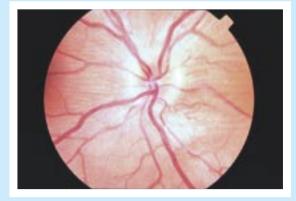
CLINICAL CHALLENGE & DÉFI CLINIQUE

Ophthaproblem

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Left eye

28-year-old obese white woman presents to Ayour office with a 3-month history of increasingly diffuse headaches associated with bilateral transient blurred vision and occasional "swooshing" sounds in her right ear. She is sometimes nauseated but does not vomit. She denies having paresthesias or weakness. She has mild photophobia but no phonophobia. She does not complain of scintillating scotomas and has no family history of migraine headaches. Her temperature is 37.4°C and her blood tests are normal.

Her medical history includes low-grade chronic sinusitis for which she took oral antibiotics 5 months ago. Her physical examination reveals no focal neurologic signs. Fundoscopic examination shows that her best-corrected visual acuity is decreased at 20/50 in both eyes.

Which of the following interventions is most appropriate for this patient?

- 1. An immediate lumbar puncture because meningitis is most likely given the history of sinusitis
- 2. Referral to an optometrist for correction of vision because a refractive error is the cause of her symptoms
- 3. Recommending weight loss and diuretics
- 4. A computed tomography scan of the head to rule out an intracranial mass.
- 5. Both 3 and 4

Answer on page 592

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Answer to Ophthaproblem

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5. Both 3 and 4

This patient has chronic papilledema L secondary to pseudotumour cerebri (PTC) (benign or idiopathic intracranial hypertension). Her presentation is typical for this condition. Lack of focal neurologic signs makes an intracranial mass unlikely. An infectious cause is also unlikely because her temperature and blood tests are normal.

Pseudotumour cerebri is a condition of unknown cause characterized by increased cerebrospinal fluid (CSF) pressure and papilledema.^{1,2} Incidence of this condition is at least 1/100 000 in the general population and increases to 19/100 000 among obese women of child-bearing age.¹

Pseudotumour cerebri presents with symptoms and signs of increased intracranial pressure (ICP) and no other focal neurologic abnormalities.3 Affected patients usually complain of severe headache, nausea, vomiting, transient visual obscurations (dimming of vision), neck or shoulder pain, and pulsatile tinnitus.¹⁻³ Horizontal diplopia might also be a symptom because increased ICP causes sixth-nerve palsy.^{1,3} The classic finding of papilledema occurs in more than 90% of cases and is usually bilateral and occasionally unilateral.^{1,3} Visual acuity is usually normal, but visual fields sometimes show an enlarged blind spot.^{1,4}

The exact cause of PTC is unknown, but it has been associated with endocrine or metabolic dysfunction, pregnancy, vitamin A use, tetracycline, nalidixic acid, cyclosporine, steroid withdrawal, and oral contraceptives. 1,3 Incidence of PTC peaks in the third decade of life and occurs more often in women than men. Obesity is commonly associated with it.1,2





Left eye

Diagnosis

Pseudotumour cerebri is essentially a diagnosis of exclusion.1 To diagnose PTC, physicians must first rule out an intracranial mass. Neuroimaging (computed tomography or magnetic resonance imaging) can rule out an intracranial mass, hydrocephalus, meningeal lesion, or cerebral venous lesion.^{1,3} Neuroimaging should be followed by a lumbar puncture to confirm increased ICP and to rule out meningitis by examining the composition of the CSF.^{1,3} In patients with PTC, CSF composition is usually normal, although the protein level is occasionally low. In summary, the four main criteria for diagnosing PTC are papilledema, normal results of neuroimaging, increased ICP, and normal CSF composition.⁵

Management

Family physicians should manage this condition in conjunction with an ophthalmologist, neurologist, or neuroophthalmologist. Treatment, which is both medical and surgical, is aimed at lowering ICP and treating symptoms directly (ie, headache). 1,3 The mainstay of medical management includes weight loss and diuretics (acetazolamide or furosemide).1 Steroids might decrease elevated ICP, but use of corticosteroids is controversial because steroid withdrawal

itself is also a well-known cause of PTC.3 Some neurologists have used repeated lumbar punctures to decrease ICP, but effects are short-lived and the procedure can be painful.1

Surgical management primarily involves CSF shunting (lumboperitoneal or ventriculoperitoneal) or decompression of the optic nerve sheath. This maneuver is usually reserved for patients with intractable headaches or progressive visual loss despite maximal medical treatment.³

The prognosis of PTC is highly variable. Most cases last several years; some patients have short-lived episodes that resolve within months. The recurrence rate is approximately 10%, and symptoms can reappear weeks to years from initial presentation.¹

References

- 1. Corbett JJ. Idiopathic intracranial hypertension (pseudotumor cerebri). In: Albert DM, Jakobiec PA, editors. Principles and practice of ophthalmology. Vol 4. Philadelphia, Pa: WB Saunders Co; 1994. Chap. 214, p. 2698-706.
- 2. Friedman DI. Pseudotumor cerebri. Neurosurg Clin North Am 1999:4:609-21.
- 3. Newman SA, et al. Pseudotumor cerebri. In: Neuroophthalmology. American Academy of Ophthalmology basic and clinical science course. 2001-2002 ed. San Francisco, Calif: The Foundation of the American Academy of Ophthalmology; 2001. Section 5, p. 136-8.
- 4. Wall M, George D. Idiopathic intracranial hypertension. A prospective study of 50 patients. Brain 1991;114:155-80.
- 5. Kline LB, Bajandas FJ. The swollen optic disc. In: Neuroophthalmology, American Academy of Ophthalmology basic and clinical science course. 5th ed. Thorofare, NJ: Slack Inc; 2001. Chap 9, p. 149-60.