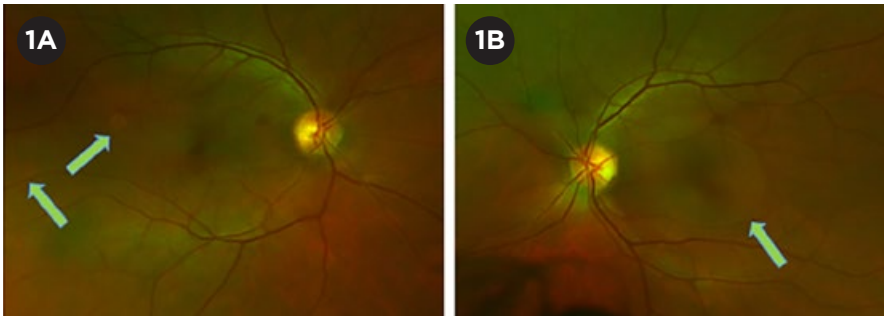
The Case of the Black Spot and Blurred Lines

A few weeks before the holidays, while working long hours, Sean Rodriguez* began to notice a new black spot in his vision. The spot lingered in the center of the vision in his left eye and would not disappear. At first, he convinced himself that stress and weariness were causing him to imagine things. But the spot grew in size, and the letters on his computer screen began to blur. After 3 weeks, he grew frantic and rushed to see us.



WE GET A LOOK. *His right eye (1A) has a small PED superotemporally at the edge of the macula (arrows); his left eye (1B) has a large serous detachment (arrow).*

The Presentation

Mr. Rodriguez sat in the exam room chair, tapping his foot anxiously.

Medical history. The 34-year-old Hispanic male was generally healthy. He sometimes suffered from bouts of asthma, for which he used an albuterol or fluticasone-salmeterol inhaler.

Ocular history. He had previously been diagnosed with anatomic narrow angles and had undergone bilateral laser peripheral iridotomies in 2012.

Social history. He worked as a hospital case manager during the day and was attending night school to fulfill his dreams of becoming a nurse practitioner. He was feeling overwhelmed with his workload.

The Exam

On initial examination, vision with correction was 20/20 in the right eye; it was 20/200 in the left eye, improving with pinhole to 20/60. Mr. Rodriguez's pupils were round and reactive with no relative afferent pupillary defect. Intraocular pressure was 16 mm Hg in both eyes. Extraocular movements and confrontation visual fields were full.

The anterior segment exam was notable for patent superior peripheral iridotomies in both eyes. On posterior segment examination, a small pigment epithelial detachment (PED) was noted superotemporally at the edge of the macula of his right eye (Fig. 1A), and there was a large serous detachment in the macula of his left eye (Fig. 1B).

Notably, there were no retinal tears or breaks, retinal hemorrhages, drusen, or optic nerve pits in either eye.

Differential Diagnosis

The leading diagnosis at this time was central serous retinopathy (CSR), given the presence of a macular serous detachment in a young male with active stressors.

Other potential diagnoses included an optic pit with associated serous retinal detachment (although no optic pits were noted on exam), age-related macular degeneration (although the patient was younger than the usual demographic affected by this condition), or an inflammatory choroidal disorder.

Tests and Final Diagnosis

Optical coherence tomography (OCT) imaging confirmed a small PED in the superotemporal macula of the right eye (Figs. 2A, 2B). In the left eye, there was a large amount of subretinal fluid, a small superonasal PED, and a thickened choroid (Figs. 2C, 2D).

Autofluorescence images revealed a circular area of hypoautofluorescence in the area of the small PED in the right eye (Fig. 3A) and a large area of central hypoautofluorescence corresponding to the area of subretinal fluid in the left eye (Fig. 3B).

Lastly, a fluorescein angiogram (FA) showed a circular area of leakage in the area of the right eye's PED (Fig. 3C) and a progressive pattern of leakage

BY **NANDINI VENKATESWARAN, MD,** AND **HARRY W. FLYNN JR., MD.**
EDITED BY STEVEN J. GEDDE, MD.

resembling a "smokestack pattern" in the left eye's macula (Fig. 3D).

All of these findings confirmed our leading diagnosis of bilateral CSR, which was symptomatic in the left eye and asymptomatic in the right eye.

