Answer to Ophthaproblem *continued from* ***page 901***

3. Idiopathic intracranial hypertension

Idiopathic intracranial hypertension (IIH), also known as *pseudotumour cerebri* or *benign intracranial hypertension,* is a neurologic disorder characterized by elevated intracranial pressure (ICP) in the absence of any apparent cause. Although more common in women of childbearing age, it can affect men and women of all ages, including children. The highest incidence is among obese women between the ages of 20 and 44 years.[1]

The pathophysiology of ICP elevation in IIH is poorly understood, in large part owing to the difficulty of applying animal models to human physiology. In general, IIH is thought to result from a multifactorial process affecting the balance between cerebrospinal fluid (CSF) production and absorption; various theories exist, including intracranial venous hypertension, vitamin A toxicity, venous sinus stenosis, and metabolic or neuroendocrine causes.[2] Obese individuals might be at greater risk due to increased intra-abdominal pressure, which impairs venous return, leading to cerebral venous hypertension.[3] Idiopathic intracranial hypertension has also been associated with various systemic conditions and certain medications (eg, tetracycline antibiotics and corticosteroids).[2]

The clinical manifestations of IIH are related to increased ICP, with headaches being the most common presenting symptom.[4] Headaches in IIH can vary in type, but are typically described as diffuse and throbbing, worse upon wakening, and accompanied by photophobia and phonophobia. Visual symptoms in the form of "transient visual obscurations"[5] are also common, occurring in approximately 75% of cases. Transient visual obscurations are brief episodes (30 to 60 seconds) of unilateral or bilateral visual dimming thought to result from optic nerve ischemia.[5] Other possible symptoms include "pulse-synchronous tinnitus,"[6] horizontal diplopia (from abducent nerve palsy), and nonspecific symptoms such as nausea, vomiting, and dizziness.[6] In some cases, the condition is asymptomatic.

The hallmark sign of IIH is bilateral papilledema. With proper training and practice, primary care physicians can observe papilledema with a direct ophthalmoscope, even through an undilated pupil (although pupil dilation with a short-acting mydriatic eye drop is accepted practice in family medicine clinics). The disk might appear hyperemic with blurred disk margins, loss of the physiologic cup, and occasional optic disk hemorrhage. It should be noted that several conditions, including drusen of the optic nerve head, myelinated nerve fibres, and other forms of optic neuropathy, might mimic the appearance of papilledema.

Management

From a primary care standpoint, the key to managing IIH is early recognition and prompt referral or investigation. Any suspicion of IIH or papilledema warrants urgent referral to an ophthalmologist or neurologist, as well as neuroimaging in the form of magnetic resonance imaging or contrast-enhanced computed tomography to exclude a space-occupying lesion. Although often normal in appearance, neuroimaging of patients with IIH might occasionally show slit-like ventricles or an "empty sella sign" (caused by flattening of the pituitary).[7] Once a space-occupying lesion has been ruled out, diagnosis of IIH can be confirmed by performing a lumbar puncture, which would demonstrate an elevated opening pressure but normal CSF composition.[8] Of note, IIH is the only instance in which lumbar puncture is not contraindicated in a patient suspected of having high ICP (other contraindications, such as coagulopathy, thrombocytopenia, and infection at puncture site, would still apply).

The treatment goals with IIH are primarily to relieve headache and prevent vision loss. Management is multidisciplinary and often involves an ophthalmologist, neurologist, and neurosurgeon in addition to the primary care physician. Although some patients can be managed conservatively with weight loss, a low-sodium diet, and close observation, many require treatment to lower ICP. This can be done medicinally in the form of acetazolamide or surgically in the form of CSF shunting or optic nerve sheath fenestration.[9] Morbidly obese patients resistant to medical and surgical treatment should be considered for bariatric surgery.[10]

Recommendations

Prompt recognition of papilledema or IIH is important to prevent permanent vision loss. Primary care physicians trained in ophthalmoscopy are able to directly identify papilledema. Any suspicion of papilledema warrants referral and neuroimaging.

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Competing interests
None declared

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