

The Case of a High School Graduate With a New Blind Spot

Julia Young* was preparing to celebrate her high school graduation by visiting New York City. While packing, the 18-year-old noticed a dark spot just to the right of her central vision in the right eye. Her parents brought her to our institute's comprehensive ophthalmology clinic for urgent evaluation.

Presentation

When Ms. Young presented to our comprehensive ophthalmology colleagues, she described an egg-shaped black dot just off-center in her right field of view. It had been present for about 12 hours and was fixed. She denied flashes of light, floaters, or history of refractive error. She had no relevant medical history. She was using oral contraceptives and topical retinoid lotion for acne but was not taking any systemic retinoid medications. Her weight was normal, and she said that it had been stable. She also denied any headaches or recent illness.

The exam. At this initial examination, Ms. Young had an uncorrected visual acuity of 20/20–1 in the right eye and 20/20 in the left eye. Her intraocular pressures were 22 mm Hg in the right eye and 23 mm Hg in the left. Her pupils were equal, round, and briskly reactive with no afferent pupillary defect (APD). Slit-lamp biomicroscopy showed that the anterior segment was unremarkable. Posterior segment examina-

tion revealed an elevated right optic disc with mild obscuration of the margins inferiorly; the left optic disc demonstrated 360 degrees of elevation with obscuration of the margin. There were no spontaneous venous pulsations in either eye.

Fundus autofluorescence (FAF) showed hyperfluorescence of the inferior right disc (Fig. 1) and 360 degrees of hyperfluorescence of the left optic disc.

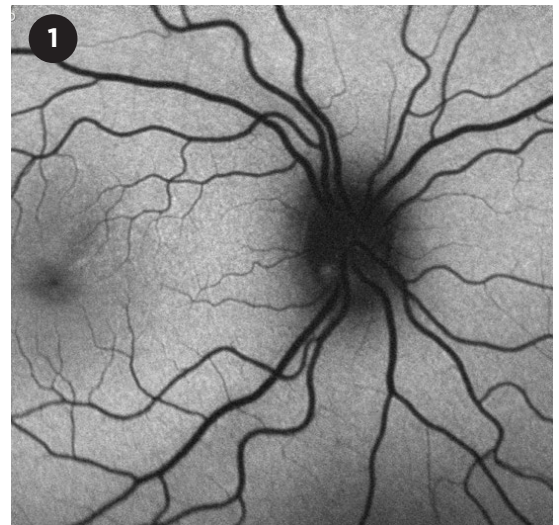
Ms. Young was diagnosed with buried optic disc drusen and sent to neuro-ophthalmology for further evaluation.

Referral to Neuro

The next morning, we examined Ms. Young in our neuro-ophthalmology clinic. She continued to report that she was experiencing an oval-shaped blind spot just off-center in her right eye and said that the scotoma had been present for more than 24 hours and had not changed in size or appearance. She was unable to see through the spot. Her examination was unchanged. Formal 24–2 Humphrey visual fields were obtained and noted to be reliable and full.

Further Testing and Review

We sent Ms. Young for retinal nerve

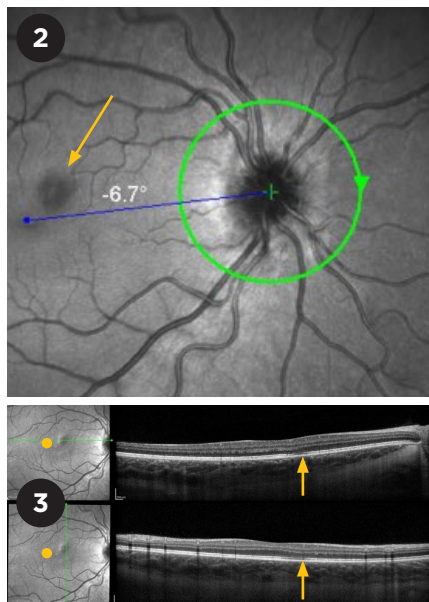


INITIAL IMAGING. Fundus autofluorescence (FAF) demonstrating hyperfluorescence inferiorly, consistent with disc drusen of the right optic nerve.

fiber layer optical coherence tomography (RNFL OCT). These images were consistent with drusen but did not explain her vision loss. At this point, we discussed the patient's symptoms in more detail. She very clearly stated that when she looked straight ahead and fixated with her right eye on a Snellen visual acuity line, the third letter was missing. We showed her several lines of visual acuity at the same exam, and she repeatedly described a blind spot just temporal to center.

