SURVEY OF OPHTHALMOLOGY VOLUME 55 • NUMBER 3 • MAY–JUNE 2010

ELSEVIER

CLINICAL CHALLENGES

PETER SAVINO AND HELEN DANESH-MEYER, EDITORS

Can't Hear, Can't See, and Too Sore to Play

Ryan A. Scheurer, BS,[1] Gregory S. Kosmorsky, DO,[2] Gary S. Hoffman, MD,[3] Carol Farver, MD,[4] Michael S. Lee, MD,[1] and Dean M. Cestari, MD[4]

[1]Department of Ophthalmology, University of Minnesota, Minneapolis, Minnesota, USA; [2]Departments of Ophthalmology, Cleveland Clinic Foundation, Cleveland, Ohio, USA, [3]Rheumatology, Cleveland Clinic Foundation, Cleveland, Ohio, USA; and [4]Neuro-ophthalmology Unit, Massachusetts Eye and Ear Infirmary, Boston, Massachusetts, USA.

(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 52-year-old man presented to neuro-ophthalmology with intermittent, bilateral, transient visual obscurations lasting seconds. He denied headache, double vision, or audible bruit. He denied scalp tenderness, jaw claudication, or history of oral or genital ulcers.

He had a past history of steroid responsive, transient, migratory polyarthralgias for 3 years. One year prior to presentation, he developed persistent leukocytosis and thrombocytosis leading to a bone marrow biopsy, which demonstrated one non-caseating granuloma. Chest x-ray and CT (computed tomography) revealed a resolving right pulmonary infarction. Normal test results included CBC, complete metabolic panel, rapid plasma reagin, fluorescent treponemal antibody absorbed, erythrocyte sedimentation rate, Lyme titer, angiotensin converting enzyme, anti-nuclear antibody, protein purified derivative, liver function tests, serum protein electrophoresis, anti-neutrophil cytoplasmic antibody, gallium scan and human leukocyte antigen B27. His c-reactive protein was 2.4 (normal <0.5). A year later, audiometry revealed bilateral high- and mid-frequency hearing loss. An EMG was normal.

He took enalapril for mild hypertension. He had a history of a basal cell carcinoma removed from his left scalp. He was adopted. He drank two beers/week and smoked 10 cigarettes a day for 25 years. He traveled extensively outside the United States.

Visual acuitiy was 20/15 and J1 in each eye. His neuro-ophthalmologic examination was unremarkable except for bilateral optic disk edema (Fig. 1). Kinetic perimetry revealed an enlarged blind spot OU and mild superior constriction OS.

What is your differential diagnosis and how would you proceed?

Comments

COMMENTS BY DEAN CESTARI, MD

The optic nerve is composed of 1.2 million axons, and each axon must maintain active axonal transoport in the orthograde and retrograde directions. The subarachnoid space of the brain is continuous with that of the optic nerve sheath, and a myriad of insults may inhibit axoplasmic transport resulting in optic disk edema. When the etiology of optic nerve

