, it is important to discuss with the cardiologist the possibility of stopping amiodarone. Many neuro-ophthalmologists will err on the side of caution and suggest stopping the amiodarone among patients who may have a clinical picture that looks like unilateral NAION.

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“I Have Melanoma, and I Am Seeing Double”

Dan R Gold DO

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

PD-1 inhibitor-associated myasthenia gravis

Teaching Points

Immune checkpoint inhibitor (ICI) medications including pembrolizumab, nivolumab, and ipilimumab are typically used for solid organ tumors, including non-small cell lung carcinoma, urothelial carcinoma, renal cell carcinoma, and melanoma, among others. Although neurotoxicity from PD-1 inhibitors is rare, reported neurological complications have included cranial neuropathies, multiple sclerosis, myasthenia gravis, transverse myelitis, myositis, Guillain-Barré syndrome, encephalitis, and meningitis, which notably, may affect either central or peripheral nervous systems. The most common presenting symptoms include weakness (eg, myasthenia gravis, myositis, transverse myelitis, Guillain-Barré syndrome), seizure (eg, encephalitis or meningitis), or change in mental status (eg, encephalitis).

Myasthenia gravis is a well-described autoimmune adverse event of checkpoint inhibitors, typically occurring within 1 to 16 weeks of initiation. Although ICI-induced MG (iMG) may look the same as traditional MG at the bedside from a motility (eg, any extraocular muscle may be involved) and eyelid (eg, Cogan lid twitch, fatigable, and variable ptosis) standpoint, iMG has distinctive features. Clinically, iMG patients often progress quickly and exhibit early (and potentially lethal) bulbar and respiratory symptoms and signs that require hospitalization, with nearly half requiring respiratory support. About one-third of iMG patients also experience ICI-induced myositis, which can cause or aggravate iMG-related weakness.¹⁻⁴

Diagnostically, although clinical suspicion should be high for iMG when diplopia and/or ptosis is present in a patient on an ICI drug, the diagnosis can be challenging to confirm from a laboratory and electrophysiologic standpoint. Acetylcholine receptor antibody testing is less sensitive in these patients, and when positive, titers can be comparatively low. Currently, there are no reports of positive muscle specific kinase (MUSK) antibodies in patients with pembrolizumab-induced myasthenia gravis. Characteristic neuromuscular junction/MG EMG findings are also present less than 50% of the time in iMG.

Therapeutically, when iMG is suspected steroids are initiated, and there should be multidisciplinary consideration of drug cessation with oncology. Discontinuation of the ICI may not be needed when the symptoms are mild and responsive to steroids. In cases with more severe sequelae, discontinuation of

the drug and initial therapy with intravenous immunoglobulin or plasma exchange may be needed, especially if the patient is steroid unresponsive.¹⁻⁴

Take-Home Points

1. When myasthenia gravis is suspected, a thorough review of the medical history including medications is essential (especially for solid organ tumors, PD-1 inhibitors, respectively).
2. Although patients with iMG may present similarly to those with traditional MG, in iMG, bulbar and respiratory symptoms occur earlier, and other immune-mediated toxicity syndromes may co-occur (eg, myositis, myocarditis). Therefore, these patients are at risk for significant morbidity and mortality.
3. Acetylcholine receptor antibodies are not typically detected in iMG, and when present, titers tend to be low.
4. Given the long pharmacodynamic and pharmacokinetic effects, inflammation can persist for weeks to months following cessation of ICIs such as pembrolizumab. Therefore, early initiation of immunotherapy, including steroids, intravenous immunoglobulin, and/or plasma exchange, is necessary to minimize the likelihood of severe or irreversible neurotoxicity.

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"I Am on Ethambutol— What Should I Look For?"

Nailyn Rasool MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Ethambutol optic neuropathy

Teaching Points

Ethambutol is important in current drug regimens for treating tuberculosis (TB) and *Mycobacterium avium* complex (MAC). It was developed in 1961 as a synthetic bacteriostatic antimycobacterial agent for individuals with TB. However, soon after its introduction, it was recognized to be associated with a partially reversible optic neuropathy that is both dose and duration dependent.^{1,2} Optic neuropathy is rare with doses of 15 mg/kg/day, but it can be seen with dose escalation greater than 25 mg/kg/day. At doses of 35 mg/kg/day, the incidence of optic neuropathy can be 18% or more, with higher risk seen in those with abnormal kidney function.^{3,4}

Patients with ethambutol optic neuropathy present with subacute bilateral painless loss of central vision with dyschromatopsia. Symptoms can begin anywhere from 1 month to 3 years following the initiation of treatment, most commonly between 4 and 12 months after beginning the medication.⁵ Classically, visual fields show a central or cecocentral defect, and dyschromatopsia is often found to involve the red-green axis; however, changes along the blue-yellow axis may also be seen.⁶ Rarely, a bitemporal visual field defect may be present.⁷ The optic nerve initially appears normal, with pallor developing at later stages. Ancillary testing such as visually evoked potentials (VEP) may demonstrate abnormalities in the p100 wave in amplitude and latency. OCT has demonstrated a decrease in retinal nerve fiber layer thickness in patients with significant vision loss. Its ability to detect subclinical disease is still uncertain.

Prevention of ethambutol optic neuropathy is key. Prior to beginning treatment with ethambutol, all patients should undergo a baseline screening examination including visual acuity, formal visual fields, color vision testing, and a dilated fundus examination. Patients must be well informed of the visual complications of the medication and advised to be vigilant about monitoring their vision periodically, checking visual acuity and central visual field on a home Amsler grid. They should be advised to present immediately should they notice a change in their vision and to inform their infectious disease specialist or internist. Follow-up screening should be performed monthly in high-risk patients (high dose, renal dysfunction, and unable to recognize subtle visual decline) and less frequently in lower-risk patients.

Currently, there is no evidence-based treatment for ethambutol optic neuropathy except prompt discontinuation of the medication. Up to 64% of patients demonstrate some improvement in their visual function months following cessation of ethambutol. Reversibility of the visual symptoms is in part age related,

as one study identified 20% visual improvement in patients over 60 years of age and up to 80% improvement in patients younger than 60.⁸ Because copper and zinc chelation by ethambutol is thought to play a role in its pathogenesis, zinc, copper, and cobalamin supplementation have been tried; however, this is not yet proven to have significant benefit.⁹

Ethambutol optic neuropathy is one of the most common drug-related optic neuropathies and may be reversible if diagnosed early and managed appropriately. Education, thorough evaluations, and frequent monitoring are essential to prevent visual loss.¹⁰

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"I Cannot See, and Now I Cannot Hear"

Peter W MacIntosh MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Sensorineural hearing loss from teprotumumab

Teaching Points

Thyroid eye disease (TED) has an incidence of 1.9 cases per 10,000 population per year,¹ with a female preponderance,² and occurs in about 40% of patients with autoimmune thyroid disease. Important risk factors for developing TED include poorly controlled thyroid disease, older age, and smoking,³ with the latter being the most important modifiable risk factor. It runs an active progressive phase with variable orbital inflammation for 2-3 years followed by an inactive stable phase.⁴

TED is known to be caused by autoantibodies targeting the thyrotropin receptor leading to hyperthyroidism. There are thought to be additional autoantigens and antibodies that drive the orbital findings in TED, including insulin-like growth factor I receptor (IGF-R). IGF-R is overexpressed by orbital fibroblasts and B and G cells in TED and thus is critical to its development.^{5,6}

Teprotumumab is a fully human monoclonal antibody that can block pathologic immune responses in active TED. In Phase 2 and 3 studies, patients with moderate to severe active TED experienced significant improvement in proptosis, double vision, soft tissue inflammation, and quality of life. During a randomized, double-masked, placebo-controlled Phase 3 multicenter trial of 41 patients assigned teprotumumab and 42 assigned placebo, most adverse events reported were of grade 1 or 2. Eighty-five percent of patients experienced adverse events, and 10% had hearing loss that was reversible upon stopping the drug. Of the 41 patients in the teprotumumab group, 5 had hearing impairment, but none of the 42 placebo patients did. In all cases, the hearing impairment resolved.⁷

