Case 9485

Idiopathic intracranial hypertension
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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Case Type: Clinical Cases
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Patient: 24 years, female

Clinical History:
A 24-year-old lady presented with gradually progressive vision loss and history of unrelenting headaches. Clinical examination revealed mild bilateral papilloedema. Neurological examination was non-contributory.

Imaging Findings:
MR imaging of brain revealed essentially normal-appearing supratentorial and infratentorial structures. No intracranial space-occupying lesion was seen. Note was made of partial empty-sella turcica as evidenced by partial flattening of the pituitary gland along the floor of sella. MR venogram was within normal limits. Additionally present were fluid-distended bilateral optic nerve sheaths with tortuous and buckled bilateral optic nerves. Mild flattening of the bilateral posterior globes was seen in the region of optic disc.

Based on these clinico-radiological findings the diagnostic possibility of idiopathic intracranial hypertension was suggested which was confirmed on lumbar puncture. The patient was subsequently initiated on Diamox and has been doing well.

Discussion:
Idiopathic intracranial hypertension (IIH), previously referred to as benign intracranial hypertension or pseudotumor cerebri, is a neurological disorder characterised by increased intracranial pressure in the absence of an intracranial tumour or other pathologies such as hydrocephalus or cerebral venous sinus thrombosis. Although classically described in obese young female patients, IIH can be seen in both genders and all age groups. Chief clinical symptoms include headache, nausea, vomiting, pulsatile tinnitus, double vision and gradual vision loss. Headache is frequently the main presenting complaint and is typically intractable and disabling. It is characteristically worse in the morning, generalised in character and throbbing in nature. If untreated, IIH is a serious threat which can cause permanent vision loss; hence, early recognition is vital, as timely intervention can preserve vision.

IIH is usually a diagnosis of exclusion [3]. It has been conventionally diagnosed by measuring an increased CSF pressure at lumbar puncture in patients with characteristic clinical features. Lumbar puncture not only measures the opening pressure, but also provides cerebrospinal fluid (CSF) for examination to exclude alternative diagnoses. If the opening pressure is increased, CSF can also be removed for symptomatic relief. Neuroimaging is performed to confirm the diagnosis, as well as to exclude alternative causes, such as intracranial mass lesion or hydrocephalus. MR venography should always be performed to exclude the possibility of cerebral venous sinus thrombosis or venous obstruction. Imaging findings suggesting the possibility of IIH include: empty sella turcica (flattening of the pituitary gland due to increased pressure), small or slit-like ventricles, optic nerve sheath distension, optic nerve tortuosity, and posterior globe flattening [1, 4, 2]. Although individual MRI findings lack sensitivity in diagnosis of IIH, a combination of signs in appropriate clinical context are very helpful. As per one of the studies [2], posterior globe
flattening is the only sign that, if present, strongly suggests the diagnosis of IIH (specificity 100%, 95% CI 93.6% to 100%; sensitivity 43.5%, 95% CI 27.3% to 60.8%; positive likelihood ratio 49.7).

The treatment of IIH is basically aimed at alleviating symptoms and preserving vision. Medical management is limited to the use of diuretics particularly acetazolamide and encouragement of weight loss [5]. Acetazolamide (Diamox) acts by inhibiting the enzyme carbonic anhydrase thus reducing CSF production. Surgical intervention such as CSF diversion procedures and optic nerve fenestration are opted only in cases of failed medical management.

**Differential Diagnosis List:** Idiopathic intracranial hypertension, Intracranial tumour, Cerebral venous thrombosis

**Final Diagnosis:** Idiopathic intracranial hypertension

**References:**


Description: Bilateral optic nerve sheaths are fluid-distended and are causing mild posterior globe flattening. The optic nerves display an unusual tortuous course. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
Description: Bilateral optic nerves are encircled by prominent CSF-distended optic nerve sheaths suggesting an abnormally elevated intracranial pressure. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
Description: Bilateral optic nerve sheaths are distended. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
Description: Distension of the optic nerve sheaths represent consequence of long-term increased pressure within CSF spaces. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
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Description: Note is made of partial empty-sella turcica as evidenced by partial flattening of the pituitary gland along the floor of sella. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
Figure 5

Description: Normal intracranial MR venogram. Origin: Ankur Arora, Dept of Radiodiagnosis, ILBS Hospital, New Delhi, India.
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