We then reviewed the RNFL OCT, which showed a wedge-shaped abnormality in the near infrared imaging in the supratemporal macula that corresponded to the blind spot that Ms. Young had described (Fig. 2). Review of her FAF from the previous day also

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FURTHER IMAGING. (2) The infrared imaging obtained during RNFL OCT demonstrated a wedge-shaped hypofluorescent area in the supratemporal macula (arrow) that corresponded to the patient's described visual disturbance. (3) SD-OCT showed disruption of the ellipsoid line (arrow) corresponding to the wedge-shaped hypofluorescent area seen in the infrared (dot), which is consistent with AMN.

revealed a retinal abnormality consistent with these findings.

The Final Diagnosis

We suspected that her diagnosis was acute macular neuroretinopathy (AMN), and we sent her to the retina clinic. There she underwent additional imaging and evaluation. Spectral-domain optical coherence tomography (SD-OCT) of her macula was significant for hyperreflectivity of her outer retina (see arrow in Fig. 3). This anomaly corresponded to the wedge-shaped alteration that we had noticed in the near infrared and FAF imaging. This finding confirmed the diagnosis.

Discussion

AMN, first described by Bos and Deutman, is an uncommon retinal disorder that affects the outer retinal layers.¹

Most patients are young women who present with sudden-onset unilateral or bilateral visual change with a paracentral or central scotoma.^{2,3} The disorder has typically been associated with viral illness and oral contraceptive use; more rarely, trauma and vaccination administration have been implicated.^{3,4} Ischemic injury to the outer retinal capillary network has been proposed as an underlying mechanism.⁵

Classically, examination reveals a wedge-shaped lesion pointing to the macula.¹ And near infrared imaging demonstrates hyporeflexive changes.⁵ In

fact, the near infrared imaging associated with Ms. Young's OCT RNFL for optic nerve head drusen prompted the diagnosis. SD-OCT of the retina or macula is quite helpful in making the diagnosis, especially in the acute phase when there are hyperreflective plaques at the level of the outer plexiform layer or outer nuclear layer.⁶

This case highlights the utility of multimodal imaging in making the diagnosis of AMN: FAF, near infrared imaging, and macular OCT all were helpful ancillary tests.

Our patient—a young woman with a history of oral contraceptive use—fits the classic description of patients at risk for AMN; however, the finding of optic nerve head drusen temporarily steered the referring physician away from the correct diagnosis.

Ms. Young's Current Status

We have seen Ms. Young several times since her initial visit. She still uses contraceptives. There has been little change in her symptoms, and her visual acuity remains the same. We obtained OCT angiography to examine for choriocapillary dropout, but the results of that were unrevealing.

Her OCT demonstrates slight improvement in the disruption of the ellipsoid line in the right eye, and the left eye imaging continues to be normal.

* Patient name is fictitious.

1 Bos PJ, Deutman AF. *Am J Ophthalmol.* 1975; 80(4):573-584.
 2 Turbeville SD et al. *Surv Ophthalmol.* 2003;48 (1):1-11.
 3 Bhavsar KV et al. *Surv Ophthalmol.* 2016;61(5): 538-565.
 4 Shah P et al. *Ophthalmic Surg Lasers Imaging Retina.* 2018;49(10):165-168.
 5 Fawzi AA et al. *Retina.* 2012;32(8):1500-1513.
 6 Munk MR et al. *Br J Ophthalmol.* 2017;101(2): 160-165.

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Six Medical Mysteries

Which Morning Rounds case reports have clocked up the most views? Of the cases published in the last two years, the six below have racked up more than 20,000 views. For links to all six, see this month's Morning Rounds online at aao.org/eyenet.

- **Aches and Pains and Blurry Vision.** A 28-year-old woman had flulike symptoms followed by red, painful, photophobic eyes and decreased vision.
- **Bitemporal Visual Field Defects.** The 47-year-old had dry eye disease secondary to Sjögren syndrome. She had recently started hydroxychloroquine therapy.
- **"Doctor, There's a Screaming Sound in My Ears."** The 11-year-old girl was plagued by increasing headaches and a "screaming" that she sometimes heard for hours.
- **Red Eyes and Red Ears.** The 57-year-old's eyes were red, painful, and photophobic; his ears, painful and tender.
- **Pixelated Vision.** The 65-year-old insisted that he had a 10%-15% reduction in vision, which he described as pixelated.
- **An Unusual Case of Left-Sided Vision Loss.** At first, she had visual field defects; two weeks later, she had altered mental status and was blind.