While most studies during clinical trials found a 10% rate of hearing loss, with most or all of them reversible without intervention,^{7,8} subsequent case reports and series have reported long-term and potentially irreversible hearing loss.⁹⁻¹²

IGF-1 receptors are expressed in the cochlea on outer hair cells, and as such IGF-1 is an important player in the development, maintenance, and protection of the inner ear and hearing function.¹³ Inborn IGF-1 deficiency leads to profound sensorineural hearing loss in both humans and mice. Later in life, bioactive IGF-1 circulation levels are reduced, and this reduction is associated with human frailty and cognitive decline along with hearing loss.¹⁴ IGF-1 deficiency leads to increased inflammation and failure of intracellular cell renewal mechanisms. This process has important consequences in the inner ear as these cell types cannot regenerate.¹⁵

Studies have shown that topical IGF-1 therapy can have a positive effect on sensorineural hearing loss.¹⁶ These data support that blocking IGF-1 activity with treatments such as teprotumumab can in theory, and now in practice, lead to iatrogenic sensorineural hearing loss.

Teprotumumab is a novel treatment for TED, and there are scant comprehensive studies of its effect on hearing. As a result, many clinicians recommend prospective investigations of hearing loss in the setting of teprotumumab treatment, with a surveillance protocol to include an audiogram and tympanometry before, during, and after infusion, as well as when prompted by new symptoms of hearing dysfunction.¹⁰

While it is prudent to consider teprotumumab as the cause of hearing loss in any patient taking that medication, one must also rule out other causes of hearing loss. This includes ototoxic medications such as aminoglycoside antibiotics and asking about a history of head trauma and exposure to loud noises or explosions.

There is no proven treatment for teprotumumab-related hearing loss, and further studies are warranted. Clinicians may monitor closely or consider discontinuation of the medication. In one case report, oral steroids were given as a treatment based on the known mechanism of IGF-1-induced inflammation with resolution of symptoms, and the patient completed teprotumumab treatment.¹⁷ If there is no improvement after significant hearing loss, consideration can be given to sound amplification devices, as suggested for our patient.

Teprotumumab has been a welcome addition to the toolbox for the treatment of TED. We can expect many more patients to receive this medication and be exposed to its potential side effects, including hearing loss. Furthermore, our understanding of TED is evolving, and the role of teprotumumab in inactive TED is also under investigation,¹⁸ which could further expand the population of patients eligible for its use. Further studies to guide management of adverse effects will be necessary.

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“I Have Blurred Vision and Swollen Nerves”

Amanda D Henderson MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Pseudotumor cerebri (PTC) secondary to minocycline

Teaching Points

PTC is a potentially vision-threatening condition associated with increased intracranial pressure that occurs in the absence of a space-occupying intracranial lesion or venous sinus thrombosis. PTC can be primary or secondary. Pseudotumor cerebri syndrome is diagnosed based on the following:

1. Signs of increased intracranial pressure: papilledema or sixth nerve palsy or 3+ neuroimaging criteria, including empty sella, posterior globe flattening, optic nerve sheath distention, and transverse venous sinus stenosis
2. Absence of localizing findings on neurologic examination, with the exception of sixth nerve palsies
3. MRI brain without structural lesion or meningeal enhancement (MRV also required in atypical patients)
4. Normal CSF composition
5. Elevated lumbar puncture opening pressure¹

Cases of primary pseudotumor cerebri typically occur in obese women of child-bearing age. In patients not fitting these demographics (ie, a patient of normal weight, as in our case, or a male patient), secondary causes of increased intracranial pressure should be suspected. The first step in management of these cases is urgent neuroimaging to exclude space-occupying lesions or venous sinus thromboses. Once this has been completed, other underlying causes deserve consideration.

Secondary PTC may occur due to medication use, as in the case described. At the time of initial presentation, several potential factors contributed to the increased intracranial pressure, including obstruction, infectious or inflammatory CSF, and minocycline. However, through the evaluation and clinical course, minocycline was ultimately determined to be the most likely underlying cause.

In a recent meta-analysis evaluating cases of PTC secondary to medication use, vitamin A and its derivatives (including vitamin A supplementation, isotretinoin, and tretinoin) and tetracycline antibiotics (including doxycycline, tetracycline, and minocycline) were most often associated with the development of secondary PTC, followed by recombinant growth hormone and lithium.² Both tetracycline antibiotics and systemic retinoids are commonly used to treat acne vulgaris, rosacea, and other dermatologic conditions, and in fact, some patients may be treated with both of these medication classes simultaneously.³ All-trans retinoic acid (ATRA) is part of the standard treatment protocol for acute promyelocytic leukemia, and secondary PTC may develop in this setting. It has been suggested that the risk for PTC may be higher when ATRA is combined with arsenic trioxide (ATO).⁴

The mechanisms by which these medications contribute to the development of increased intracranial pressure have not been fully elucidated. Importantly, although some reports have suggested an association between oral contraceptive pills (OCPs) and PTC, this relationship has not been confirmed in well-designed case control studies, and thus the observed coincidence of OCP use and PTC is likely explained by the overlapping demographics of these 2 groups (ie, they are largely women of child-bearing age).⁵

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Section II: Swollen Optic Nerve

Young Patient With Acute Vision Loss and a Swollen Nerve

Neena R Cherayil MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Right optic neuritis in setting of myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOGAD)

Teaching Points

Optic neuritis (ON) is an inflammatory optic neuropathy typically characterized by subacute monocular vision loss often associated with orbital pain worse on eye movement, reduced contrast and color vision, and a relative afferent pupillary defect. ON is a clinical diagnosis, and Petzold et al recently proposed diagnostic criteria for ON to include both clinical and paraclinical features, including OCT, MRI, and serum antibody testing.¹

ON is perhaps best known for its association with multiple sclerosis (MS), where acute demyelinating ON is the first clinical event in 15%-25% of patients;² however, ON has a broad differential diagnosis and is associated with other demyelinating diseases, such as MOG antibody-associated disease (MOGAD) and neuromyelitis optica spectrum disorder (NMOSD). In addition, optic neuritis can be caused by a wide variety of infectious and parainfectious disease, including syphilis, Lyme disease, and neuroretinitis of any cause, as well as inflammatory disorders such as sarcoidosis, Behçet syndrome, granulomatosis with polyangiitis, systemic lupus erythematosus, and Sjögren disease. Paraneoplastic optic neuritis in the setting of antibodies to anti-collapsin-responsive mediator protein 5 (CRMP5) has been seen in patients with small-cell lung cancer and thymoma.¹

ON is the most frequent presenting feature of MOGAD.³ The MOG antibody was first discovered in 2007, but commercially available testing was only developed years later and the international diagnostic criteria for MOGAD was not published until 2023. The serum cell-based assay for MOG immunoglobulin has superior diagnostic accuracy over other assays, such as ELISA, which are nonspecific. Unlike MS and

NMOSD, MOGAD does not appear to have a racial or gender predilection. Moderate to severe optic disc swelling and bilateral involvement is more common in MOGAD-related ON than in MS- and NMOSD-related ON. MOGAD-ON patients also often have severe vision loss at nadir³ but tend to have good recovery, leading to better outcomes than NMOSD-related ON. Radiographically, longitudinal and perineural optic nerve enhancement increases the diagnostic suspicion for MOGAD-ON.⁴ MOGAD patients can also present with acute disseminated encephalomyelitis (ADEM) or transverse myelitis with or without concurrent optic neuritis. Brainstem or cerebellar inflammation and cerebral encephalitis are other less common presentations of MOGAD.³

MOGAD-ON is often briskly steroid-responsive but can sometimes relapse with steroid weaning; patients should therefore be given a 1-2 month steroid taper after an attack. Early clinical recognition of MOGAD-ON is crucial as there is some suggestion that early high-dose steroids may be associated with improved visual outcomes.⁵ About half of MOGAD-ON will have a monophasic course. As such, chronic immunotherapy is recommended only for those with relapsing disease or significant disability after the initial attack. Traditional medications for MS are ineffective, and MOGAD patients are most often treated with IV immunoglobulin, tocilizumab, rituximab, azathioprine, or mycophenolate mofetil.