Discussion

CSR—first described by J. Donald M. Gass, MD, in 1967—is a condition characterized by serous retinal detachments and/or retinal PEDs, often found in the macular region.[1,2]

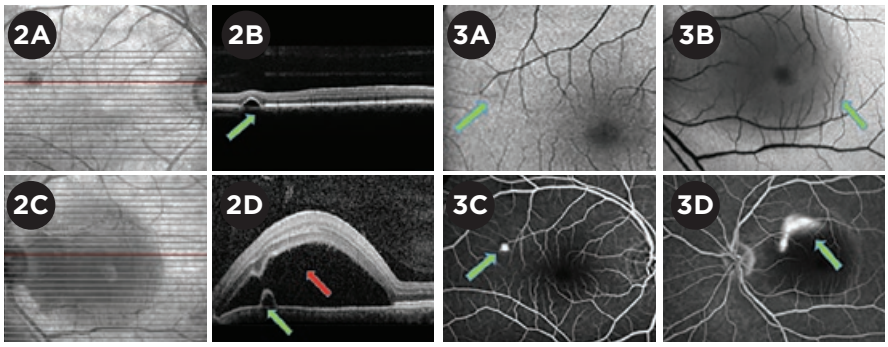
Symptoms. Typical symptoms include blurred vision, micropsia, metamorphopsia, scotomas, and decreased contrast sensitivity; however, patients can often be asymptomatic if the macula is not involved.[2]

Demographics. CSR classically affects men in their third to fifth decades of life. Asians and African Americans are thought to be at greater risk.[2]

Risk factors. Major risk factors for the condition include high levels of stress and/or a type A personality,[3] exogenous steroid or testosterone use, Cushing syndrome, and pregnancy,[4] all of which can cause elevations in systemic corticosteroids or cortisol.

Pathogenesis. Rises in serum glucocorticoid or catecholamine levels affect vascular autoregulation and retinal pigment epithelium (RPE) function. Many hypothesize that in CSR the choroidal vasculature becomes hyperpermeable.[5] Vessel hyperpermeability leads to an increase in hydrostatic pressure that overwhelms the barrier function of the RPE. Consequently, there may be progressive choroidal thickening, PED formation, and subretinal fluid accumulation.[2,5]

Imaging. Multimodal imaging is powerful in establishing this diagnosis. OCT can identify neurosensory retinal detachments and PEDs. Enhanced-depth OCT images, in particular, can measure choroidal thickness; a thickened choroid is characteristically seen in patients with CSR. FA can identify active leaks causing accumulation of subretinal fluid. Patterns of leakage observed include an inkblot pattern, where a small, circular, hyperfluorescent leak increases gradually in size, or a smokestack pattern, where a point of leakage expands and extends vertically to resemble a plume of smoke, as was seen in our patient. Indocyanine green imaging often shows dilated and engorged choroidal vessels with concomitant leakage, corresponding to the areas of leakage identified on FA.[2]



IMAGING. *OCT of the right (2A, 2B) and left (2C, 2D) eyes confirmed small PEDs in each (green arrows) and a large amount of subretinal fluid in the latter (red arrow). Autofluorescence images showed areas of hypoautofluorescence: In the right eye (3A), this was circular and in the area of the small PED (arrow); in the left eye (3B), this was larger and corresponded to the area of subretinal fluid (arrow). Fluorescein angiograms showed leakage in both eyes: In the right eye (3C), it was circular and in the area of the PED (green arrow); in the left eye (3D), there is a progressive pattern of leakage resembling a smokestack in the macula (green arrow).*

In a subset of patients, CSR can be a chronic condition. For those patients, subretinal fluid often lasts for more than 3 months, and patients are at a 30%-50% risk for having recurrences. On exam and imaging, patients with chronic CSR can have unremitting intraretinal cystoid edema and RPE atrophy. In rare cases, choroidal neovascular membranes (CNVM) can be identified.[2] (Also, see Journal Highlights, page 21.)

Management

The treatment goals for CSR are to improve or preserve visual acuity, induce reattachment of the retina, and prevent further recurrences of the condition.

The condition is often self-limiting, usually requiring a 3-month period to allow for the subretinal fluid to resolve spontaneously. Lifestyle modification is paramount; patients must reduce stressors and discontinue steroid use (oral, intranasal, or topical).[6]

Several additional management options have been investigated. Trials in the 1980s looked at laser coagulation to the RPE to hasten resolution of subretinal fluid, but this led to formation of permanent scotomas, enlargement of laser scars, or CNVM formation.[2]

Photodynamic therapy (PDT) has been used, as it induces choroidal hypoperfusion and vascular narrowing and remodeling, and is thought to tighten the blood retinal barrier. Studies have shown resolution of macular detachments with improved vision with half-dosage or half-fluence therapy, but risks include choroidal ischemia, RPE atrophy, and CNVM formation.[6]

