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CLINICAL CHALLENGES

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Can Lightning Strike Twice?

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(In keeping with the format of a clinical pathologic conference, the abstract and key words appear at the end of the article.)

Case Report. A 73-year-old woman presented with complaints of decreased vision in her right eye (OD). She described a darkness in her vision OD for 2 days. On further questioning she described jaw stiffness and pain with chewing for the 2 weeks prior to presentation. The patient had a known history of biopsy-proven giant cell arteritis (GCA) diagnosed 5 years previously. She presented at that time with a loss of vision in her left eye (OS), scalp tenderness, and jaw stiffness. Her funduscopic examination revealed a swollen optic disk OS. She was treated with high-dose prednisone and had successfully completed a taper over the ensuing 13 months. Her vision OS remained hand motion. She had been off corticosteroids for 54 months during which time she never had a recurrence of any of her symptoms, further visual loss or an erythrocyte sedimentation rate (ESR) $>$ 12 mm/hr. She noted that her current jaw stiffness was similar to what she experienced 5 years before.

Visual acuity was 20/25 OD and hand motion OS. She correctly identified 9/10 color plates OD but was unable to identify the test plate OS. Her slit-lamp examination was unremarkable. She had a prominent left relative afferent pupillary defect (RAPD). Automated perimetry on the right and the optic disks are seen in Figs. 1 and 2, respectively.

What further information and/or testing do you suggest?

What are the diagnostic possibilities?

Comments

Comments by Rod Foroozon, MD

Looking at the fundus photographs, the right optic disk is pale (except for a small temporal portion) and swollen. Preservation of the temporal portion and therefore the papillomacular bundle is presumably the reason the visual acuity has been relatively spared. The degree of pallor is perhaps not as severe as that typically suggested by the "chalky white" appearance of arteritic anterior ischemic optic neuropathy (AION), a hallmark of GCA;[14,26,27] however, the funduscopic appearance in Fig. 2 is consistent with AION from GCA. The optic disk findings on the previously affected left side are a bit unexpected in that the degree of optic disk cupping is not as great as that typically seen after arteritic AION.[8,10] There is mild retinal arteriolar attenuation on the left that can be seen both in ischemic optic neuropathy and after central retinal artery occlusion (CRAO). A large left RAPD would be more suggestive of an optic neuropathy than a retinopathy. Although AION is the most

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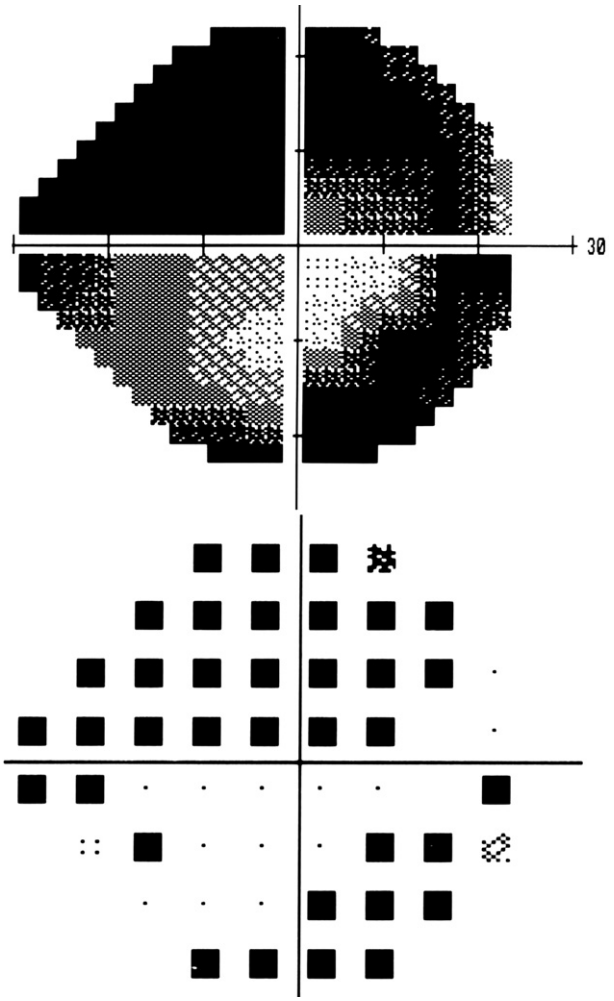

Fig. 1. Humphrey perimetry showing a superior altitudinal defect in the patient's right eye; she was unable to do formal perimetry for the left eye because of poor acuity.

common cause of visual loss in GCA, obtaining the prior records would confirm the site of visual loss in the left eye.