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Older Patient With Vision Loss and Swollen Nerve

Marc H Levin MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Optic neuropathy due to neurosyphilis

Teaching Points

Sexual transmission of the spirochete *Treponema pallidum* causes syphilis, which may present early or late after infection in ocular tissues without systemic manifestations. Syphilitic optic neuropathy (in the secondary or tertiary phase)—and involvement of other ocular tissues (secondary phase)—has become increasingly common in recent years in individuals with and without HIV coinfection.^{1,2} Neurosyphilis should be considered in patients across all demographics who present with atypical inflammatory optic neuropathy and unexplained optic atrophy. Syphilis can cause retrobulbar optic neuritis, papillitis with retinal vasculitis, neuroretinitis, or optic perineuritis. In addition to optic neuropathy, syphilis can affect any part of the eye, including uveitis and chorioretinitis, which can occur in isolation or in conjunction with the optic neuropathy.

Serum RPR and Venereal Disease Research Laboratory (VDRL) testing are useful screening tools, with occasional false positives and negatives in neurosyphilis, and FTA-ABS testing is almost universally positive when syphilis infiltrates the optic nerve. Positive CSF VDRL confirms the diagnosis of tertiary neurosyphilis, although it is frequently negative, and elevated CSF white blood cell and protein concentration are used to follow disease activity before and after treatment. Tertiary optic neuropathy must be treated with 2 weeks of IV penicillin, and all cases of syphilis should be reported to the local public health department. In this case, repeat CSF analysis is planned, and MRI orbits can be repeated to evaluate for resolution of the contralateral right intraconal orbital lesion.

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65-Year-Old With Vision Loss and Swollen Nerve

Anita A Kohli MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Arteritic anterior ischemic optic neuropathy due to giant cell arteritis (GCA)

Teaching Points

The patient's presenting symptoms, demographics, and presence of pallid disc edema with a peripapillary cotton wool spot and elevated inflammatory markers were highly concerning for GCA, which was confirmed with a positive temporal artery biopsy.

The prevalence of GCA is highest in those over 70 years and is very rare in those less than 50. Vision loss, one of the most concerning features of GCA, is usually profound, with less than 40% of patients retaining vision 20/200 or better.¹ Between 8% and 28% of patients have transient vision loss as the initial symptom, and 20%–62% of patients develop vision loss in both eyes.² Patients will often have systemic symptoms, such as weight loss, unexplained fevers, and jaw scalp tenderness and/or temple pain, with jaw claudication reported to be the most suggestive of GCA.³ However, up to 20% of patients with vision loss from GCA do not have systemic symptoms,² so a high index of clinical suspicion must be kept for any elderly patient with vision changes.

GCA may be life threatening due to possible aortic dissection, rupture of aortic aneurysm, or myocardial infarction.⁴ This occurs due to granulomatous inflammation of the medium and larger arteries, with vision loss occurring primarily due to involvement of the ophthalmic artery and posterior cilioretinal arteries, leading to arteritic anterior ischemic optic neuropathy. Two-thirds of the time, there is pallid disc edema in the acute setting,² which can be considered the most pathognomonic examination finding for GCA. Other forms of intraocular ischemia, such as cotton wool spots or arterial occlusions (classically involving the cilioretinal artery or central retinal artery), can greatly increase the suspicion for GCA in the appropriate clinical setting. Rarely, other intracranial arteries are affected and can lead to infarcts of the occipital cortex and resultant homonymous hemianopia, in about 3% of patients.⁵ Both the afferent and efferent visual pathways are susceptible, and in the latter diplopia may result, as in this patient. The diplopia can be transient or constant and can be in any pattern, though these patients often have at least 1 systemic symptom and/or elevated inflammatory markers.⁶

While they are nonspecific markers of inflammation, ESR and CRP are usually elevated in GCA and can help increase the likelihood of diagnosis.⁷ Elevated platelets are also often present. If the diagnosis is unclear and/or homonymous visual field defects are present, neuroimaging, such as MRI/MRA, should be acquired. In the setting of vision loss from presumed GCA,

high-dose IV steroids should be started immediately. Temporal artery biopsy should occur within 1-2 weeks of starting steroids to increase yield.⁸ While the temporal artery biopsy is the gold standard for diagnosis, it is important to note that the inflammation of the temporal artery may be discontinuous, and these skip lesions can occur in up to 28% of specimens.⁹ Therefore a negative biopsy does not rule out GCA, and corticosteroid treatment should be continued if the clinical suspicion remains high and other etiologies have been ruled out.

Temporal artery ultrasound (color Doppler sonography) has emerged as an adjunctive method to diagnose GCA, but it requires skilled sonographers, which may limit its generalizability. Abnormalities on ultrasound can include a hypoechoic halo and occlusion or stenosis of the temporal artery. A meta-analysis found that color duplex ultrasound had a sensitivity of 0.76 and specificity of 0.93 for GCA as compared to clinical diagnosis,¹⁰ suggesting that temporal artery ultrasound may be additive in diagnosing GCA. Other ancillary testing that can support the diagnosis of GCA include delayed choroidal filling on fluorescein angiography,¹¹ enhancement of the temporal arteries on high-resolution MRI,¹² and/or evidence of large vessel vasculitis (eg, aortitis) on CTA.¹³

In 2016 and 2017, 2 randomized, double-blinded clinical trials demonstrated that tocilizumab (Actemra), an IL-6 inhibitor, in conjunction with a prednisone taper was found to be superior to placebo with a prednisone taper for patients with GCA when assessing for steroid-free remission.^{14,15} Consultation with rheumatology regarding the treatment regimen is prudent.

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"I Have Eye Pain, and My Nerve Is Swollen"

Gregory P Van Stavern MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Idiopathic orbital inflammatory syndrome

Teaching Points

Patient's original presentation (optic neuropathy with limitation of eye movements) would localize to the right orbit, particularly the right orbital apex. The subacute, painful onset suggested inflammatory syndrome. Inflammatory orbital disorders encompass all conditions resulting in inflammation to any portion of the orbital compartment. In practice, this is most commonly secondary to bacterial infection and orbital cellulitis. In diabetics and immunocompromised patients, fungal involvement is a major concern, particularly mucormycosis. Thyroid orbitopathy remains the most common cause of noninfectious orbital inflammation in adults.

Noninfectious and nonthyroid orbital inflammation has a large differential diagnosis, including autoimmune diseases (granulomatosis with polyangiitis, sarcoid, IgG4-related disease [IgG4-RD]) and infiltrative neoplasms (lymphoma, metastasis, and others). The most common associations are sarcoidosis, granulomatosis with polyangiitis, and IgG4-RD.

IgG4-RD is a systemic condition characterized by 2 cardinal histopathological features: tissue infiltration by IgG4-bearing plasma cells and fibrosis, which is usually storiform in character. Although raised serum IgG4 titers are common, they are not specific, and the diagnosis rests on pathology from affected tissues. Patients with IgG4-RD can present with isolated orbital inflammation. In one Japanese study, biopsy-confirmed IgG4-RD accounted for 22% of orbital lymphoproliferative cases. Enlargement of the infraorbital nerve can be seen in some cases. Biopsy is essential to confirm the diagnosis.

