“Optic neuritis” that is neither

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A 28-year-old African American woman developed periocular pain worsening with eye movement and decreased vision in her left eye. A diagnosis of left optic neuropathy was made based on the findings of decreased vision in the left eye, a left relative afferent pupillary defect, and mild optic disc hyperemia in the left eye (figure 1), with normal retinæ and maculae bilaterally. Automated perimetry showed a small central scotoma in the left eye. The pain with eye movements suggested an inflammatory mechanism (i.e., optic neuritis). Her neurologic examination was normal. MRI of the brain showed 2 small T2 hyperintense lesions in the right inferior frontal lobe and left peritriangular white matter. The lesions did not enhance. Angiotensin converting enzyme level was slightly elevated.

Figure 1 Fundus photograph of the left eye at the time of the episode of optic neuritis

There is mild optic nerve edema, and the optic nerve is hyperemic.

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A chest CT was negative for pulmonary sarcoidosis. She did not receive steroids, and her visual function improved spontaneously over a few weeks. She had no neurologic symptoms and the diagnosis of clinically isolated syndrome was made. The patient declined treatment with an immunomodulatory agent.

Five years later, she noted again decreased vision in her left eye. Initial evaluation by an optometrist documented visual acuity of 20/20 in the right eye and 20/40 in the left eye, with a small left relative afferent pupillary defect, and the diagnosis of recurrent optic neuritis was made. She was then sent to our institution for repeat MRI, lumbar puncture, neuromyelitis optica antibody testing, treatment with IV steroids, and discussion of long-term treatment for presumed demyelinating disease. She was seen in neuro-ophthalmology, 4 weeks after the onset of symptoms, at which time vision in the left eye was already spontaneously improving. She denied pain with eye movement. Visual acuity was 20/15 in the right eye and 20/25 in the left eye. Color vision was full and she had a trace left relative afferent pupillary defect. Humphrey visual fields showed a central scotoma in the left eye (figure 2A).

Fundus examination showed mild optic nerve pallor on the left, consistent with her previous episode of optic neuritis (figure 2B), but there were pigmentary changes and elevation of the macula in the left eye (figure 2C), suggesting a maculopathy. Imaging of the macula with Cirrus spectral-domain optical coherence tomography (Carl Zeiss Meditec, Dublin, CA)

Figure 2 Evaluation at the time of the second episode of visual loss in the left eye (5 years after the photograph shown in figure 1)

(A) Humphrey visual field of the left eye (gray scale at the top and pattern deviation at the bottom) shows a central depression in the left eye (black squares on the pattern deviation). (B) Left eye fundus photograph centered on the optic nerve shows the absence of disc edema; there is mild optic nerve pallor compared with figure 1. (C) Left eye fundus photograph centered on the macula shows the absence of the normal foveal reflex, a few exudates, and elevation of the fovea, which looks like a blister (arrows). (D) Spectral domain optical coherence tomography of the left macula shows the blister with subretinal fluid (red arrow) nasal to the fovea. (E) Late phase of the retinal fluorescein angiography obtained on the left eye shows the typical focal leakage of fluorescein seen with central serous chorioretinopathy (note the hyperfluorescent spot superonasal to the fovea [arrow], which represents fluid leakage from the choroid through the retinal pigment epithelium).

showed an exudative macular detachment in the left eye (figure 2D). Retinal fluorescein angiography demonstrated focal leakage superonasally to the fovea (figure 2E) consistent with central serous chorioretinopathy. Her vision further improved spontaneously over the next few weeks.

DISCUSSION

Central serous chorioretinopathy (CSR) is a common retinal disorder (incidence 10 per 100,000 in men and 1.7 per 100,000 in women)¹ which usually presents as isolated acute unilateral visual loss. It often improves spontaneously, similar to idiopathic optic neuritis.² Because CSR is a localized disorder of the retina, patients have no pain, normal pupils with no relative afferent pupillary defect, but have decreased visual acuity and a central scotoma when it involves the macula.³ Our patient had a previous history of optic neuritis, explaining why she had a small residual relative afferent pupillary defect and mild optic nerve pallor when seen 5 years later.

Because a diagnosis of recurrent optic neuritis would have led to the diagnosis of probable multiple sclerosis⁴ in our patient, with important implications regarding the patient's treatment and prognosis, it is important to be aware of mimics of optic neuropathies, such as CSR.^{5,6} Furthermore, steroids, if given to treat her "optic neuritis," could have worsened her CSR, which represents a classic contraindication to steroid treatment.³ Fortunately, a comprehensive ophthalmologic examination led to the correct diagnosis of a benign and usually self-limited retinal condition in our patient. This case demonstrates the importance of collaboration between ophthalmologists and neurologists when patients experience visual loss.⁷

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