Benign Intracranial Hypertension

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Benign intracranial hypertension (pseudotumor cerebri) is a syndrome of intracranial hypertension that classically presents with headaches and visual disturbance. Physical examination discloses papilledema. Diagnosis is confirmed by a normal cranial computed tomographic scan or magnetic resonance image and the presence of a markedly increased opening pressure on lumbar puncture. Treatment is directed to underlying causes, hypertension, and withdrawal of offending medications. Repeated lumbar puncture, diuretic therapy, and surgery are occasionally used. Careful follow-up and visual testing are imperative. **J Fam Pract 1990; 30:290-292**

Benign intracranial hypertension (pseudotumor cerebri) is a syndrome of intracranial hypertension without associated intracranial mass. Classically, signs of the condition include headaches, visual disturbance, and a finding of papilledema. It is important for the family physician to include this disorder in the differential diagnosis of headache, especially in obese women of childbearing years.

CASE REPORT

A 22-year-old obese woman came to the Family Health Center with the complaint of headaches for 6 years. Usually her headache would last for 3 to 4 days and spontaneously resolve. The patient had occasionally experienced scotomata preceding the headaches but denied photophobia, nausea, or vomiting. She had been treated with a variety of nonsteroidal anti-inflammatory agents without benefit. During the 3 weeks before admission, the headaches became increasingly severe and were associated with mild photophobia. She was also nauseated and vomited several times. There were no localizing neurologic symptoms. The patient denied any fever, chills, confusion, or seizure activity.

On physical examination, the temperature was found to be 37.0°C (98.6°F) orally, blood pressure 100/76 mm Hg, pulse 112 beats per minute. The patient was an obese woman in some distress because of her severe headache. On head, ear, eye, nose, and throat examination, pupils were found to be equal, round, and reactive to light. Accommodation and extraocular movements were intact. The discs showed bilateral papilledema. The neck was slightly rigid. The neurologic examination was otherwise unremarkable. Cranial nerves II through XII were intact. Motor examination revealed that her muscle strength was 5/5, and her deep tendon reflexes were 2+/4+. Sensation was intact to light touch. There were no cerebellar signs. The complete blood count and electrolytes were normal.

The patient underwent an emergency cranial computed tomographic (CT) scan, with normal results except for a small ventricular system and no clearly demonstrated sulci space. These findings were compatible with increased intracranial pressure. The differential diagnosis entertained at that time included infection and pseudotumor cerebri. A lumbar puncture was performed, which demonstrated clear fluid and a pressure exceeding the maximum top of the manometer, ie, greater than 56 cm of water. Latex agglutination tests on the cerebrospinal fluid for streptococcal pneumonia, *Neisseria meningitides*, and *Cryptococcus* were negative. Acid-fast baccilus staining revealed no organisms. The microscopic examination was remarkable for several leukocytes. There was ultimately no growth either for acid-fast bacilli or fungal organisms.

Neurology consultation was obtained, and the presumptive diagnosis of pseudotumor cerebri was made. Findings from magnetic resonance imaging (MRI) of the head, obtained to exclude a venous sinus thrombosis, were completely normal. A vitamin A level, thyroid index, parathyroid hormone level, and cortisol determination were also obtained and all were normal.

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A therapeutic lumbar puncture was performed with enough fluid withdrawn to lower closing pressure to 16 mm of water. The patient was begun on furosemide, 40 mg twice daily, and acetazolamide, 250 mg 4 times daily. A dietary consultation was obtained for a weight-reducing diet, and an ophthalmology consultation was obtained to evaluate the patient's visual fields. The patient was discharged on the third day of hospitalization on a regimen of sustained-release acetazolamide, 500 mg twice a day, furosemide, 40 mg orally twice a day, and a 1800-cal weight-reduction diet.

Since the time of her discharge, the patient has had several repeat lumbar punctures. Prednisone was added to the regimen for a 2-week trial at 40 mg twice daily. Despite medical therapy, her headaches persisted, but her visual fields have remained intact by objective examination.

A surgery consultation was obtained. Because she had a good response to lumbar punctures, it was felt that a lumbar peritoneal shunt would be appropriate. The shunt was therefore placed at the spinal cord level of L4-L5. The surgery was tolerated well. At 1-month follow-up, the patient has continued to have incapacitating headaches and intermittent poor vision. While this poor response may indicate shunt failure, it is hoped the patient will improve over the next several months.

DISCUSSION

Quincke first described the syndrome of increased intracranial pressure in the absence of a space-occupying lesion in 1897.[1,2] "Ottic hydrocephalus" was further characterized in patients with thrombosis of the dural sinuses following ear infection.[2] *Benign intracranial hypertension* was a phrase coined by Foley[3] and is now used along with the term *pseudotumor cerebri* to describe a syndrome of increased intracranial pressure without the presence of an intracranial mass.

Benign intracranial hypertension usually presents with headache, ranging from minor to severe, which is typically worse with the Valsalva maneuver and head movement and is more pronounced in the morning.[4–8] Visual disturbance is seen in approximately 35% of patients and is generally preceded by headache. There may be episodes of horizontal double vision or other tangential visual obscurations.[6–9] Less frequently, there is pain in the neck, shoulders, back, arms, and occasionally, pulsatile tinnitus.[4] Notably there is no disruption of consciousness.