However, a significant proportion of cases of orbital inflammation have no underlying cause and are labeled as idiopathic orbital inflammatory syndrome (IOIS). This was previously known as “orbital pseudotumor.” The idiopathic form is essentially a diagnosis of exclusion, although a careful history and office-based testing may help narrow the differential diagnosis considerably and allow for a more selective diagnostic workup. Most patients have unilateral onset, although pediatric IOIS has a higher likelihood of bilateral presentation.

Orbital imaging (CT or MRI of the orbits with and without contrast) is indicated for all patients with suspected orbital inflammation. The pattern of involvement in the orbit is variable. In some cases, there is intense enhancement and enlargement of the extraocular muscles, with involvement of the tendon called “orbital myositis” (differentiating it from thyroid orbitopathy, which is tendon sparing). Other findings include diffuse enhancement of the orbital fat (“fat stranding”). If the optic nerve is involved, the most common pattern is perineural enhancement (“optic perineuritis”). Optic perineuritis can also occur in isolation and has a similar differential diagnosis to IOIS. Isolated lacrimal gland involvement has been described as well (dacryoadenitis). Although bone destruction and intracranial extension have been reported, they are rare and should suggest another diagnosis.

Further evaluation depends upon the presentation, as well as patient demographics, but often includes laboratory studies for some of the disorders mentioned above (syphilis serology, ANCA, etc.). Systemic workup for an underlying autoimmune or neoplastic disease is indicated, although the degree and extent are driven by the patient presentation. Imaging of the chest, preferably with CT scan, is often used to screen for sarcoid and lymphoma. PET-CT scan is an option as well since this has higher sensitivity for both sarcoid and lymphoma. Orbital biopsy was traditionally reserved for patients with atypical presentations, those refractory to treatment, and those with a relapsing course, but some specialists favor early biopsy for more definitive diagnosis and treatment.

The mainstay of treatment is high-dose corticosteroids. These should be initiated once an infectious cause has been adequately excluded. For most cases, there is a dramatic and rapid response to steroids, with improvement in orbital findings, motility, and often visual loss. Although there is no high-quality evidence guiding treatment beyond immediate use of steroids, the general recommendation is to taper the prednisone slowly to avoid flareups since these are common. The patient may need to be carefully monitored during the steroid taper, and steroids need to be increased if patients experience a return of their symptoms. The adverse effects of steroids experienced by patients and/or higher rate of relapse in many make steroid-sparing agent therapy a viable option. There is no consensus on which specific steroid-sparing therapy is most effective, and consultation with a rheumatologist may be necessary to choose the best agent. Mycophenolate mofetil, methotrexate, azathioprine, and rituximab have been reported to be efficacious in case reports and case series, but there are no randomized clinical trials to guide therapy.

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“My Nerve Is Swollen, and I Cannot See”

Laura Bonelli MD

DIAGNOSIS & TEACHING POINTS

Diagnosis

Our patient presented with slowly progressive loss of vision, visual field defect, and mild optic nerve edema in the left eye; neuroimaging revealed thickening and circumferential enhancement along the optic nerve consistent with optic nerve sheath meningioma.

Teaching Points

Differential diagnosis in this case includes other causes of visual loss with optic disc edema. Optic neuritis and optic perineuritis are considerations, but the slowly progressive and painless course are not consistent. Similarly, ischemic optic neuropathy would be more rapid in onset. Other orbital compressive or infiltrative lesions typically would produce proptosis or other orbital signs.

Optic nerve sheath meningiomas (ONSM) are rare tumors of the arachnoid cap cells within the optic nerve sheath. They constitute 1%-2% of all meningiomas and are the second-most common intrinsic optic nerve neoplasm after optic nerve gliomas.

ONSM can be classified as primary (pONSM), arising from the intraorbital or intracanalicular portion of the optic nerve, or secondary (sONSM), arising from an intracranial location with extension to the optic nerve and orbit. Secondary ONSM are more common than pONSM. ONSM are more frequent in females (F:M = 3:2), and they generally present between 30 and 50 years of age.

The classic presenting triad of progressive vision loss, optic nerve atrophy, and optociliary shunt vessels (retinochoroidal collaterals) has been described as pathognomonic for ONSM, but few patients present with all 3 findings. The most common presentation is unilateral, progressive decline of vision and visual field with optic nerve pallor or edema.

Sex hormone receptor expression is common in meningiomas—progesterone 88%, estrogen 40%, and androgen 39%. As a result, meningiomas can have increased growth during pregnancy.

Diagnosis is based on clinical presentation and neuroimaging findings. The high-resolution images provided by MRI studies have improved our ability to diagnose ONSM earlier (“Tram-Track sign”). Incisional biopsy is rarely necessary to diagnose ONSM and is generally avoided due to the high risk of severe vision loss.

Management strategies include observation, radiotherapy, and surgical intervention. Minimally symptomatic ONSM can be observed and monitored with clinical examinations, visual field test, OCT studies, and neuroradiologic imaging. As new radiation therapy techniques have evolved, studies have shown very good short- and long-term results of ONSM treatment. The primary current modality is stereotactic fractionated radiotherapy. Surgical treatment of pONSM with total resection is usually associated with severe loss of vision due to damage of the optic nerve vascular supply through the pial plexus and is not done routinely.

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Section III: Neuro-Ophthalmic Mimickers and Visual Disturbances

“I Am Blinded by the Light”

Susan P Mollan MBChB

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Likely sporadic late-onset cone dystrophy

Teaching Points

Late-onset cone dystrophy is a group of rare disorders that affect the cone cells in the retina. While the majority of cone dystrophies are hereditary in origin (autosomal dominant, autosomal recessive and X-linked) and occur earlier in life, around 40% are sporadic and can occur at any time throughout life. It is estimated that 1 in 30,000 people in the general population can be affected. The predominant symptoms are hemeralopia, color vision impairment, and reduced visual acuity.¹⁻³

Hemeralopia is a symptom often misinterpreted in the literature.⁴ The word is derived from ancient Greek; Hemera was the goddess of the day, and *alaos* means “blindness”: “hemeralopia” literally means “day blindness.” While initially the person presented here was thought to have photophobia, on direct questioning her symptom was revealed as a difficulty seeing in the daylight, rather than an aversion to light. Dark glasses helped, which may have enforced the perceived symptom of photophobia.

The differential diagnosis here could have included persistent visual aura, as the condition became evident in temporal proximity to the severe migraine. However, while visual aura occurs in 15% to 20% of migraineurs, it is by definition is a transient phenomenon. Photopsia, scintillating scotoma, and teichopsia are commonly reported by people who suffer from migraine, along with other visual symptoms such as visual distortion, sense of heat wave, blurring, and hemianopsia.⁵ The history of a persistence of the visual phenomenon following a migraine almost certainly led to this patient’s neurology consult. Where persistent visual aura is reported, serious neurological causes such as temporal lobe epilepsy or an occipital space-occupying lesion need to be excluded.⁶ A detailed previous medical and surgical history should be taken in anyone presenting with medically unexplained visual loss, to note if there is a prior history of cancer, intraocular surgery, intracranial surgery, or potentially toxic drugs or medications that could cause the symptoms and focal retinal dysfunction.

As the condition progresses, more objective signs can become obvious. Central visual field loss may be better demonstrated with a Humphrey 10-2 visual field. OCT imaging findings, such as focal disruption of the inner segment and outer segment photoreceptor junction, need to be carefully excluded.⁷ The fundus autofluorescence may be disturbed in the macula, with some people demonstrating a frank bull’s eye maculopathy of the classic ring of hyperfluorescence surrounding a residual

island of hypofluorescence.¹ Likewise in those with more severe forms, there may be pigmentary changes in the macula or temporal disc pallor due to the loss of the retinal nerve fiber layer from the papillomacular bundle.^{2,3}

The diagnosis is confirmed with ocular electrodiagnostic testing. The full-field electroretinography (ERG) can be normal, with the multifocal ERG confirming macular dysfunction. The photopic ERG may be delayed or absent, with a normal scotopic ERG that isolates the cone dysfunction.^{2,3} Often the visual outcome is poor; however, there is variability in those diagnosed with late-onset sporadic progressive cone dystrophy.^{2,3}

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“I Have Normal Vision, but My Optic Nerves Are Pale”

Tatiana Bakaeva MD PhD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Our patient has bilateral superior optic disc hypoplasia related to maternal type 1 diabetes.

