

Headache in an Obese Adolescent Male: A Nonclassical Presentation of an Uncommon Disease

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Abstract

Background. Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a diagnosis of exclusion in the clinical scenario of increased intracranial pressure without an identifiable cause. This case report provides a brief review of current literature on IIH, important differential considerations to rule out prior to diagnosis, and relevant imaging findings of IIH. **Case Presentation.** An 18-year-old male presenting with headache and blurry vision was found to have signs of increased intracranial pressure on imaging without other abnormalities to explain the cause, suggesting IIH as a diagnosis. **Conclusion.** IIH is classically seen in overweight females of childbearing age but should be considered in all obese patients, including the pediatric population given the increasing rate of childhood obesity.

Keywords

idiopathic intracranial hypertension, pseudotumor cerebri, headache, obesity

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Case Report

An obese 18-year-old male presents to the emergency department with a 2-day history of recurrent headaches. His headaches are located in the occipital region with a dull, aching quality. Patient also endorses nausea, vomiting, and vision changes during his headaches. He denies recent head trauma, loss of consciousness, or history of migraine headaches. He has been taking ibuprofen for his pain without much relief. Neurologic examination was negative with intact cranial nerves II–XII, normal sensory and motor function in all extremities, and negative cerebellar examination. A computed tomography (CT) scan of the brain was negative for masses, bleed, midline shift, and hydrocephalus but demonstrated an empty sella (Figure 1) and indentation of the posterior sclera of both globes at the optic nerve head insertion sites (Figure 2a and b), both of which are radiographic indicators of increased intracranial pressure (ICP). A follow-up magnetic resonance imaging (MRI) scan of the brain revealed enhancement and protrusion of the prelaminar optic nerves bilaterally and tortuosity of the right optic nerve (Figure 3), which are consistent with prolonged elevation of ICP. Magnetic resonance venogram (MRV) of the head demonstrated a right transverse sinus “flow gap,” a common finding in



Figure 1. Sagittal noncontrast head computed tomography shows an empty sella, a sign of increased intracranial pressure.

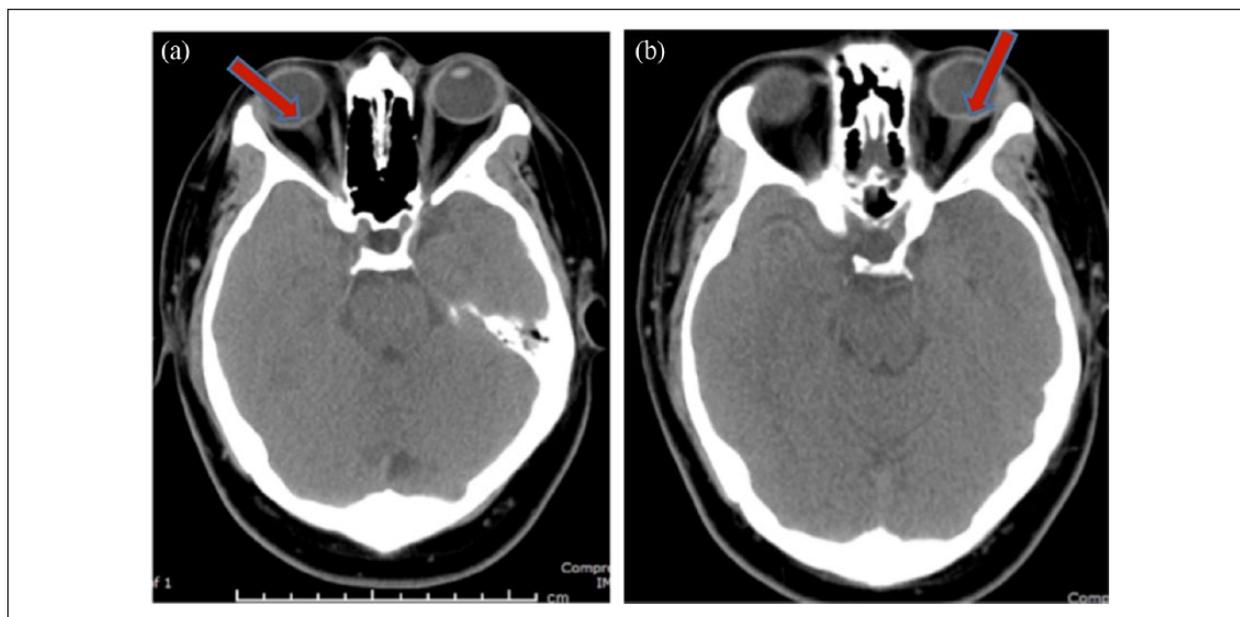
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Figures 2. Axial noncontrast head CT shows indentation of the posterior sclera of the right (a) and left (b) globes, respectively, a sign of increased intracranial pressure.

