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Intracranial Hypertension (Pseudotumor Cerebri)

Idiopathic intracranial hypertension (IIH) in post-pubescent teenagers and adults is a disorder causing elevated intracranial pressure which occurs typically in young obese women. The etiology of this disorder is unclear, although it may be related to abnormal CSF absorption and abnormal venous drainage.

Clinical symptoms include headaches, transient visual obscurations defined as dimming of vision lasting seconds with standing up and Valsalva maneuvers, pulsatile tinnitus that goes with the heartbeat, diplopia from pseudosixth nerve palsy, and vision loss which usually affects the peripheral vision first and infrequently causes loss of acuity.

Triggers for IIH include recent weight gain, use of vitamin A products including isotretinoin, tetracyclines, and withdrawal from chronic corticosteroid use.

Clinical exam shows bilateral optic disc edema which may be asymmetric usually with normal visual acuity and visual fields which can range from normal to severe defects.

Differential diagnosis of optic disc edema includes papilledema from intracranial mass, hydrocephalus, meningitis. Other differential diagnoses include optic disc drusen, anomalous optic disc, anterior ischemic optic neuropathy, and anterior optic neuritis.

The modified Dandy criteria for IIH includes an opening pressure >25 mm $\rm H_2O$, normal constituents in the spinal fluid without evidence of infection, MRI or CT brain without mass or hydrocephalus, and no other cause elevated intracranial pressure. If the case is atypical, a MR or CT venogram should be performed to exclude venous sinus thrombosis.

Management is based on visual function (visual acuity and Humphrey visual field) and severity of papilledema. Weight loss is recommended for all as a 5% loss has been shown to improve IIH. Patients with normal visual function and mild papilledema may not require any treatment with medications. With moderately decreased visual function, acetazolamide is used with dosing

from 1000-4000 mg daily. For patients with severely decreased or worsening visual function despite medical treatment, interventions such as optic nerve sheath fenestration, lumbar or ventriculoperitoneal shunt, or venous sinus stenting may be required. Repeat lumbar punctures are not helpful for the routine treatment of IIH since the CSF which is drained off is quickly remade.

The prognosis is typically good. Usually IIH resolves with treatment but can be recurrent or chronic. Neuro-ophthalmologists provide an important service to monitor the visual function and prevent irreversible vision loss.

All patients with headache require an initial examination of the optic disc to look for possible papilledema.

By: Thong Pham, MD

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