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Clinical challenge

Beyond what the eye can see

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1. Case report

A 45-year-old woman presented with a 4-week history of visual loss. She described an initial central scotoma and color desaturation in her left eye. Over several days, her peripheral vision became involved. Two weeks after onset, she noted similar symptoms in her right eye, which also progressed to severe visual loss. There was no associated eye pain, headache, or systemic symptoms.

She had a history of chronic kidney disease, diabetes mellitus, hearing loss, and preeclampsia. Preeclampsia had developed during all 3 of her pregnancies and resulted in 1 stillborn child and 2 premature deliveries. The preeclampsia was presumed to have resulted in chronic kidney disease. Diabetes had been diagnosed 8 years before presentation, and the patient had moderate to poor blood sugar control with oral hypoglycemics. Sensorineural hearing loss occurred suddenly and simultaneously 7 years previously, with no recovery. Investigations at the time failed to reveal an underlying cause.

Social history was unremarkable. She did not drink alcohol or smoke cigarettes. She denied illicit drug use or recent vaccination. She was a stay-at-home mother to her 2 teenage children.

Family history was relevant for late-onset diabetes mellitus in her mother and maternal grandmother. Her mother had congenital hypoplasia of the left side of the body, which included left-sided hearing loss. This was attributed to an intrauterine injury.

Initial examination revealed best corrected visual acuities of 6/60, N24 in the right eye, and count fingers at 1 m in the left

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eye. There was a left relative afferent pupillary defect. There was no evidence of ocular inflammation. Funduscopy revealed widespread granularity of the retina. There were no abnormalities of the optic disks. Fluorescein angiography was unremarkable (Fig. 1).

What is the differential diagnosis of the visual loss, and how would you proceed?

2. Comments

2.1. *Comments by Jason J. S. Barton, MD, PhD, FRCPC*

Acute bilateral visual loss has a fairly long list of possibilities, ranging all the way from the retina to striate cortex. The fact that this was sequential, with one eye affected before the other, is helpful: it excludes retrochiasmal lesions, which if sequential would affect one hemifield before the other instead. Hence, the main possibilities are bilateral retinopathy or optic neuropathy.

Could this be bilateral inflammatory retinopathy? Young women in particular are vulnerable to an array of retinal disorders known by their acronyms: AZOOR, PIC, ARPE, AMPPE, multiple evanescent white dot syndrome, and so on. These generally present in similar subacute fashion, can affect visual acuity, and some have a tendency to bilateral involvement. However, most have characteristic white spots or other lesions on funduscopy and abnormalities on fluorescein angiography. Acute idiopathic blind spot enlargement syndrome and multiple evanescent white dot syndrome can have "granularity," although usually limited to the macula and evolving in later stages.[9,31] The widespread nature of the granular changes mentioned here sounds different.

Bilateral sequential optic neuritis is a possibility. Somewhat atypical is the lack of pain. In the Optic Neuritis Treatment Trial, 92% of patients had pain, and in 87%, it was worse or only present with eye movement.[1] Nevertheless, common things being common, this has to be a leading candidate. Although bilateral sequential involvement can occur with multiple sclerosis, this should always raise suspicion for neuromyelitis optica, especially if the patient is Asian.

Infectious causes of inflammatory optic neuropathy are more unusual but can present bilaterally in a subacute fashion as well. Aspergillus and mucormycosis are fungal infections that involve the sinuses: these occur more frequently in patients with diabetes or other conditions that compromise the immune system. Syphilis, Lyme disease, and Bartonella are also important to consider, particularly if there are retinal exudates—the so-called macula star. Tuberculosis can cause a granulomatous basal meningitis that can affect both optic nerves.

Bilateral acute sequential visual loss can occur with anterior ischemic optic neuropathy, for which diabetes and hypertension are risk factors[3]; however, disk edema should be evident and only in a minority does visual loss progress over the days after onset. Posterior ischemic optic neuropathy is far less common and occurs primarily in the setting of giant cell arteritis, for which she is too young, or severe prolonged hypotension, often in the perioperative state.[3,10]

We are not told her medications, but we can infer from her past history that she is probably not taking ethambutol or amiodarone. Methanol is a cause of acute bilateral optic

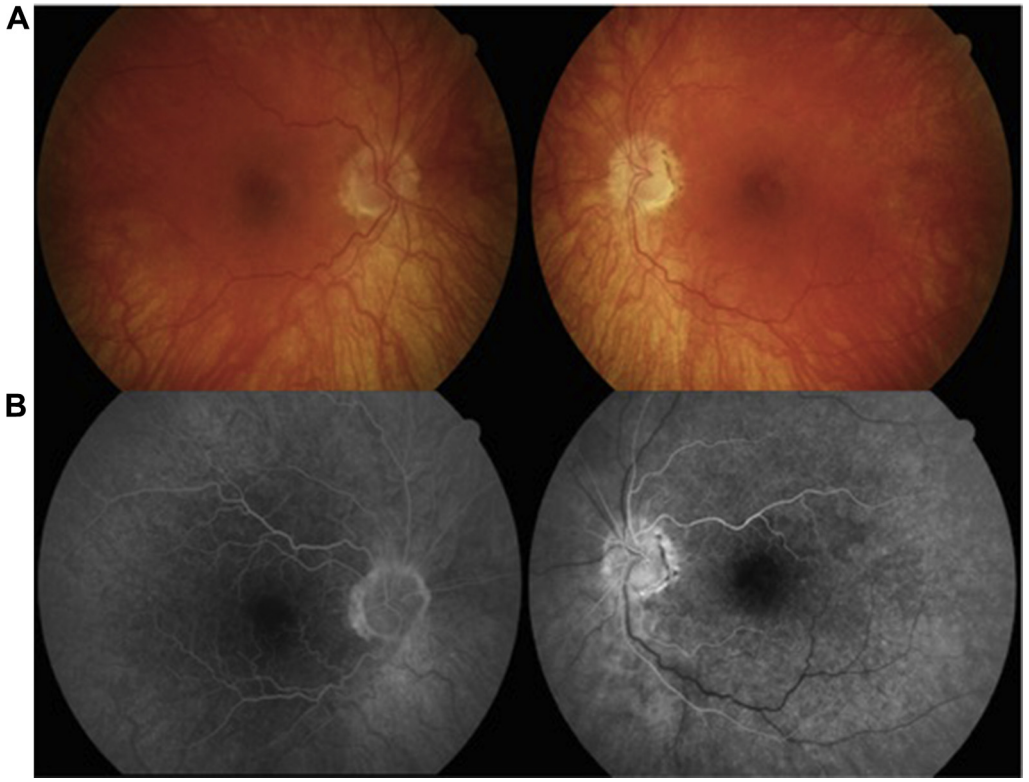

Fig. 1 – *A*: Color fundus photos right and left eye. *B*: Normal arterial phase fluorescein angiogram.