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CLINICAL CHALLENGES

PETER SAVINO AND HELEN DANESH-MEYER, EDITORS

Glazed (Vision) and Confused

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

Case Report

A 60-year-old man presented for neuro-ophthalmic consultation for progressive blurred vision over 2 weeks associated with black spots in all visual fields. He denied double vision or positive visual phenomena. He had had a normal screening ophthalmologic examination 4 months before.

He had been well until 3 months earlier when he had flu-like symptoms of fever, fatigue, and malaise for several days followed by the onset of severe frontal headaches without phonophobia or photophobia. One month after the headaches started he developed depressed mood and anxiety. Three months after the headaches started he became confused and forgetful. He forgot the code for his house alarm and how to use a cellular phone. He had an episode when he awoke confused and then had brief loss of consciousness with his eyes rolled back. He had no recollection of this event. The visual symptoms started 1 week later. Review of systems included a 45-pound weight loss attributed to medication, occasional dizziness, diffuse pain, and trouble walking due to hip pain.

He was evaluated by a neurologist, endocrinologist, otorhinolaryngologist, and psychologist prior to his neuro-ophthalmology referral. He was admitted to the hospital twice: once for evaluation of headache and once after the episode of losing consciousness.

Laboratory testing during his illness included normal complete blood count, basic metabolic panel, liver studies, B12, folate, urinalysis, fasting lipid panel, and iron studies. Lead and arsenic were undetectable. Thyroid studies were normal. Testosterone was 132 (normal 241–827). Lyme and rapid plasma reagin (RPR) testing were negative. Lumbar puncture 1 month after the start of headaches did not provide any relief. Cerebrospinal fluid (CSF) examination revealed 0 white blood cells(wbc)/μL, 40 red blood cells(rbc)/μL, 76 mg/dL protein, 64 mg/dL glucose, negative venereal disease research laboratory, no cryptococcal antigen, and no malignant cells. Electroencephalogram (EEG) performed after his transient loss of consciousness did not reveal epileptiform activity. Echocardiogram was unremarkable.

Magnetic resonance imaging (MRI) of the brain two weeks after the onset of headaches demonstrated an opacified left frontal sinus and no parenchymal abnormalities. Repeat MRI of the

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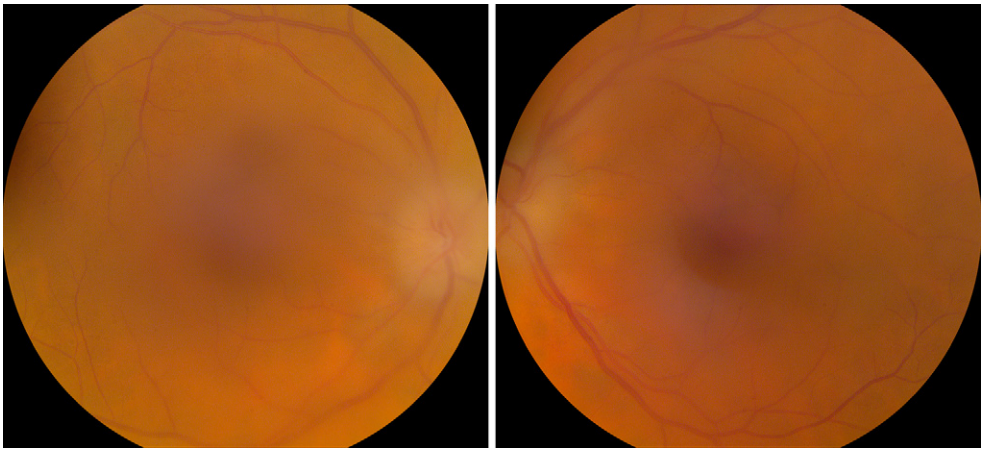

Fig. 1. Bilateral optic disk swelling; the vitreous haze accounts for the poor view.

brain three months into his symptoms was unchanged. Computed tomography (CT) angiogram of the head was unremarkable 3 months after the onset of headaches. Chest X-ray showed no acute disease and was stable when compared with a study from 6 years prior.

He was started on testosterone gel for low testosterone 2 months into his illness without any improvement. Therapeutic trials for his headaches and psychiatric symptoms included antibiotics, nonsteroidal anti-inflammatory agents, metoclopromide, topiramate, valproic acid, methylprednisolone, oxycodone, clonazepam, and escitalopram. None provided significant symptom relief.

He had no known drug allergies. Medications included levothyroxine, testosterone gel, fluticasone/salmeterol inhaler, fluticasone nasal spray, montelukast, clonazepam, escitalopram, multivitamins, folic acid, B-vitamin complex, fish oil, and ibuprofen as needed. Past medical history included a benign spinal cord tumor that was removed in June 2003, hypothyroidism, and sinus surgery.

He was married. He was self-employed in sales until 3 months into his illness when he became unable to work due to his cognitive impairments. He had a 25-pack an year smoking history. He quit smoking 14 years prior to presentation. He denied illicit drug use. Family history was remarkable for cardiac disease in both parents who died at 78 and 86 years of age and an aunt with epilepsy.

On examination he was normotensive weighing 228 pounds. Visual acuity was 20/80 −1 OD improving to 20/60 −1 with pinhole and 20/50 −2 OS improving to 20/30 −2 with pinhole. He saw only 2 of 8 Ishihara color plates with the right eye, and 6 of 8 with the left. Visual fields to confrontation, pupils, eyelids, and eye movements were all normal. Slit-lamp examination showed no cells in the anterior chamber. He had bilateral optic disk swelling and bilateral vitreous haze due to vitritis (Fig. 1).

On neurological examination he was agitated, with decreased attention, poor effort, and labile affect. He was not oriented to city or year, he did not know the president, and he could not recall three objects after 5 minutes. Spontaneous speech was perseverative and contained many curses. He could not follow embedded commands. Naming and repetition were intact. He had normal strength, sensation, coordination, gait, and deep tendon reflexes.

What are the diagnostic possibilities?

How would you proceed?

Comments

COMMENTS BY JOSEP DALMAU, MD, PHD

This is the case of a middle-aged man with subacute cognitive decline, visual changes, and loss of conciousness starting after a febrile illness. Based on this history a central nervous system (CNS) process with brain parenchymal and/or leptomeningeal involvement causing functional impairment and possibly seizures is suggested. A negative work-up at this juncture should not distract from this localization. A normal interictal EEG does not rule out seizures unless an event is captured and parenchymal inflammation causing clinical neurological compromise may be below the resolution of MRI and CSF sampling, particularly early in the presentation of many leptomeningeal processes.

Visual acuity changes and color vision loss suggest an optic nerve process with or without involvement of the retina; identification of disk swelling confirmed involvement of the optic nerve. The vitritis implies an inflammatory, infectious, or neoplastic etiology. The