On physical examination, the most frequent deficit found is papilledema, which is generally bilateral but can occasionally be seen unilaterally.[7–9] Up to 25% of patients will have a sixth cranial nerve palsy related to their increased intracranial pressure.[4–7] On close visual testing, many patients will be found to have an enlarged blind spot and inferior defects in their nasal-visual field.[7–9]

TABLE 1. DIFFERENTIAL DIAGNOSIS OF BENIGN INTRACRANIAL HYPERTENSION

Tumors
Brain
Spinal cord
Metastatic
Arteriovenous malformations
Encephalitis
Sarcoidosis
Pseudopapilledema

Differential Diagnosis

When the patient presents with headaches and papilledema, a number of diagnostic entities should come to mind in addition to benign intracranial hypertension[5–7] (Table 1). To rule out an intracranial mass, cranial CT scanning or MRI should be undertaken. The findings on CT scan will be normal or occasionally show small ventricles in the case of pseudotumor cerebri.[10,11] Only if the CT or MRI scanning results are normal may a lumbar puncture be performed to confirm increased intracranial pressure. If a lumbar puncture is performed in the presence of a mass lesion with increased intracranial pressure, fatal complications may ensue. An opening pressure greater than 200 mm of water is diagnostic. Glucose and cell count in the cerebrospinal fluid will be normal while protein is normal or low.[5–7]

Causes of Benign Intracranial Hypertension

Benign intracranial hypertension is seen in approximately one out of 100,000 patients.[12] The diagnosis is made most frequently in obese women of childbearing age. Once the diagnosis of benign intracranial hypertension has been made, a variety of causes should be ruled out[5–7] (Table 2). A thorough history of medication use, nutritional status, personal history, gynecologic problems, and evidence for selected autoimmune and endocrine disorders should be sought.[5–7]

Treatment

Initial treatment is aimed at correcting underlying causes of benign intracranial hypertension. Withdrawal of offending medications, weight reduction, and the treatment

TABLE 2. CONDITIONS IMPLICATED IN CAUSING BENIGN INTRACRANIAL HYPERTENSION

Metabolic or Endocrine	**Venous Sinus Thrombosis**
Obesity	Supprative otitis media
Pregnancy	Head trauma
Pituitary tumor	Jugular vein occlusion
Addison's disease	
Menstrual irregularity	
Hypoparathyroidism	
Thyroid abnormalities	
Medications	**Other**
Steroids	Systemic lupus erythematosus
Oral contraceptive pills and estrogens	Guillain-Barré syndrome
Thyroid replacement	Iron deficiency anemia
Tetracyclines	
Nalidixic acid	
Vitamin A	
Lithium	

of hypertension are important.[5–7] Because 10% to 24% of patients with benign intracranial hypertension suffer serious visual obscurations—and impairment does not correlate with the duration of symptoms, degree of papilledema, or number of occurrences—static perimetry is essential for all patients.[7] The loss is often seen in peripheral fields initially; therefore, Snellen testing or routine visual field screening is insufficiently sensitive to detect such a loss. After the initial screening, the asymptomatic person should have a follow-up eye examination in 1 to 3 months. Acetazolamide in doses of 500 mg twice a day to three times a day, or 40- to 160-mg doses of furosemide may be helpful for symptoms. Potassium supplements are usually needed in conjunction with these medications. In addition, numbness, renal stones, and mild metabolic acidosis are associated with acetazolamide. Steroids, when used for a short time, may be helpful, but recurrent headache and visual disturbance are generally seen after withdrawal. Indeed, steroids when used on a prolonged basis, have been associated with the benign intracranial hypertension syndrome.

Lumbar puncture has been widely used for the treatment of headache. It is recommended that approximately 30 cm of fluid be removed or that the opening pressure be reduced to 50% of the initial reading. While there is no correlation between the intracranial pressure and the occurrence of headache, some patients appear to benefit consistently from this technique. For patients with progressive visual loss or headache unrelieved with any of the above therapies or more traditional analgesics, the lumbar peritoneal shunt can be helpful.

Lumbar peritoneal shunting effectively treats benign intracranial hypertension, and symptoms and signs will generally respond within 1 month of the operation.[13,14] In a recent series of patients with recalcitrant benign intracranial hypertension, all responded favorably to lumbar peritoneal shunt.[14] Nonetheless, over 50% required shunt revision, and there is concern that shunting will prolong what is usually a self-limited process. Thus, the lumbar peritoneal shunt is a treatment of last resort, reserved for patients with unremitting symptoms or rapid visual loss.

Long-term follow-up of patients with benign intracranial hypertension is essential. Visual loss has been reported years after initial presentation.[7,8] While most patients will have a course lasting 6 to 12 months, there is recurrence in 10%, and up to 25% may have a protracted course.[15]

It is important for the family physician to recognize benign intracranial hypertension as an entity in the differential diagnosis of headache. A thorough examination of the optic disc will provide the key to initial diagnosis, which can be confirmed by the absence of a mass on CT scanning and the presence of markedly increased intracranial pressure on lumbar puncture. Screening for visual losses and long-term follow up are essential.

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