Headache and Visual Symptoms in a Child

Tabarki. B [1,2]

[1] Headache and Visual Symptoms in a Child
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Clinical Presentation :

An 11-year-old previously healthy female presented with a two-week history of progressively worsening headaches associated with transient episodes of blurred vision and pulsatile tinnitus. The headaches were diffuse, throbbing, and more pronounced in the morning, occasionally waking her from sleep. There was no history of fever, vomiting, recent infection, head trauma, or medication use.
On examination, it was noted that she was overweight (BMI > 95th percentile for age) and alert. Additionally, she displayed mild right sixth-nerve palsy. The rest of the neurological and systemic examinations were unremarkable. Funduscopic examination and a brain MRI were conducted, as shown in Figures 1a, 1b, and 2.

Figure 1 : (a) Axial T2-weighted brain MRI showing optic-nerve tortuosity and bilateral papillary protrusion (arrow). (b) Sagittal T1-weighted image demonstrating a partially empty sella (circle) and low-lying cerebellar tonsils (line).

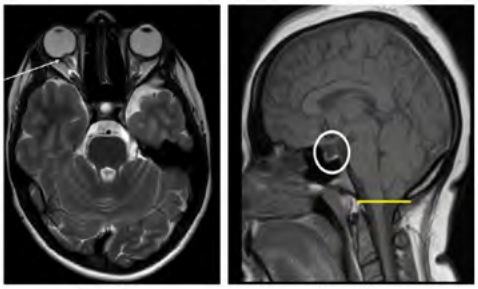

Figure 1a Figure 1b

Figure 2 : Fundus photograph showing bilateral papilledema (Grade 3).

Figure 2

Questions :

1. What are the findings inferred from Figures 1a, 1b, and 2?
2. What is the most likely diagnosis?
3. How is the diagnosis confirmed?
4. What is the treatment?

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ANSWERS AND DISCUSSION :

1. Brain MRI reveals optic-nerve tortuosity, bilateral papillary protrusion, an empty sella, and low-lying cerebellar tonsils (Figure 1a and 1b).

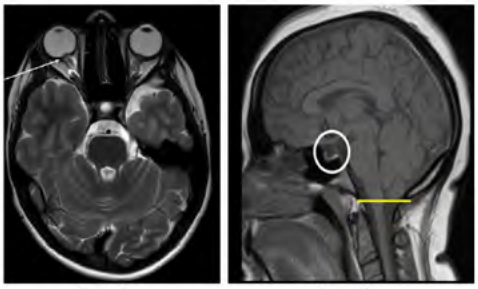

Figure 1a Figure 1b

Magnetic resonance venography (MRV, not shown) was normal, showing no signs of venous sinus thrombosis or stenosis. Figure 2 shows bilateral papilledema at grade 3.

Figure 2

2. Based on the clinical presentation, funduscopic findings, and neuroimaging, the most likely diagnosis is idiopathic intracranial hypertension (IIH). The primary risk factor identified was being overweight.
3. Confirmation of IIH requires measuring the cerebrospinal fluid (CSF) opening pressure. A lumbar puncture revealed an elevated opening pressure of 30 cm H_2O (normal < 25 cm H_2O) with normal CSF composition.
4. The main treatment for IIH are acetazolamide therapy and weight reduction. Acetazolamide inhibits carbonic anhydrase, thereby reducing CSF production. The recommended dose for children is 25 mg/kg/day (up to 2 g/day) divided into two or three doses. It is important to monitor and correct serum electrolytes and blood gas, as necessary. Furosemide or topiramate may be used as adjunctive agents to further lower intracranial pressure. Most pediatric cases respond well to first-line medical therapy (1,2). If IIH is refractory to medical treatment, surgical options, such as optic-nerve-sheath fenestration, CSF shunting, serial lumbar punctures, or venous-sinus stenting; may be considered (1-3).
Pediatric IIH is rare but clinically significant, with an estimated incidence of approximately 1 case per 100,000 children. The incidence increases during adolescence, particularly between 12 and 15 years, with a female predominance (1,2).
According to the revised criteria (4,5), the diagnosis of IIH requires:
1.Papilledema is present in 90 to 95 % of patients, either unilaterally or bilaterally. However, even in its absence, the diagnosis should not be ruled out if clinical and CSF findings support raised intracranial pressure. Optical coherence tomography is a useful tool to assess the thickness of the retinal nerve fiber layer surrounding the optic disk. Both visual field and visual acuity should be assessed in these cases.
2.Elevated intracranial pressure (> 25 cm H_2O in non-obese, unsedated children, or > 28 cm H_2O in obese or sedated children). The procedure must be performed in the lateral decubitus position for accurate pressure measurement;
3.Normal neurological examination except for cranial-nerve palsies; 4. Normal CSF composition;
5.Normal neuroimaging with no secondary cause of intracranial hypertension.
The most concerning complication of IIH is irreversible visual impairment, which may occur in up to 10% of children (2). Regular ophthalmologic monitoring is essential to prevent permanent visual loss.

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