CHAPTER 33

THE WOMAN WITH GENERALIZED HEADACHE AND TRANSIENT VISUAL OBSCURATIONS

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Case History

A 42-year-old obese woman developed, over the last year, a constant holocranial headache associated with intermittent nausea, pulsatile tinnitus, and transient visual obscurations (TVOs). She had been consuming acetylsalicylic acid on a daily basis for her headaches. Her menses were often irregular. She had a prior history of migraine without aura which was usually triggered by menstruation and relieved with each of her pregnancies. A neurologic examination was normal except for the presence of bilateral papilledema.

Questions about This Case

- What is your diagnosis and differential diagnosis?
- What diagnostic procedures would you perform and why?
- How would you manage the patient's headaches?
- What is the major morbidity associated with this disorder?

Case Discussion

This 42-year-old woman with a prior history of migraine developed a chronic daily headache associated with papilledema due to intracranial hypertension. A mass lesion must be ruled out by neuroimaging, preferably magnetic resonance imaging (MRI). The MRI was done and was normal except for an empty sella, which is often associated with increased intracranial pressure. A magnetic resonance venogram showed no evidence of venous obstruction. A lumbar puncture (LP) was next performed. This is obligatory as chronic meningitis can mimic the syndrome of idiopathic intracranial hypertension (IIH). The opening pressure was 300 mm of cerebrospinal fluid (CSF). The cell count, protein, and glucose were normal. Fungal cultures and cryptoccocal antigens were negative. The patient had no acquired immunodeficiency syndrome (AIDS) risk factors and no evidence of hypoparathyroidism or systemic lupus erythematosus (SLE), and she did not take any of the drugs associated with increased intracranial pressure.

Intracranial hypertension may be either: (1) idiopathic, with no clear identifiable cause, or (2) symptomatic, as a result of systemic lupus, renal disease, hypoparathyroidism, venous sinus occlusion, radical neck dissection, vitamin A intoxication, or drug side effects (nalidixic acid, danocrine, steroid withdrawal) (Table 33–1). Symptomatic intracranial hypertension can be secondary to changes in cranial venous outflow, which may influence intracranial pressure by increasing cerebral blood

TABLE 33–1. Syndromes of Increased Intracranial Pressure

Primary
A. Idiopathic intracranial hypertension with papilledema
B. Idiopathic intracranial hypertension without papilledema
Secondary
A. Hydrocephalus
B. Mass lesion, neoplasm, stroke—hematoma
C. Meningitis/encephalitis
D. Trauma
E. Major intracranial and extracranial venous obstruction
F. Drugs: vitamin A, nalidixic acid, anabolic steroids, steroid withdrawal
G. Systemic disease: renal disease, hypoparathyroidism, SLE

TABLE 33–2. Features of Idiopathic Intracranial Hypertension

1. Headache: chronic tension-type headache with migrainous features, may be present upon awakening; Can be intermittent or absent
2. Associated features: pulsatile tinnitus, transient visual obscurations, diplopia, visual loss, shoulder and arm pain
3. Patients: predominantly obese women aged 20 to 50
4. Physical and neurologic examination: within normal limits except for papilledema, visual loss, obesity, and a sixth nerve palsy (occasionally bilateral)
5. Neuroradiology: CT or MRI show no evidence of intracranial mass, hydrocephalus, or venous sinus thrombosis. (Empty sella may be present.)
6. Lumbar puncture: demonstrates increased CSF pressure with a normal composition. (May show decreased protein.)
7. No other causes of increased CSF pressure present.

volume, producing brain edema, and impairing CSF absorption. Intracranial venous outflow obstruction can be caused by tumors, head trauma, chronic otitis, and hypercoagulable states.

Idiopathic intracranial hypertension (pseudotumor cerebri) is a disorder of increased intracranial pressure of unknown cause which occurs predominantly in obese women of childbearing age. Its major morbidity, visual loss, occurs in 80% of patients; blindness occurs in 10%. Headache occurs in most, but not all, patients. Transient visual obscuration, an episode of visual clouding in one or both eyes, usually lasting seconds, is common and occurred in this patient. Transient visual obscuration is seen in all forms of increased intracranial pressure with papilledema but is not a specific symptom. Transient visual obscuration can occur even in patients without increased intracranial pressure who have elevated optic disks (not papilledema) from other causes (drusen, coloboma, disk edema, and optic nerve sheath tumors). Other common symptoms of IIH include diplopia, visual loss, and pulsatile tinnitus. Some patients have shoulder and arm pain (perhaps secondary to nerve root dilation) and retro-orbital pain. Signs include papilledema and sixth nerve palsy.

Idiopathic intracranial hypertension occurs with a frequency of about 1 case per 100,000 per year, rising to 19.3 cases in obese women between the ages of 20 and 44. The patient with IIH is commonly a young, obese woman with chronic daily headache (CDH), normal laboratory studies, normal neurologic examination (except for papilledema), and empty sella (Table 33–2). Idiopathic intracranial hypertension is not associated with pregnancy, hypertension, diabetes, thyroid disease, iron deficiency anemia, or the use of tetracycline or oral contraceptives.

The pathophysiology of IIH is unknown. Postulated mechanisms include: (1) increased rate of CSF formation; (2) increased intracranial venous pressure; (3) decreased rate of CSF absorption; and (4) increased interstitial fluid in the brain (edema). Recent studies suggest that a decreased rate of CSF absorption at the arachnoid villi and interstitial brain edema are the major contributors.

