CLINICAL VIGNETTE

Idiopathic Intracranial Hypertension: A Case Report

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Case Study

A 42-year-old female had been having nearly daily headaches for several months. She described the headaches as throbbing and associated with nausea and flashing lights in her peripheral visual fields. The headaches were effectively treated with ibuprofen, caffeine and rest. Her other presenting symptom was mild dyspepsia, worse with recumbency. The headaches would occur in the morning and would intensify with change in position until she received her daily dose of ibuprofen. One month prior to presentation, the patient began having blurred vision. She was unable to associate the visual changes with the headache, as both happened on a daily basis. The visual changes were described as vision blurring lasting several minutes, with spontaneous resolution. As the patient worked for hours at a time in front of a computer monitor, she attributed the symptoms to eye fatigue and visited her optometrist to evaluate her prescription. After being told she had papilledema, she presented for further evaluation.

She had a past medical history of obesity, hypertension, polycystic ovarian syndrome, and migraine. Her family history was significant for migraine and diabetes in her mother and thyroid disease in one sister. There was no personal or family history of cancer, heart disease, or neurologic disease. Her medications included a multivitamin and metformin 500mg once daily. She smoked a half pack of cigarettes daily for the past 22 years. She drank alcohol periodically and did not use intravenous drugs. Her review of systems was negative other than the stated symptoms.

On exam, the patient was an overweight female (BMI 32), with hypertension (BP 145/89). She had a non-focal neurologic exam. Her cranial nerve exam was normal. She had no deficits in balance testing and a Mini Mental examination revealed a score of 27/30. She had moderate exopthalmos, which had been observed on previous exams. The patient's fundoscopic exam revealed papilledema with flattened optic discs bilaterally.

The patient subsequently was evaluated by neurology and underwent imaging with an MRI of the brain. This revealed an empty sella turcica without other abnormalities. Lumbar puncture was remarkable for an opening pressure of 280 mmH2O. Ophthalmologic exam revealed peripheral visual field deficits without significant diminishment in acuity.

Discussion

Idiopathic Intracranial Hypertension (IIH) should be considered in the setting of intractable headaches with visual disturbance. The overall incidence of IIH is 1.5/100,000 with the highest risk group being overweight women aged 15 to 45[1]. As was the case in this presentation, the most typical symptom is headache with transient visual symptoms. Other symptoms that suggest IIH are diplopia, tinnitus, photopsia and neck pain. The headaches are typically severe and, because of the associated visual symptoms, nausea and vomiting, are often confused with migraines. Pulsatile tinnitus is present in the majority of cases, with patients typically describing an amplified transmitted heartbeat. The visual symptoms are typically transient loss of acuity, lasting seconds to minutes. Additionally, visual scotoma with peripheral flashes of light and peripheral visual losses are reported in more than half of IIH cases. Diplopia and sustained loss of visual acuity are less common, occurring in roughly 30 percent of cases[2].

On examination, the critical finding is papilledema. Comprehensive ophthalmologic examination may reveal visual field losses, afferent pupillary defect, and retinal edema or detachment. Cranial nerve deficits may also be observed, particularly a sixth nerve palsy.

Once a diagnosis of IIH is considered, imaging should be obtained to exclude other causes of elevated intracranial hypertension, such as malignancy, subarachnoid hemorrhage, or venous sinus thrombosis. MRI will reveal such structural lesions. If negative, a lumbar puncture with opening pressure readings should be performed. The diagnosis is confirmed with an elevated pressure (over 200mmH2O) with an otherwise normal CSF profile[3].

The prognosis associated with IIH is reflective of the major morbidity of the disease, visual loss. Rates of blindness associated with the disease vary among studies, ranging from 6-24%[4].

Treatment approaches to IIH center on the goal of reducing vision loss. Studies of weight loss in obese patients with IIH have demonstrated a reduction in both papilledema and visual decline. Typically, weight loss is prescribed in conjunction with medical therapies. These treatments aim to lower the intracranial pressure, thereby reducing compression of the optic nerve. The first line treatment is typically acetazolamide 500mg twice a day[5]. This dose stabilized vision loss in over half the treated patients. Furosemide is often used as an adjunct to acetazolamide when single drug therapy has been unsuccessful at alleviating visual symptoms. In patients whose visual symptoms fail to improve on medical therapy, two surgical options are available. CSF shunting is effective in relieving headache and has a high rate of visual stabilization or improvement (up to 95%). Optic nerve sheath fenestration is equally effective in treating visual symptoms, but has less effect on headache than shunting. Both these surgical procedures have a high degree of complication[6].

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