Teaching Points

Optic nerve hypoplasia (ONH) is a congenital anomaly of one or both optic nerves characterized by decreased number of optic

nerve axons. It can be mild or severe, segmental or diffuse. Vision can range from 20/20 to light perception. ONH is listed as the third most prevalent cause of vision impairment in children younger than 3 years of age.¹

On fundusoscopic exam, hypoplastic optic disks appear small (one-third to one-half normal size), pale, surrounded by a ring of sclera and annular pigmentation, a “double ring sign.” Fundusoscopic optic disc anomalies in cases of mild or segmental hypoplasia may be subtle.

ONH that only involves the superior segment is called superior segmental optic nerve hypoplasia (SSONH). Patients with SSONH often have normal visual acuity and may have visual field defects that they may be unaware of.

ONH can be isolated or associated with cerebral structural defects. Septo-optic dysplasia (SOD, de Morsier syndrome) is an association between ONH and absence of septum pellucidum, agenesis of the corpus callosum and pituitary gland abnormalities. Patients with ONH often have endocrine abnormalities and developmental delays.^{2,3}

Numerous gestational risk factors for ONH have been reported, including maternal smoking, first trimester bleeding, young maternal age, primiparity, low maternal weight gain or weight loss, preterm labor, low birth weight, and intrauterine growth restriction. An association of maternal diabetes with SSONH is well described.⁴

The exact pathophysiologic mechanism of ONH is unclear. Fetal vascular insult and primary failure of development of retinal ganglion cells have been hypothesized. Several gene mutations have been shown to affect optic nerve and hypothalamic-pituitary axis development, though no specific genotype-phenotype correlation has been found to explain most of the cases of ONH.

All children with suspected hypothalamic dysfunction should have an ophthalmoscopic evaluation to look for ONH. Brain MRI and endocrinology evaluation are recommended for all cases of confirmed ONH. Children with ONH should be monitored for growth and development and often need occupational, physical, and speech therapy. Their vision should be monitored, and treatments for amblyopia, strabismus, and low vision interventions should be offered when needed.

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“My Vision Is Normal, but I Cannot See”

Rudrani Banik MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

The patient was diagnosed with visual snow syndrome in the setting of post-concussion syndrome and migraine.

Teaching Points

Visual snow syndrome (VSS) is a neurological condition characterized by the presence of persistent visual disturbances, described as television static-like or flickering patterns throughout the entire visual field in both eyes. Individuals with VSS typically report a variety of visual and nonvisual symptoms. Visual symptoms include visual snow, photophobia, floaters, nyctalopia, blue field entoptic phenomena, and palinopsia (persistent afterimages and trailing images). Nonvisual symptoms may include tinnitus, paresthesias, depersonalization, and derealization. Many patients also have psychiatric issues such as anxiety and depression associated with their VSS symptoms.

The diagnosis of VSS is based on specific diagnostic criteria in which there is the presence of typical visual symptoms and the absence of any underlying structural abnormalities or eye conditions. Ancillary testing such as neuroimaging and electrophysiologic testing (electroretinography [ERG], visual evoked potential, and multifocal ERG) are typically not needed for diagnosis in the context of typical symptoms.

The prevalence of VSS is estimated to be 1%-2% in the general population. VSS may be seen in migraine or postconcussion syndrome, though in many patients it develops spontaneously. The exact cause of VSS is unknown, but it is believed to be related to abnormal functioning of the visual processing pathways in the brain. Two theories of VSS include thalamocortical dysrhythmia and neuronal hyperexcitability within the visual cortex.

Currently, there is no evidence-based cure or treatment for VSS. However, several management strategies can help alleviate symptoms and improve quality of life. These include lifestyle modifications (eg, reducing stress, avoiding triggers), prescription medications (such as lamotrigine, topiramate, or clonazepam), and nonpharmacological approaches (eg, cognitive behavioral therapy, biofeedback).

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“I Have a Visual Field Defect, but My Nerve Looks Normal”

M Tariq Bhatti MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Vitreomacular traction¹

Teaching Points

The type of visual field defect can often determine the anatomic location of the lesion. In general, the following characteristics of a visual field defect should be noted:

- Monocular vs. binocular
- Pattern of defect (eg, arcuate, paracentral, central, cecentral, altitudinal)
- Homonymous vs. heteronymous
- Complete vs. incomplete
- Incongruous vs. congruous (Historically, a congruent homonymous hemianopia was considered to originate from a lesion involving the occipital lobe, but this paradigm has been shown not to be a reliable localizing sign.)²

The differential diagnosis of a monocular visual field defect is broad, but there are several general concepts that should

be kept in mind. Aside from a few instances (anterior chiasm or anterior occipital lobe), a monocular visual field defect is indicative of a lesion of the retina or optic nerve. Anterior segment pathology such as a cataract rarely can cause a visual field defect.³

Unless the process is acute (less than 4-6 weeks), a lesion of the optic nerve will often be associated with either optic disc pallor on clinical examination or changes on the OCT (ie, peripapillary retinal nerve fiber layer [pRNFL] and/or ganglion cell–inner plexiform layer thinning). Typically, retinal disease does not result in optic disc pallor, but in some cases, such as retinitis pigmentosa, there can be optic disc pallor. Several important clinical and testing findings can help differentiate an optic neuropathy from a retinopathy/maculopathy; see Table 1.⁴

OCT has become an invaluable component in the evaluation of a patient with visual loss. When interpreting an OCT, it is important not only to view the pRNFL thickness plot but also to carefully review all the macular B-scans (source images) and remain cognizant of any imaging artifacts.⁵

In terms of our patient’s final diagnosis, vitreomacular traction is characterized by focal adhesion and drag by the vitreous, resulting in anatomical abnormalities of the underlying retina. Secondary retinal pathologies include loss of the foveal contour, retinal folds, cystoid macular edema, retinoschisis, tractional retinal detachment, retinal tears, macular hole formation, and avulsion of retinal vessels with vitreous hemorrhage.⁶

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Table 1

Symptom	Optic Neuropathy	Retinopathy/Maculopathy
Metamorphopsia	Rare	Common
Dyschromatopsia	Common	Rare
Pain	Common in optic neuritis	None
Photophobia/glare	Rare	Sometimes
Photopsia	Rare	Common
Paraclinical testing		
Amsler grid	Absent line (scotoma)	Distorted lines
Ophthalmoscope	Normal, pale, or swollen optic disc	Normal (occasionally pale optic disc)
Photostress test	Normal	Delayed
Reduced visual acuity	Common	Common
Relative afferent pupillary defect	Present if unilateral or asymmetric bilateral	None, unless extensive retinal damage
Visual field defect	Central scotoma, arcuate	Central scotoma

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“I Have Light Sensitivity”

Samuel Spiegel MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

This patient was diagnosed with photophobia after concussion.