0039-6257/10/$–see front matter
doi:10.1016/j.survophthal.2009.09.002

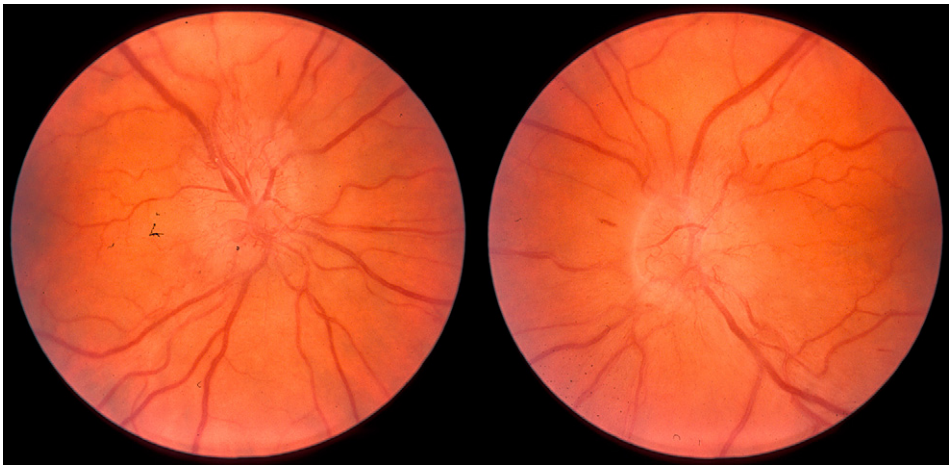

Fig. 1. Both optic disks are swollen with a splinter hemorrhage bilaterally.

dysfunction is elevated intracranial pressure, disk edema is bilateral and it is referred to as *papilledema*.

The ophthalmoscopic characteristics of papilledema include blurring of the optic disk margins, filling-in of the optic disk cup, elevation of the optic nerve head, edema of the retinal nerve fiber layer, and retinal/choroidal folds. Vascular signs include venous congestion of arcuate or peripapillary vessels, papillary and peripapillary hemorrhages and/or exudates, retinal nerve fiber layer infarcts (cotton-wool spots), and hyperemia of the disk.

Symptoms of increased intracranial pressure include headache and brief transient obscurations of vision (TOV). Less commonly patients may complain of blurred vision, double vision, and, late in the process, loss of peripheral vision.

The differential diagnosis of papilledema is disk edema not caused by increased intracranial pressure (compressive optic neuropathy, papillitis, diabetic papillopathy, vitreo-papillary traction,[3,18] and anterior ischemic optic neuropathy, to name a few) and pseudopapilledema.

This patient has a history of TOV that lasts seconds. It is important to recognize that very brief episodes of TOV can be reported by patients with optic nerve swelling from any cause, not just papilledema. In this patient, the fundus examination reveals bilaterally swollen optic nerves without exudation, hemorrhage, or optic disk drusen. Given the symptoms of TOV and the bilaterally swollen optic disks, elevated intracranial pressure must be excluded as the etiology. The patient should have an MRI of the brain with gadolinium to exclude a space occupying lesion or an obstructive hydrocephalus, especially given his history of a bone marrow biopsy that shows a non-caseating granulomatous disease. If the magnetic resonance imaging (MRI) is normal, a magnetic resonance venogram (MRV) should be obtained to exclude a dural venous sinus thrombosis that can cause intracranial hypertension.

Following these tests, a lumbar puncture should be obtained to measure the opening pressure; routine cell count and chemistries should be obtained to exclude an infectious, inflammatory, or malignant process.

Case Report (Continued)

MRI of the brain/orbits with gadolinium was normal. MR venogram was not performed. A lumbar puncture demonstrated an opening pressure of 170 mm H20, 11 white blood cells (WBCs) (lymph 62%, mono 20%), and normal protein and glucose. Special stains, cultures, and cytology were negative. Two months later, he developed a posterior vitreous detachment with prominent Weiss rings over the disk in each eye, and the disk edema had improved (Fig. 2). The patient was diagnosed with pseudodisc edema secondary to vitreopapillary traction.[3,18]

Meanwhile, azathioprine 150 mg and prednisone 20 mg were instituted for worsening arthralgias and myalgias. He did not complain of ocular symptoms for 7 months until he developed a sudden loss of visual field OD. His acuities were 20/30 OD and 20/20 OS. Examination revealed a trace relative afferent pupillary defect (RAPD) OD, blurred optic disk margins, and 2+ vitreous cells OU. Visual fields showed a superior altitudinal defect OD. Patches of retinal edema were noted in the inferior macula OD, along with sheathing and occlusion of an inferotemporal retinal artery. Fluorescein angiography revealed leakage from both optic disks (Fig. 3).

What is your differential diagnosis and how would you proceed?