Most patients describe their headache as their most severe ever. Common features include daily headache present upon awakening and pulsating in character, retro-ocular pain with eye movement, and nausea, vomiting, and pulsatile tinnitus.

In one series of patients, the mean age was 31 years and headache was reported by 92% (73% had CDH, 93% said it was the most severe ever, and 83% said it was pulsatile). Nausea occurred in 57%, vomiting in 38%, and orbital pain in 43%. Transient visual obscuration was present in 71%, diplopia in 38%, and visual loss in 31%.

Occasionally patients with IIH are incidentally found to have papilledema while being examined for another purpose. Five to 10% of patients are essentially asymptomatic. Loss of visual field and visual acuity are the only significant complications of IIH with papilledema.

An ophthalmologic examination should include intraocular pressure, visual fields (Goldmann or Humphrey), optic disk photos, visual acuity, and search for a relative afferent pupil.

In some patients, intracranial hypertension without papilledema has been described. Patients, particularly obese women, with CDH and symptoms of increased intracranial pressure, i.e., pulsatile tinnitus, history of head trauma or meningitis, an empty sella on neuroimaging studies, or a headache that is not relieved by standard therapy, should have a diagnostic LP. The clinical, historic, radiographic, and demographic characteristics are identical to those of patients with papilledema except for: (1) possible association with prior head trauma or meningitis; (2) extended delay in diagnosis, which requires an LP in the absence of papilledema; and (3) no evidence of the visual loss seen in patients with IIH.

The reason there is no papilledema in these cases of intracranial hypertension is not known. Congenital or acquired optic nerve sheath defects, "chronic IIH"with resolution of papilledema, or early IIH are alternative explanations.

In patients with suspected intracranial hypertension (with or without papilledema), the diagnosis is based on an LP following neuroimaging. If the LP is unremarkable and intracranial pressure is elevated to greater than 200 mm of water (in nonobese subjects), then IIH is the likely diagnosis. Routine blood chemistries, prothrombin time (PT), partial thromboplastin time (PTT), antinuclear antibodies (ANA), venereal disease research laboratory (VDRL), chemistry profile, thyroxine (T_4), and thyroid-stimulating hormone (TSH) are helpful.

TABLE 33–2. Treatment of Idiopathic Intracranial Hypertension

1. Eliminate symptomatic causes
2. Weight loss, if obese
3. Standard headache treatment
4. Carbonic anhydrase inhibitors and loop diuretics
5. Short course of high-dose corticosteroids
6. Serial lumbar punctures
7. Lumboperitoneal or ventriculoperitoneal shunt
8. Optic nerve sheath fenestration

Management Strategies

Once the diagnosis of IIH is made, secondary causes should be sought and eliminated. Over 50 diseases, conditions, toxins, or pharmaceuticals have been associated with IIH. Obese patients should be encouraged to lose weight. If the patient is asymptomatic and has no visual loss, then no treatment is indicated. Careful ophthalmologic follow-up is needed. If there is no papilledema, or papilledema with no visual loss, and the only complaint is headache, then the headache should be treated aggressively. Headache associated with IIH and papilledema has been reported to frequently respond to standard headache treatment (Table 33–3).

If rigorous headache therapy is unsuccessful, or if there is visual loss, then a 4- to 6-week trial of furosemide or a potent carbonic anhydrase inhibitor (acetazolamide) should be given. These drugs decrease elevated intracranial pressure. The use of high-dose steroids (prednisone or dexamethasone) is controversial but may be effective in IIH. Headache commonly recurs when steroids are withdrawn.

Lumbar puncture typically relieves headache in IIH. Since CSF is rapidly replaced, prolonged symptomatic relief may reflect a persistent CSF leak. Alternatively, transient reduction of CSF pressure may allow decompression of the arachnoid villi, allowing for prolonged enhanced CSF absorption. Patients with IIH who have visual loss or severe incapacitating headache that does not respond to medical therapy or repeated LP may need surgical management.

Some suggest treating IIH with a lumboperitoneal shunt, but this often has to be repeated and has the potential for development of hindbrain herniation and a new headache. For others a ventriculoperitoneal shunt is the preferred shunting procedure.

Optic nerve sheath fenestration (ONSF) entails surgical incision of the dura mater covering the intraorbital optic nerve. The proposed mechanism is improved optic nerve axoplasmic flow and continuous intraorbital CSF drainage. Sixty-five to 76% of patients get relief of medically uncontrolled headache with ONSF. Although ONSF has been performed on patients with unilateral papilledema, to our knowledge it is untried in patients with IIH without papilledema. Without the threat of loss of vision, the small risk of visual loss due to the surgery probably outweighs the potential benefits.

Selected Readings

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Editorial Comments

The finding of papilledema in a patient with headache usually suggests an ominous etiology; however, when initial imaging procedures are normal, the diagnosis of "idiopathic intracranial hypertension" or what used to be called "pseudotumor cerebri" is entertained. In its idiopathic form this disorder almost approaches a syndromic status. Dr. Silberstein clearly overviews this disorder in this case, along with differential diagnostic concerns and management strategies. It is wise to exclude "secondary," treatable etiologies in all cases. As outlined by Dr. Silberstein, careful attention to visual symptoms and their progression is a high priority in such cases, as other symptoms frequently abate and resolve themselves over time.