Teaching Points

Light sensitivity, or photophobia, is a common presenting symptom in many conditions, including ocular surface disease, intraocular inflammation, retinal degenerations, migraine, and other neurologic pathology. Therefore, careful ophthalmologic examination with review of patient history is important to rule out other causative etiologies. In absence of ocular pathology, photophobia, in conjunction with a history of head trauma, even mild trauma, should raise the possibility of concussion and related photophobia.¹ Mild head injuries can have associated symptoms that linger for weeks, and nearly half of all individuals diagnosed with postconcussive syndrome following mild traumatic brain injury report symptoms lasting up to 3 months. Those who suffer postconcussive syndrome commonly retain an increased sensitivity to light.²⁻⁴

For photophobia secondary to concussion, clinicians should focus on treating reversible causes, appropriate expectation setting, and reassurance. Concrete ophthalmologic management strategies should include the use of tinted lenses (FL-41), minimal use of sunglasses indoors, discouraging other maladaptive behavior, and frequent use of topical ocular lubrication.⁴⁻⁷ It is important to consider other associated features and address those that are applicable, such as co-occurrence of migraine. Given the commonalities and overlap in associated conditions and therapy, a multimodal approach can help mitigate symptoms. Multidisciplinary involvement of other specialties, such as neurology, occupational therapy, or psychotherapy, may be warranted.^{2,5} Recognizing the potential impact on an individual's daily life is important as well. Overall, a combination of targeted ophthalmologic related interventions tailored to an individual's needs can effectively alleviate symptoms and improve quality of life.

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Section IV: Double Vision—What to Do?

Case of Acute Double Vision in a Younger Patient

Lauren C Ditta MD

DIAGNOSIS & TEACHING POINTS

Trochlear nerve palsy (cranial nerve IV palsy) represents the most common paralysis of a single cyclovertical muscle (superior oblique).¹ Most adult cases are congenital in etiology; however, many do not present with symptoms until adulthood, when patients can no longer maintain fusional vergence control of their hyperphoria and develop diplopia. Trauma is a common cause of acquired CN IV palsy due to vulnerability of the nerve in its extended course from the dorsal midbrain to the superior orbital fissure. In the absence of a supportive history, it can be challenging to decipher patients with other causes for acute-onset vertical diplopia and/or new nonspecific visual symptoms, who may require extensive diagnostic workup.

In the acute setting of an acquired, unilateral CN IV palsy, most patients experience intractable, symptomatic vertical diplopia, along with diagonal or torsional symptoms.¹ Patients may manifest a compensatory head tilt and turn away from the affected side. Or, in bilateral CN IV palsies, a patient may have a chin-down posture to compensate for an induced V pattern strabismus caused by reduced tertiary abducting function of both superior oblique muscles. Observation for subtle facial asymmetry, specifically relative midface hypoplasia, may be a nonspecific sign of a congenital CN IV palsy on the affected side, caused by a compensatory torticollis to maintain fusion dating back to infancy or early childhood.²

Diagnostic criteria for an isolated, unilateral CN IV palsy include a hypertropia of the affected eye that increases on adduction and ipsilateral head tilt, which can be clinically identified by the Parks-Bielschowsky 3-step test. A helpful fourth step to differentiate a CN IV palsy from skew deviation is the upright supine test.³ The 3-step test is reliable to identify an isolated cyclovertical muscle palsy in the acute setting; however, it does not differentiate congenitally decompensated superior oblique palsies from acquired cases, and it can be confounded by restrictive strabismus, multiple cranial nerve palsies, and prior trauma or strabismus surgery. Like other cranial nerve palsies, myasthenia gravis, and thyroid disease, CN IV palsies create incomitant strabismus, and careful assessment of ductions and versions should reveal expected incomitance patterns. Understanding extraocular motility patterns in all positions of gaze allows for a better understanding of the patient's symptoms and can be particularly helpful with management, including surgical planning (eg, diplopia worse in contralateral down gaze—the field of action of the superior oblique muscle—versus diplopia in ipsilateral or contralateral up gaze, due to a tight superior rectus or compensatory inferior oblique overaction, respectively.) Additionally, patients with a longstanding CN

IV palsy may have large vertical fusional amplitudes (ability to fuse more than 2-4^Δ in the vertical plane), whereas patients with newly acquired palsies struggle to fuse small amounts of vertical prism. Finally, torsion should be subjectively assessed by the double Maddox rod test as patients with an acquired CN IV palsy often have a measurable degree of torsion, compared to congenital cases. Torsional diplopia measuring >10 degrees may be indicative of a bilateral CN IV palsy. Fundus excyclotorsion can also be noted on ophthalmoscopy.

There is good evidence to support early neuroimaging in all patients who present with acute, isolated ocular motor palsies.⁴ This is especially important in younger adults (<50) and pediatric patients who have a high prevalence of structural, infectious, and inflammatory causes where a comprehensive evaluation, including MRI of the brain and laboratory workup, is recommended.⁵

In the case presented, neuroimaging was important to identify a schwannoma, which is a benign skull base lesion that can often be missed on initial neuroimaging studies when contrast is not used due to the small size of the fourth cranial nerve. Schwannomas represent 8% of primary intracranial tumors and are uncommonly seen in association with the ocular motor cranial nerves. In general, isolated CN IV palsies resulting from an intracranial tumor are rare.⁴

Management of CN IV palsy is primarily dictated by the underlying pathology and the patient's specific symptoms in the setting of the clinical examination. For schwannomas of the trochlear nerve, the primary goals of treatment are to relieve a patient's symptoms and to prevent tumor progression. Partial occlusion can serve as a conservative, short-term option for immediate relief of diplopia. If the vertical deviation is small and relatively comitant, base-down Fresnel prisms can be placed on spectacles in the acute setting or ground into glasses for stable deviations. For patients who are surgical candidates, proper surgical planning includes a thorough understanding of the patient's symptoms, measuring all 9 diagnostic fields of gaze to identify specific patterns of incomitance, and performing an intraoperative examination with oblique traction testing.

It is reasonable to delay surgery (6-12 months) to monitor for spontaneous resolution and stable measurements.¹ Deviations $\geq 15^{\Delta}$ generally require 2 muscle surgeries to achieve orthotropia. While there is no standard treatment recommendation for trochlear nerve schwannomas, it is reasonable to monitor small, isolated, asymptomatic lesions with clinical and radiological observation with MRI with/without contrast, as these lesions often remain stable over time.⁶ Stereotactic radiosurgery may be indicated for larger, progressive, and symptomatic lesions because of its precise and targeted delivery of radiation and low morbidity.⁷ When counseling patients on management, it is important to explain that symptoms following surgical treatment are expected to improve, but often persist.

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Case of Acute Double Vision in an Older Patient

Crandall E Peeler MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Right sixth nerve palsy secondary to IgG4-related disease

Teaching Points

Isolated cranial nerve VI dysfunction is the most commonly encountered ocular motor palsy, and in adults over the age of 50, it most often results from microvascular injury in the setting of classic cardiovascular risk factors (ie, systemic hypertension, hyperlipidemia, diabetes, and smoking).^{1,2} Microvascular sixth nerve palsies tend to resolve spontaneously, and several practice guidelines recommend clinical monitoring in older adults without initial MRI as a cost-savings measure, with neuroimaging reserved for those who progress or fail to improve over 4 to 12 weeks.^{3,4} However, studies of early neuroimaging in cases of presumed microvascular sixth nerve palsy in adults over 50 estimate that a causative structural lesion may be present in up to 15% of cases. Many providers also feel that early, normal neuroimaging has significant value in terms of allaying patient fear and anxiety in the setting of new-onset diplopia.¹ As a result, the question of whether to image early in these cases remains far from settled.

When considering the appropriate workup in a case of presumed microvascular sixth nerve palsy, careful history taking

is crucial to confirm whether the new deficit is truly “isolated.” Patients with microvascular cranial nerve injuries commonly report mild to moderate headache and periorbital pain, so experts will continue to label patients with these associated symptoms as “isolated” cases.¹ However, any signs of orbital disease (ie, proptosis or periorbital swelling), other neurologic symptoms (ie, additional cranial nerve palsies, numbness, weakness, or ataxia), severe headache, a history of prior malignancy, or symptoms of systemic infection, inflammation, or giant cell arteritis should trigger an immediate workup.³ In the case presented here, the patient’s symptoms of ipsilateral mastoid tenderness as well as tinnitus were concerning for focal infection, inflammation, or neoplasm with possible spread to the central nervous system. However, he did not readily offer up these associated symptoms, as he was most bothered by the new-onset diplopia.

