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CLINICAL CHALLENGES

A Weed by Any Other Name

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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 39-year-old man presented with complaints that his vision had grown progressively gray or cloudy over the past month, with a 'smoky' appearance and episodes of intermittent tunnel vision. In addition, he complains of persistent headache. He denied any diurnal variation and weakness, numbness, or paresthesias. He was a contact lens wearer. Current medical diagnoses include anxiety, depression, and obstructive sleep apnea. He used a CPAP machine at night and was on Paxil. His father, who died at age 45, had a brain stem tumor. He used recreational marijuana and alcohol, but denied tobacco abuse.

Best corrected visual acuity was 20/20 OU, and confrontational visual fields were full to finger counting in all four quadrants, with intraocular pressures of 17mmHg in both eyes. AOHRR color plates were 15/15 OU, and his pupils were equal and reactive, with no relative afferent papillary defect. The slit-lamp examination was normal except for slight inferior corneal pannus OS, consistent with his contact lens use. Automated perimetry is seen in Fig. 1, and dilated fundus examination, in Fig. 2. The macula and periphery were normal in both eyes.

What is the differential diagnosis?

What work-up would you propose?

Comments

COMMENT BY VALERIE PURVIN, MD

The patient is a generally healthy man with recent headaches and bilateral, asymmetric optic disc elevation. Despite his description of 'smoky' vision, optic nerve function is in fact normal. His visual field shows enlargement of the physiologic blind spot, which is simply a reflection of the disc swelling, not an indication of optic nerve dysfunction. The differential diagnosis of optic disc edema with normal optic nerve function is wide, but some causes can be ruled out easily.

We might first consider whether his disc elevation might represent pseudopapilledema, as from buried drusen. Careful inspection of the fundus photographs, however, shows definite opacification of the retinal nerve fiber layer and subtle Paton lines adjacent to the disk's edge, confirming that this is acquired disc edema rather than congenital elevation. In individuals with Leber hereditary optic neuropathy the optic discs often give the appearance of swelling, termed pseudo-edema, but increased blind spots and Paton- lines are not present.

The first consideration in a patient with bilateral acquired disc edema and normal optic nerve function is increased intracranial pressure (ICP), which can be due to a mass lesion, hydrocephalus,

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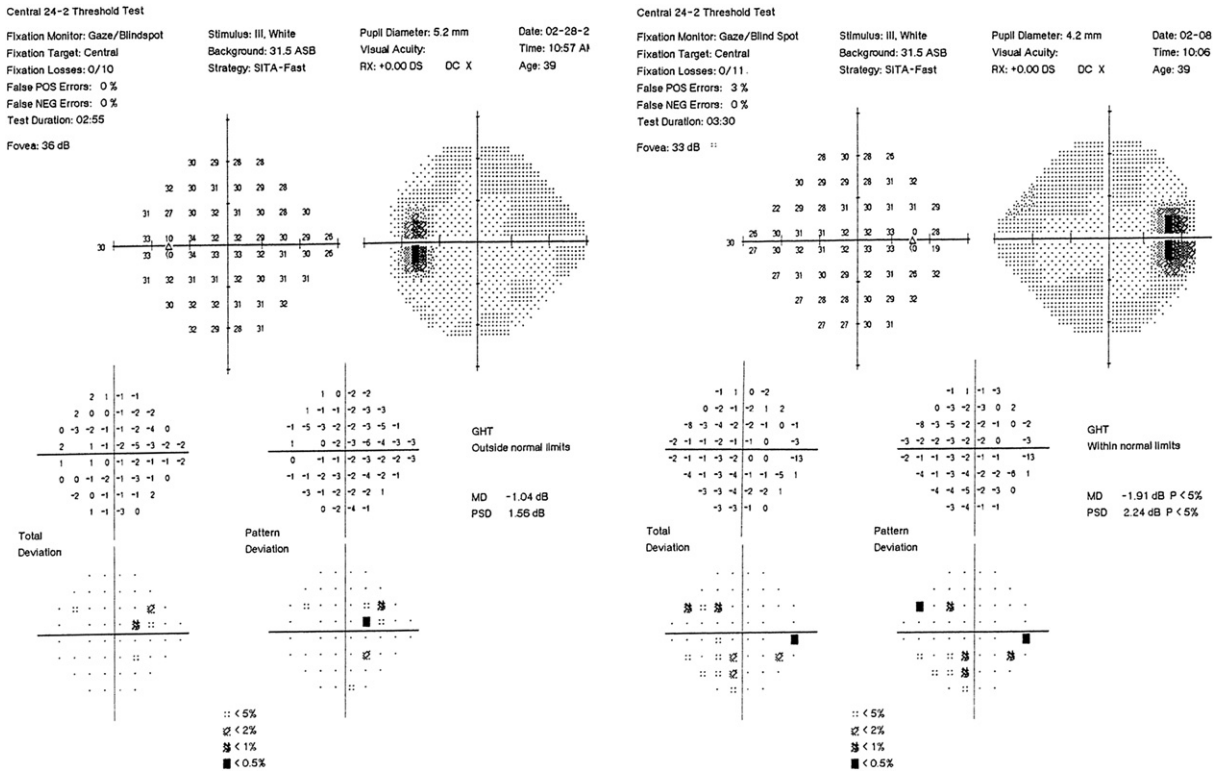

Fig. 1. Humphrey visual field 24-2 with increased blind spots in both eyes and a questionable early arcuate defect in the right eye.

meningitis or pseudotumor cerebri (PTC) syndrome. A magnetic resonance imaging (MRI) scan with contrast should be obtained to investigate these possibilities. One specific cause of PTC is cerebral venous thrombosis, which may not be visible on standard MRI sequences. To more fully investigate this possibility, an MRV could be obtained as part of the initial evaluation or, alternatively, could be added if the MRI is non-diagnostic.

Disk edema with good vision can also be an early sign of optic nerve compression, but this is unlikely to affect both eyes simultaneously. Including orbit views with fat suppression as well as brain images will investigate this possibility. In some cases of anterior ischemic optic neuropathy, there is a pre-infarctive stage in which axoplasmic transport is compromised so that disc edema is present, but neural transmission is still functioning. This too is highly unlikely to affect both eyes at the same time. Causes such as posterior uveitis and decreased intraocular pressure have already been excluded by the otherwise normal eye examination.

Certain toxic-metabolic conditions can cause bilateral disc edema in the absence of increased ICP, most notably cyclosporine and amiodarone, but these are ruled out by the history. The POEM syndrome (polyneuropathy, endocrinopathy, and monoclonal gammopathy) is a rare cause of bilateral disc edema, but its other features are lacking here.

Obstructive sleep apnea syndrome can cause disc edema, and interestingly this patient has a history of this condition. The mechanism in such cases is thought to involve nocturnal elevation of ICP from hypoxia-induced venous dilation, increased intrathoracic venous pressure from forced expiratory effort against a closed glottis, and possibly other factors. In such individuals the daytime ICP is normal or borderline, but elevations can be documented during sleep. Depending on the results of his other testing, the effectiveness of his CPAP could be re-evaluated.

If the MRI does not show a mass lesion or hydrocephalus, the next step in the work-up would be a lumbar puncture, to include measurement of opening pressure as well as routine cerebrospinal fluid tests. Additional evaluation would be based on results of the above testing.

Case Report (Continued)

MRI, MRA, and MRV were all interpreted as normal (Fig. 3). Lumbar puncture revealed an opening pressure of 41 cm water and an elevated cerebrospinal fluid (CSF) protein (79.0 mg/dl – normal 15 – 45 mg/dl). Autoimmune, endocrine,