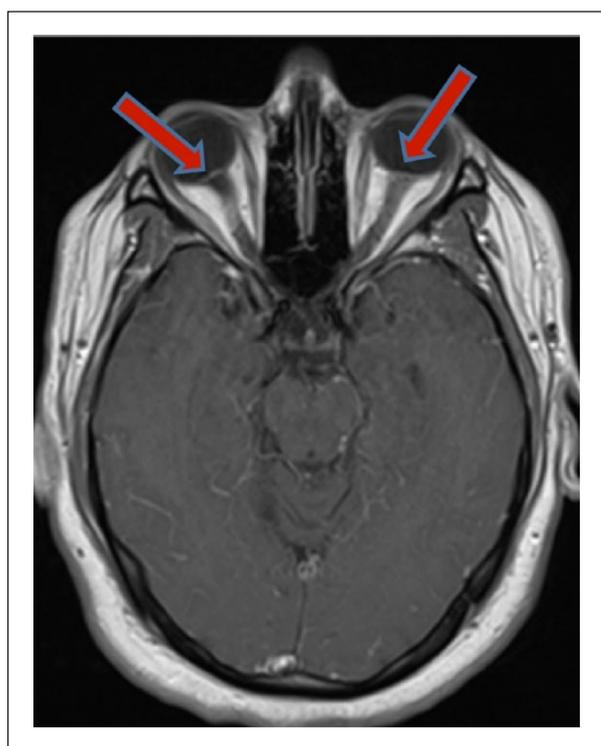


Figure 3. Contrast enhanced T1-weighted axial magnetic resonance imaging of the brain shows enhancement and protrusion of the prelaminar optic nerves and right optic nerve tortuosity, signs of increased intracranial pressure.

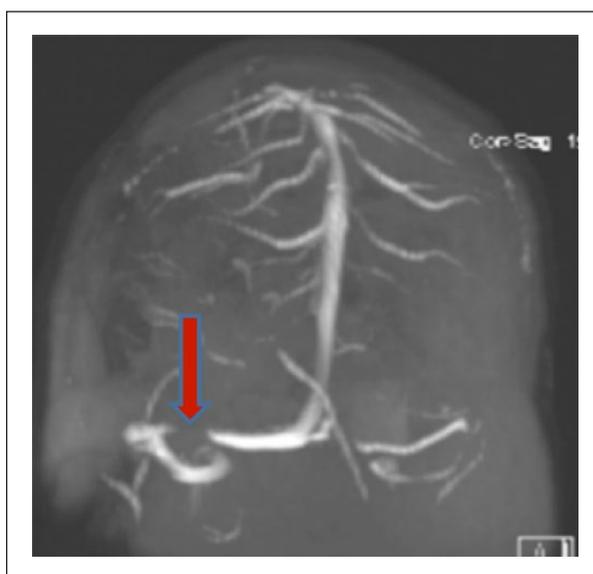


Figure 4. Time of flight magnetic resonance venogram of the head shows a right transverse sinus "flow gap" and an asymmetrically narrowed caliber left transverse sinus. There is no evidence of venous sinus thrombosis.

patients with IIH, but no evidence of venous sinus thrombosis (Figure 4). As a result of these imaging, the patient underwent a lumbar puncture, which demonstrated an opening pressure of 55 cm H₂O (normal

range = 11.5-27 cm H₂O in children¹) and normal cerebral spinal fluid analysis. He was started on acetazolamide and counseled on weight loss.