The differential diagnosis for the patient presented here was initially broad following his MRI. Fortunately, the bulk of the enhancing tissue seen on imaging was readily accessible for biopsy, culture, and histopathologic analysis. Immunoglobulin G4-related disease (IgG4-RD) is a chronic fibroinflammatory condition associated with elevated serum levels of IgG4 (≥ 135 mg/dL), storiform fibrosis, and IgG4-positive plasmacyte infiltration of affected tissues (≥ 10 /high powered field with a ratio of IgG4+ cells $>40\%$). IgG4-RD may mimic lymphoma or granuloma formation seen in sarcoidosis or tuberculosis infection, so evaluation typically involves ruling out these other conditions while ruling in IgG4-RD. Elevated serum IgG4 levels may also be seen in eosinophilic granulomatosis with polyangiitis and multicentric Castleman disease. Sites most commonly involved include the orbit and lacrimal gland, salivary glands, pancreas and bile duct, tissues of the head and neck, lung, and retroperitoneal cavity.^{5,6}

Systemic corticosteroids are an effective initial treatment of IgG4-RD, with recommended dosing between 0.6 and 1 mg/kg/day, depending on the extent of disease. Symptoms tend to improve rapidly with steroid treatment, but an extended taper is recommended given a high rate of relapse. Patients are typically managed by rheumatology and transitioned to a steroid-sparing agent, such as the anti-CD20 antibody rituximab, for long-term maintenance. Routine cancer surveillance is recommended, as a higher rate of malignancy has been identified in patients diagnosed with IgG4-RD.⁵

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Cases of Esotropia With Sixth Nerve Palsies, Sagging Eye, Heavy Eye: When to Image

Zoë R Williams MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Age-related distance esotropia secondary to sagging eye syndrome

Teaching Points

Sagging eye syndrome (SES) has been described as a cause of horizontal and/or vertical ocular misalignment in older patients, resulting from connective tissue band stretch between the superior and lateral rectus muscles with inferior displacement of the lateral rectus muscle pulley.¹ Patel et al reported the lateral rectus–superior rectus band was visible in 95% of coronal T1 weighted images, almost 70% of coronal short tau inversion recovery (STIR) images, and 70% of coronal CT images.² Chaudhuri and Demer found that the lateral rectus–superior rectus band was frequently ruptured in SES.¹ There are typically accompanying signs of adnexal connective tissue degeneration causing levator aponeurosis dehiscence and deep superior sulci.^{1,3} Abduction should be full with normal horizontal saccadic velocities, although there may be limitation of supraduction, and the esotropia for distance should be vertically comitant.¹ If there is symmetric inferior displacement of the lateral rectus muscle pulleys, SES causes a purely horizontal misalignment (esotropia greater at distance than near); however, if there is asymmetric displacement, a cyclovertical strabismus is induced (hypotropia on the side of the greater lateral rectus muscle pulley displacement).^{1,3} Age-related distance esotropia (ARDE) is defined as distance esotropia at least double the asymptomatic esotropia of ≤ 10 PD at near.⁴ The esotropia for distance can be mild to moderate (variably reported across studies as a mean distance esotropia of 4–20 PD) and is usually more than double the deviation at near.^{3,5–7}

A study of 945 patients over age 40 (mean age: 66) presenting to the Stein Eye Institute with binocular diplopia found that 31% had SES.³ SES was diagnosed if there was either ARDE, defined as symptomatic esotropia at distance with orthophoria or esophoria of ≤ 10 PD at near, or small-angle cyclovertical strabismus (CVS), defined as symptomatic vertical strabismus (hypotropic eye was more excyclorotated inconsistent with superior oblique palsy and there was minimal change with right or left head tilt and no evidence of superior oblique muscle atrophy) with or without horizontal strabismus.³ In addition, adnexal degenerative changes and normal saccades were required.³ The majority of patients were female and over age 70.³ The relative proportion of patients with diplopia from SES increased from $<5\%$ under age 50 years to $>60\%$ over age 90.³ ARDE was present in 35% and CVS in 65% of cases of SES.³

ARDE can be treated with prismatic correction or bilateral medial rectus recession vs. bilateral lateral rectus resection.^{4–6} Chaudhuri and Demer recommend the target angle for correc-

tion by medial rectus muscle recession is double the distance esotropia for ARDE.⁴ An increase in surgical dose is not recommended for lateral rectus plication/resection in ARDE.⁴ Hypertropia in CVS can be treated with either graded vertical rectus partial tenotomy or vertical rectus muscle recession or resection.³ Ocular misalignment may recur after surgery with progressive SES.^{3,7}

Neuroimaging is not required to diagnose SES but may be helpful in surgical planning in cases presenting with ARDE. If neuroimaging is deferred, it is important to ensure a complete neuro-ophthalmologic examination is performed to assess for alternate etiologies mimicking SES, such as posterior fossa lesions, hydrocephalus, and Chiari malformations.^{3,8,9} Cerebellar esotropia and skew deviation may present with similar ocular misalignment as SES and can be differentiated from SES by accompanying cerebellar signs of ataxia, gaze-evoked nystagmus, or saccadic dysmetria. Cyclovertical strabismus due to SES can mimic a CN IV palsy, and neuroimaging can be helpful to confirm absence of superior oblique muscle atrophy in SES.⁸ SES is rare in young patients; therefore neuroimaging should be performed to exclude a neurologic etiology causing divergence insufficiency type esotropia.^{3,8,9} If there is lateral incomitance of the esotropia, CN VI paresis should be suspected with assessment of saccadic velocity and full cranial nerve examination. Mild thyroid eye disease can present with a fairly comitant esotropia for distance with minimal or no apparent limitation of ductions.⁶ Exophthalmos, eyelid retraction, lid lag, or lateral flare can be helpful in differentiating thyroid eye disease from SES.

Heavy eye syndrome (HES) is a rare type of mechanical strabismus causing esotropia and hypotropia in the setting of high axial myopia with superotemporal globe prolapse outside the muscle cone.^{8,10} HES also causes degeneration of the lateral rectus–superior rectus band; however, in contrast to SES, there is more often a large angle esotropia and hypotropia with impaired abduction and supraduction due to slippage of both the lateral rectus muscle pulleys inferiorly and the superior rectus muscle pulleys medially.^{8,10} It can sometimes be difficult to distinguish HES from SES reliably without orbital imaging, which is required for a diagnosis of HES.^{8,11} Quasicoronal orbital imaging studies have shown that the angle between the superior and lateral rectus muscle centers relative to the globe center is significantly greater in HES (mean SR-LR displacement angle of 121 degrees) than in SES (mean SR-LR displacement angle of 104 degrees).¹¹ It is essential to distinguish HES from SES for surgical planning, as HES can be successfully treated by loop myopexy of the superior and lateral rectus muscles to restore the normal anatomic configuration, as described by Yokoyama,^{8,11–13} with additional medial rectus muscle recession if medial rectus muscle contracture is suspected, as described by Yamaguchi.¹³

In summary, patients with ARDE due to possible SES who are younger than the typical demographic or who have atypical features (rapid progression, significant incomitance of esotropia, or limitation of abduction) should undergo neuroimaging to exclude alternate etiologies, including neurologic mimickers. It is important to perform a complete neuro-ophthalmologic examination to assess for signs incompatible with SES. Patients with high myopia with suspected SES require orbital imaging to rule out radiologic evidence of HES, as this will change surgical management.^{8,11–13} All patients with a history of cancer, regardless of age, should undergo neuroimaging with contrast-

enhanced MRI brain and orbits to rule out metastasis in the setting of acquired binocular diplopia. It is controversial whether neuroimaging is indicated in all patients with binocular diplopia in the pattern of an isolated cranial nerve palsy.^{14,15}

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Surgical Considerations in Diplopia

Ore-Ofeoluwatomi O Adesina MD

DIAGNOSIS & TEACHING POINTS

Final Diagnosis

Chronic palsy of the subarachnoid and cavernous left CN VI in the setting of a left skull-base meningioma

Teaching Points

Our patient had a chronic left CN VI palsy with persistent, symptomatic diplopia. The clinical course of this patient highlights some important teaching points in the management of chronic CN VI palsies.

