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Clinical Challenge

Don't drink in the valley

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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.)

1. Case report

A 39-year-old previously healthy man was referred for evaluation of several months of headaches and intermittent blurred vision. There were no other neurological symptoms. At an emergency room visit 4 months previously, a complete blood count and basic metabolic profile were within normal limits, and noncontrast computed tomography (CT) of the head was interpreted as normal (Figs. 1 and 2). His headaches improved partially with acetaminophen.

Visual acuity was 20/32 OD and 20/25 OS. Pupillary reactions were brisk, with no anisocoria and no relative afferent pupillary defect. The anterior segment examination was unremarkable, and extraocular motility and confrontational visual field testing were normal. He correctly identified 8 of 10 Ishihara pseudoisochromatic color plates in each eye. Funduscopy showed optic disk edema which was mild (Frisén grade 2) in each eye with superior and nasal peripapillary hard exudates. There were no cells in the vitreous. The macula demonstrated hard exudates. The retinal periphery was normal. Blood pressure was 124/78 mm Hg.

Are the ophthalmic findings consistent with optic disk edema from papilledema?

Is a noncontrast head CT adequate for this patient?

What would you do next?

2. Comments

2.1. *Comments by Michael Vaphiades, DO*

The most common causes of bilateral disk edema with relatively good visual acuities, no relative afferent pupillary defect, and preserved visual fields are papilledema (from a varied etiologies), uncontrolled hypertension (hypertensive retinopathy), and diabetic papillopathy. This patient is not a diabetic that we know of, and presumably uncontrolled hypertension would have been detected in the emergency department. So the most logical explanation at this point is papilledema from elevated intracranial pressure which would be in-keeping with the headaches and transient visual obscurations (intermittent blurred vision). This is presumably not from a large space occupying lesion because that likely would have been detected on the noncontrasted cranial CT. The noncontrast CT is not an adequate imaging modality to detect some other processes causing elevated intracranial pressure like inflammatory, infectious or some neoplastic disorders. He is atypical for idiopathic intracranial hypertension given his gender, and we are not told his weight. We need more history (substance abuse, sexual history, travel) and should obtain a contrast-enhanced magnetic resonance imaging brain and orbits study with fat suppression and an MRV

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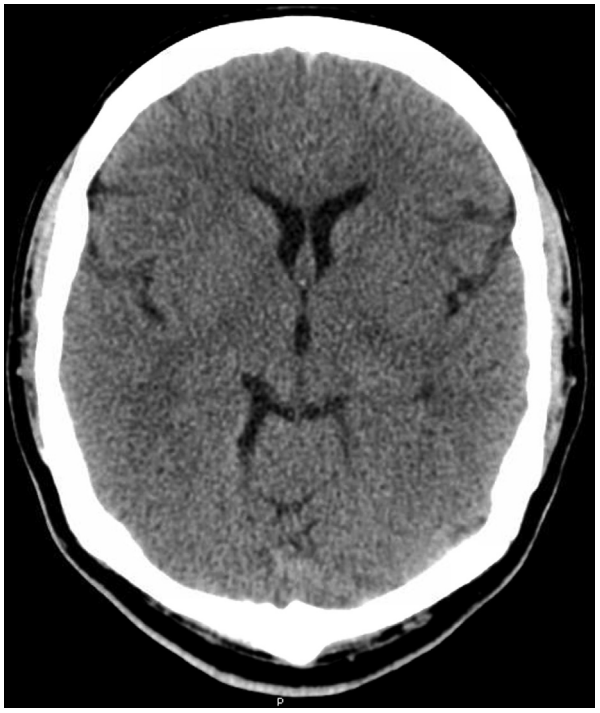

Fig. 1 – Noncontrast axial CT of the brain showed no mass lesion. CT, computed tomography.

to look for cerebral venous sinus thrombosis. I would also check a complete blood count to look for anemia, which can be associated with elevated intracranial pressure, and a basic metabolic panel, including a glucose, in case we need to treat with corticosteroids or other drugs. In addition, I would check for inflammatory disorders like sarcoidosis, lupus or infections like HIV, tuberculosis, or syphilis. The caveat is that other bilateral optic neuropathies can be confused with papilledema, another reason for better neuroimaging.

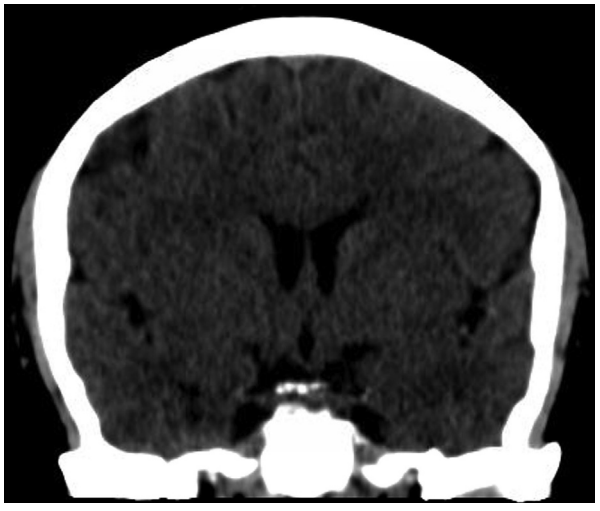

Fig. 2 – Noncontrast coronal CT of the brain showed no mass lesion compressing the optic chiasm. CT, computed tomography.

3. Case report (continued)

On repeated questioning, he reported a 10-pound weight loss and night sweats over the past year. He also reported a history of 21 years of heavy alcohol use but had quit 6 months before the onset of his headaches. Magnetic resonance imaging of the brain with contrast demonstrated extensive nodular leptomeningeal enhancement, greatest in the basilar cisterns, with extension into the foramen magnum as well as right inferior frontal and bilateral optic nerve and chiasmal enhancement with surrounding edema (Figs. 3 and 4). There was mild enlargement of the lateral and third ventricles (not depicted in the figures).

What would you do next?

4. Comments (continued)

4.1. *Comments by Dr. Vaphiades*

We need to evaluate the patient's cerebrospinal fluid (CSF) to search for infections (bacterial and fungal), inflammation, or neoplastic disorders (especially with the weight loss). We want to send a high volume of CSF for polymerase chain reaction to help rule out neoplasm. In addition, body CT or positron emission tomography scanning may help to define another area of pathology or primary neoplasm that could potentially be biopsied. In addition, I would prescribe acetazolamide to lower the intracranial pressure. Alternatively, if a ventriculoperitoneal shunt (VP) shunt was placed, a meningeal biopsy could be performed at that time for diagnostic purposes, and this would obviate the acetazolamide.

5. Case report (concluded)

After the optic disk edema was noted, the patient was sent to the emergency room where he rapidly decompensated. Given the examination findings of bilateral optic disk edema and imaging studies consistent with meningitis and communicating hydrocephalus, he was referred to neurosurgery and had a ventriculoperitoneal shunt placed and right frontal dural and leptomeningeal biopsy performed. The CSF studies at the time of shunt placement showed no pleocytosis, and all serologic testing for fungal, bacterial, syphilitic, viral, and mycobacterial etiologies was negative. Meningeal biopsy demonstrated acute and chronic inflammation, but no organisms or neoplasm. The patient had a lumbar puncture for further laboratory evaluation and was started on fluconazole 400 mg daily because of concerns for fungal infection. Although peripheral coccidioides antibodies were negative, the CSF demonstrated positive titers for coccidioides on a repeat lumbar puncture.

Three months later, vision was 20/20 in each eye with a few residual peripapillary hard exudates and no evidence of optic disk edema. His headaches had resolved, and he was continued on fluconazole.