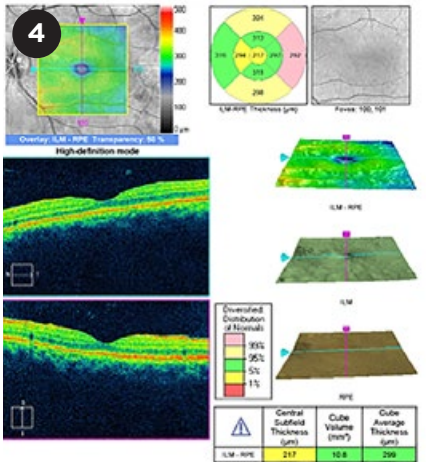
Micropulse laser (MPL) has also been used. A recent study found PDT to be more effective than MPL for chronic CSR, but additional studies are needed to further elucidate MPL's role.[7]

Anti-vascular endothelial growth factor (VEGF) drugs have been utilized for CSR and for CNVM associated with CSR, targeting hypoxic conditions in the choroid or RPE that can lead to VEGF production; however, evidence supporting their use is mixed.[8]

Mineralocorticoid receptor antagonists, such as spironolactone and epleronone, are oral medications that have shown potential in the treatment paradigm of CSR. They have shown reduction of subretinal fluid and central macular thickness in studies. However, standardized dosage and duration of therapy continue to be determined.[9]

Our Patient

We urged our patient to try to reduce his stress levels; and we also told him



SIX MONTHS LATER. *He remains 20/20 in his left eye, with OCT revealing no recurrence of subretinal fluid.*

to discontinue his fluticasone-salmeterol inhaler as tolerated. One month after his initial presentation, his vision improved to 20/30 in the left eye with inferior displacement of the subretinal fluid. Six months later, he remains 20/20 in the left eye with no recurrence of symptoms (Fig. 4). His right eye still has a small but stable PED that is not visually significant. He is followed every 6 months or sooner if needed.

* Patient name is fictitious.

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Dr. Venkateswaran is a current third-year resident at the Bascom Palmer Eye Institute. **Dr. Flynn** is the J. Donald M. Gass Distinguished Chair in Ophthalmology and Professor of Ophthalmology at the Bascom Palmer Eye Institute and is a retina specialist. *Financial disclosures: None.*

Morning Rounds: 2018 in Review

TACKLE THIS YEAR'S MEDICAL MYSTERIES. See this article online where you'll find links to the 10 cases below (aao.org/eyenet).

- **A Case of Aches and Pains and Blurry Vision.** Becky Brown,* a 28-year-old Caucasian woman, had experienced flu-like symptoms—followed by red, painful, photophobic eyes and decreased vision. (January.)
- **The Mystery Choroidal Lesion.** Tabitha Tisch* was anxious. For several hours, the 53-year-old experienced episodic diplopia, had difficulty breathing, and struggled to find words. (February.)
- **Is This Déjà Vu?** Iris Brown* is an 85-year-old woman who came to our clinic complaining of a slowly progressing, painless decrease in vision in her left eye over the last year. We noted a white-yellow opacity that resembled a nuclear sclerotic cataract—but she had undergone cataract surgery 7 years earlier! (April.)
- **An Unusual Case of Left-Sided Vision Loss.** Janet Jenkins,* an active 73-year-old woman, first presented to her optometrist with the chief complaint of a 1-week history of a new green tint to her vision. Within a few weeks, her mental status had altered and she was blind. (May.)
- **Doctor, There's a Screaming Sound in My Ears!** Laura Mitchell,* an 11-year-old girl, was plagued by worsening headaches and a "screaming" that she sometimes heard for hours. (June.)
- **"The Most Thorough Examination I've Ever Had."** It was turning out to be a long day for Gerard Gooman.* He initially saw his optometrist for a floater and was now sitting in the sub-waiting room of the busy ophthalmology office waiting for a diagnostic test. (July.)
- **Rethinking a Case of Chronic Scleritis.** Meiling Chen* is a 59-year-old Taiwanese woman. She complained of ocular irritation and redness in her left eye, starting about 4 months earlier. Despite topical nepafenac and oral ibuprofen, her left eye was still red and inflamed. (August.)
- **Foggy With a Chance of Hemorrhage.** Tatiana Ivanov,* a 79-year-old Albanian woman now living in New York City, grimaced and held her hand over her right eye. She reported 4 days of blurry vision and 2 days of severe pulsating eye pain and worsening vision. Now, she said that she could see only "fog" with her right eye. (September.)
- **The Case of Enigmatic Lid Swelling.** Charles Prince,* a 53-year-old, had a presumptive diagnosis of orbital cellulitis—but he didn't respond to intravenous antibiotics or corticosteroids. (October.)
- **A Bothersome Bump.** One morning, 8-year-old Mark Mario* woke up with a tender, swollen left eyelid. After several days of worsening swelling and pain, Mark's mother sought help. (November.)

* Patient names are fictitious.

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