

Pseudotumor Cerebri Syndrome: IIH and cerebral venous sinus disease

Neil R. Miller, MD FACS

Pseudotumor cerebri (PTC) is the term used for a clinical condition presenting with signs and symptoms of elevated intracranial pressure (ICP), such as headache, papilledema, pulsatile tinnitus, transient visual loss, VI nerve palsy, progressive visual field deficits, torticollis (in children) and cerebrospinal fluid (CSF) leakage. The current criteria for diagnosis of PTC require: (1) intracranial pressure elevation to at least 250 mm H₂O; (2) normal-sized or small ventricles; (3) absence of intracranial mass lesions; and (4) normal CSF composition. Although some cases of PTC are due to specific causes, such as drugs (eg, lithium, tetracycline), and are called “secondary PTC”, most occur in obese or overweight young women for reasons that are unclear and are referred to as “primary PTC” or “idiopathic intracranial hypertension” (IIH). It is now clear that most patients with so-called IIH have evidence of unilateral or bilateral transverse sinus stenosis. The role that this plays in the etiology of IIH is unclear and probably heterogeneous; however, stenting of the stenotic region has become a popular treatment, particularly in patients who fail medical therapy.

Appropriate and timely management of IIH usually prevents irreversible loss of vision and often can restore visual function that has been lost. In all cases, once the diagnosis has been made by neuroimaging and lumbar puncture, medical therapy (combined with weight loss in obese patients with IIH) is often sufficient to cure the condition, whereas surgery should be offered to those who fail, cannot tolerate, or decline medical treatment or those who already have significant visual dysfunction when first diagnosed. Surgical procedures currently being used include optic nerve sheath fenestration, various forms of cerebrospinal fluid diversion procedures, and venous sinus stenting. There are advantages and disadvantages to all of these procedures. Long-term monitoring with funduscopy, optical coherence tomography, and visual function examination is crucial while a patient is on medical management and after surgical treatment.

References

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