It would be nice to get the prior medical records detailing the ophthalmic findings and the prior histopathology of the temporal artery biopsy. Prior fundus photographs would also be helpful to confirm prior optic disk edema. Additional historical details that are not mentioned in the case description but might be helpful include the use of any medications that might precipitate ischemic optic neuropathy (interferon-alpha[23] for example) and a history of hypotension or anemia suggestive of shock-induced AION.[11] It would be important to know when the last ESR (and other markers of inflammation, platelets or C-reactive protein) was performed and to document other symptoms consistent with GCA such as polymyalgia, decreased appetite, and headache. A history of prior transient visual loss before the onset of fixed visual loss in the right eye would also be more suggestive of arteritic AION.

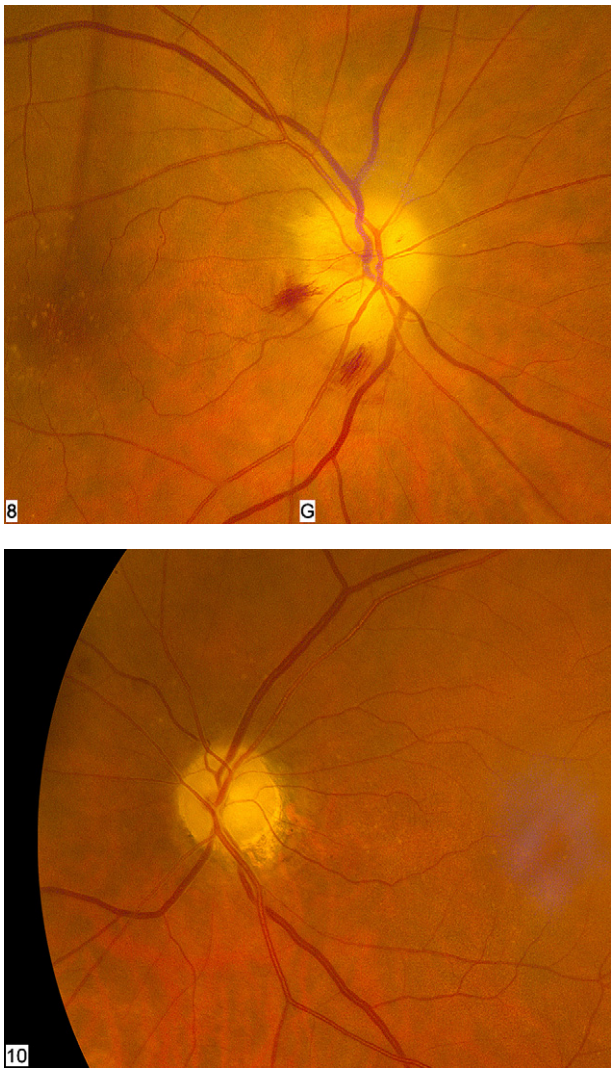

Fig. 2. *Top:* Right eye, pale swollen optic nerve with nerve fiber layer hemorrhages. *Bottom:* Left eye, pale cupped optic nerve.

From this examination we know there is evidence of an optic neuropathy on each side, with optic disk edema on the right that appears consistent with ischemic optic neuropathy. This patient's findings raise important questions:

- Can visual complications of GCA occur years after the disease is successfully treated?
- How often should a patient with biopsy-proven disease undergo blood testing?
- Is it possible to have non-arteritic AION occur in a patient known to have GCA?

The first question is much easier to answer in the affirmative. Whereas treated GCA is typically thought to have a more active phase followed by a more quiescent phase, there are reports of subsequent visual loss with findings suggestive of arteritic AION.[4,12,19] Unlike the present patient who had jaw pain with chewing, some