Timing of Intervention

Sixth nerve palsies from injury to the nerve in the subarachnoid space and cavernous sinuses can occur from a multitude of etiologies, including compression (from tumors or intracranial pressure elevation), inflammation (multiple sclerosis, sarcoidosis), infection (syphilis, petrous apex infections, cavernous sinus thrombosis), and trauma. Esotropia from chronic CN VI palsy usually requires surgery; however, because spontaneous improvement can occur after CN VI injury, intervention in the acute phase is typically limited to patching for larger deviations and temporary prisms for smaller deviations. It is prudent to wait 6-12 months until no further spontaneous improvement is seen for larger deviations prior to considering surgical intervention. In the case of this patient with a CN VI palsy from nerve compression due to a meningioma, it was prudent to wait to perform surgery until his deviation stabilized after receiving radiosurgery and administering botulinum toxin.

The Role of Chemodenervation

Contracture of the antagonist medial rectus (MR) muscle with resultant restriction of abduction can occur in CN VI palsies, and chemodenervation of the MR in both acute and chronic CN VI palsy has been employed for several decades. Although surgery tends to be more beneficial than botulinum toxin as monotherapy, the latter has been shown to significantly improve alignment for many patients¹ and can help differentiate a complete lateral rectus (LR) palsy from a partial one with MR restriction. In a study by Repka and Morrison, a course of chemodenervation significantly improved the alignment of 41% of patients who ultimately did not require surgical intervention.² In the other cases, only a modest improvement was seen;² however, the implication is that even modest improvement can reduce the surgical dosing needed to correct alignment, as was seen in this case.

Role of Prisms

Prisms can be used to alleviate symptomatic diplopia and can be incorporated into glasses or given in the form of Fresnel prism. Fresnel prisms are flexible prism membranes made from optical-grade polyvinyl chloride. One side of the prism has angular

grooves that add up to the prismatic correction of the prism, and the other smooth side is attached to the glasses after being cut to shape to fit the lens. They can be particularly helpful for patients in the first 6 months of their acute neurologic injury as a means of temporary relief of their diplopia as their recovery is monitored and for those who do not wish to receive chemodenervation of the MR. In patients who wear glasses, they can be attached to the current spectacle correction. Alternatively, they can be placed on a pair of clear lenses for those who do not wear glasses.

Fresnels come in a range of 17 powers from 1.00 to 40.00 PD. While the amount of strabismus that can be corrected by Fresnel prisms is quite large, the prisms create optical blur in direct proportion to the strength of the prism, and for larger angles of strabismus they may reduce visual acuity by several lines. Most patients do not tolerate more than 15-20 PD of correction with Fresnels, and in the case of this patient, his 30 PD LET was too large to be comfortably corrected this way. He instead opted for patching the left eye until undergoing surgery. Finally, prisms can be incorporated into spectacle correction if the angle of strabismus is amenable to prismatic correction once stable or prescribed postoperatively, which was the case in this patient's treatment.

Choice of Surgical Procedure

In cases of chronic CN VI palsies requiring surgery, the choice of surgical procedure depends upon the severity of abducens weakness after recovery of CN VI function with a stable deviation, as well as the results of forced ductions employed to evaluate for MR contracture. In situations where there is partial recovery of CN VI function, surgery usually consists of weakening the ipsilateral MR muscle and tightening the ipsilateral LR muscle using standard dosing tables. For patients with chronic complete or severe CN VI palsies with little or no ability to abduct the eye from primary position, the goal of surgical intervention is to transpose the vertical rectus muscles toward the LR, utilizing their tone to improve alignment in primary position. There should be little expectation of improved abduction of the eye with these procedures. Over the years, multiple procedures transposing the superior and inferior rectus muscles with full or partial tendon or muscle transpositions with or without tenotomy and with or without weakening of the MR muscle (based on forced duction testing for restriction) have been developed. A full review of these procedures is outside the scope of this text, but the reader is referred to an excellent 2021 review by Akbari et al⁵ for this. Anterior segment ischemia (ASI) is a rare but serious complication of transposition procedures, and its risk increases with tenotomy of multiple muscles and in patients with vascular risk factors such as diabetes.

Partial muscle transpositions, single muscle transpositions, and transpositions without tenotomy have been introduced to reduce the risk of multiple muscle manipulation and ischemia. One such procedure is the superior rectus (SR) transposition to the LR muscle with MR recession to avoid anterior segment ischemia. This was described by Hunter et al for management of Duane syndrome and CN VI palsy.³ In 2003, Nishida et al reported a new muscle transposition procedure in which the split temporal halves of the SR and IR were transposed without tenotomy or LR manipulation. In order to further reduce

operative damage, the procedure was further modified without tenotomy or muscle splitting. In this procedure, after the vertical recti were explored, 6-0 polypropylene sutures were passed through the temporal margins of each muscle at a distance of 8 to 10 mm behind their insertion points. The same sutures were then passed through each scleral wall at a distance of 10 to 12 mm behind the supero- and inferotemporal limbus. Then, the temporal margin of each vertical rectus was transposed supero- or inferotemporally, toward the LR, and anchored onto the sclera. In their 2005 review, Muraki et al performed the procedure on 9 patients with complete CN VI palsies, with 6 of 9 undergoing MR muscle recessions. The surgical correction by muscle transposition alone ranged from 24 to 36 PD, and that by muscle transposition and recession of the MR ranged from 50 to 62 PD. The mean correction was 46.3 ± 13.1 PD per eye, and no major vertical ductional disturbances or anterior segment ischemia occurred in any patients.⁴

Outcomes

The 2021 review by Akbari et al⁵ concluded that transposition procedures are highly effective in the treatment of esotropia caused by complete LR palsy, with 50% to 80% success rates.⁵ Success can be defined in different ways; in a report by Peragallo et al success was defined as the absence of diplopia without prisms or face turn, vertical deviation ≤ 2 PD and horizontal deviation ≤ 10 PD. Success rates for strabismus procedures in patients with abducens palsies were similar across all etiologies, and the frequency of reoperation was higher among those patients with neoplastic or traumatic etiologies than those with microvascular/idiopathic or other central nervous system causes.⁶ In this case, the modified Nishida procedure was performed to minimize manipulation of the vertical rectus muscles. A 6-mm MR recession was performed due to positive intraoperative force ductions and resulted in good postoperative alignment, eventually requiring a small prism correction to produce a comfortable, diplopia-free outcome.

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Index

Adesina, Ore-Ofeoluwatomi O 15, 35
Bakaeva, Tatiana 10, 28
Banik, Rudrani 10, 29
Bhatti, M Tariq 11, 30
Bonelli, Laura 9, 27
Cherayil, Neena 7, 23
Ditta, Lauren C 13, 32
Gold, Dan R 2, 18
Henderson, Amanda D 4, 22
Kohli, Anita A 8, 24
Lee, Michael S 2, 18
Levin, Marc H 7, 24
MacIntosh, Peter W 3, 20
Mollan, Susan P 10, 28
Peeler, Crandall E 13, 33
Rasool, Nailyn 3, 19
Spiegel, Samuel J 12, 31
Subramanian, Prem S 5
Van Stavern, Gregory P 8, 25
Williams, Zoë 14, 34