Final Diagnosis

Idiopathic intracranial hypertension.

Discussion

IIH, commonly called pseudotumor cerebri, is diagnosed by an elevation in ICP without an identifiable cause. While many hypotheses have been given for the increase in ICP, from increased cerebral volume to increased cerebrospinal fluid production, the exact pathogenesis remains unknown. Despite its former name benign intracranial hypertension, untreated IIH can have severe consequences. IIH causes optic nerve damage through papilledema, with studies demonstrating visual field loss in 71% of patients at time of diagnosis.² Thus, prompt treatment is required to prevent further vision loss.

IIH is an uncommon disease with an annual incidence of 1 to 2 per 100 000, but rates have been rising globally with the obesity epidemic.³ The classic IIH patient is an overweight female of childbearing age, though IIH has been seen in children as young as 4 months old.⁴ In the pediatric population in particular, obesity is a less important risk factor compared with the adult population, and there is no distinct sex predilection.⁵ Furthermore, a limited study of 10 prepubertal children with IIH reported strabismus and stiff neck as more common presenting symptoms than headache.⁶

When symptoms of increased ICP present in these nonclassical populations, it is especially important to evaluate for secondary causes of intracranial hypertension with neuroimaging prior to making a diagnosis of IIH. The differential diagnosis is broad and includes intracranial hemorrhage, central nervous system infection, stroke, neoplasm, hydrocephalus, cerebral edema, and obstructed venous drainage. In the pediatric population in particular, clinicians should assess for steroid use or withdrawal, tetracycline use, Addison's disease, hypoparathyroidism, obstructive sleep apnea, and lupus, all of which can cause elevated ICP.

In cases of acute elevation of ICP, head CT is the imaging of choice to evaluate for midline shift, effacement of the basilar cisterns and sulci, and hydrocephalus.⁷ Otherwise, MRI of the brain with contrast is the preferred study to exclude secondary intracranial hypertension, particularly subtle intracranial masses and meningeal pathologies.⁸ Image findings on MRI suggestive

of elevated ICP include empty sella, flattening of the posterior sclera, tortuosity of the orbital optic nerve, and distention of the preoptic subarachnoid space.⁹ MRV is often required in addition to MRI to exclude conditions that cause intracranial hypertension via venous outflow obstruction, such as dural venous sinus thrombosis.¹⁰ Flow gaps, seen in this patient, commonly present in IIH as an indication of transverse sinus stenosis and not thrombosis; whether their formation is the cause or consequence of elevated ICP still remains a hotly debated topic.¹¹

In this case report, the positive findings of empty sella, flattening of the posterior sclera, and optic nerve tortuosity on CT/MR, exclusion of dural venous sinus thrombosis on MRV, and increased opening pressure on lumbar puncture all together cinch the diagnosis of IIH.

Conclusion

Idiopathic intracranial hypertension is an uncommon disease that presents atypically in the pediatric population, usually without the common risk factors seen in the adult population. When suspected, other causes of intracranial hypertension need to be evaluated with neuroimaging prior to making this diagnosis of exclusion. Treatment should be initiated promptly to prevent further vision loss.

Author Contributions

BZ: Contributed to conception and design; drafted the manuscript; critically revised the manuscript; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

CY: Contributed to conception and design; contributed to analysis; drafted the manuscript; critically revised the manuscript; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

SC: Contributed to conception and design; contributed to analysis; critically revised the manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring integrity and accuracy.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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