MASKED IDIOPATHIC INTRACRANIAL HYPERTENSION

Antoine Saint-Victor[1], Lauren Zingaro[1], Zainab Chaudhary[1], Anna-lena Meinhardt MD, Melissa Wing, Kevin Mesina MD, Esther Exantus DPM, Htay Htay Kyi MD

Department of Internal Medicine, Rutgers New Jersey Medical School

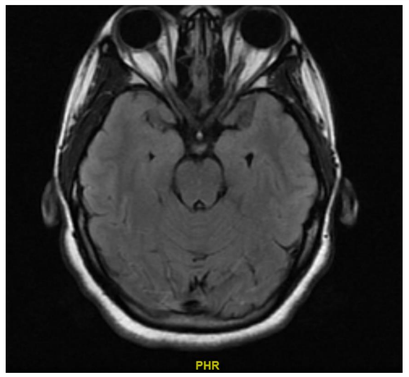
Introduction

- Idiopathic Intracranial Hypertension (IIH) refers to a buildup of cerebrospinal fluid in the skull, causing increased intracranial pressure and resulting in symptoms like headache, and visual and auditory changes. The presentation and differential diagnoses of IIH are broad, leading to an extensive work up, and ultimately the diagnosis is one of exclusion.
- Typical presentation for patients with IIH is a young obese female with history of headache, papilledema, diplopia, and pulsatile tinnitus (1). The presence of papilledema further supports the diagnosis of IIH. If papilledema is present, neuroimaging and a lumbar puncture is performed to confirm the absence of structural issues and increased ICP, respectively (2). When these parameters are met and no other diagnosis fits better, the diagnosis of IIH is made.

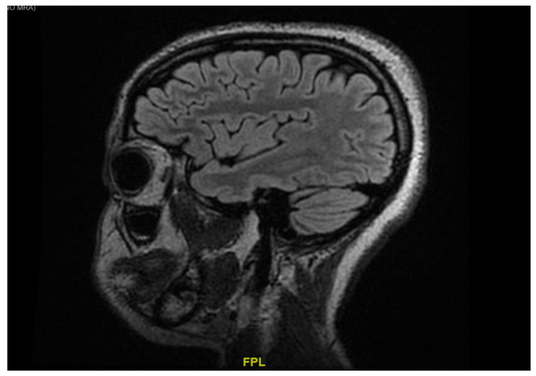
Case Presentation

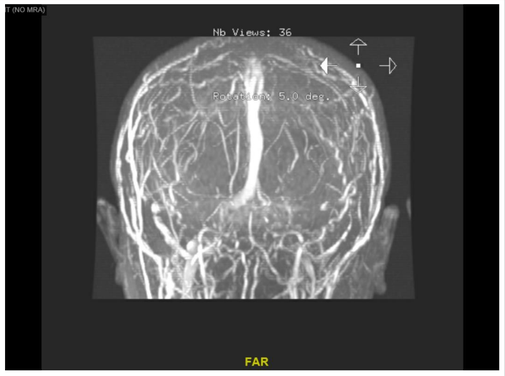
- A 17-year-old female with a past medical history significant for asthma and subclinical hypothyroidism presented with a chief complaint of persistent, progressive blurry vision for 3 weeks.
- She also had intermittent neck and shoulder pain along with mild weakness of the right upper extremity, as well as vibration and proprioception in the bilateral lower extremities. Workup during an admission 4 weeks prior for a similar complaint showed normal results. At that time, she was discharged on steroids and tizanidine.
- She then presented to the ophthalmology clinic, where a fundoscopic exam revealed bilateral papilledema. CT imaging also showed bilateral papilledema without hemorrhage, mass effect, or enhancing mass lesions.
- On admission, vitals were stable apart from tachycardia, and, notably, her BMI was 24.1.
- Rheumatology workup was negative aside from speckled ANA pattern of 1:1280, positive rheumatoid factor 1:80, and ESR of 23. Infectious workup including HIV, syphilis and a hepatitis panel were normal.
- MRI of the orbit showed bilateral papilledema with no hemorrhagic or enhancing lesions. A lumbar puncture was performed with findings only significant for an opening pressure of >55 cm H2O.
- Due to increasing pressure, she received an optic nerve sheath fenestration of her left eye. She was started on high-dose intravenous solumedrol IV as well as acetazolamide.
- A diagnostic cerebral angiogram and venous manometry showed unremarkable arterial vasculature but revealed a pressure gradient between the bilateral transverse and sigmoid sinuses, suggesting a stenosis. Subsequently a stent was placed in the right transverse sinus.
- In the following four months after her surgery, the patient continues to be followed by the outpatient neurosurgery and ophthalmology clinics. The vision in her right eye has improved while the vision in her left eye has stabilized. Her peripheral vision is slowly improving. For maintenance, she is taking acetazolamide and aspirin, and has been discontinued on clopidogrel and steroids. The patient had a follow-up MRI of her cervical spine which was unremarkable, and there are no noted neurological deficits.

Significant Findings



MRI of orbit showing bilateral papilledema with no hemorrhagic or enhancing lesions





MRI Brain venogram showing suggested stenoses at the distal transverse sinuses bilaterally. No evidence of dural venous sinus thrombosis. Protrusion of the optic nerves to the posterior globes, and relatively diminutive pituitary gland for the patient's age and gender, findings which are compatible with increased intracranial pressure.

Conclusion

- Due to the nature of IIH being a diagnosis of exclusion, it can take many laboratory and imaging studies to come to a definitive clinical conclusion.
- While fitting some of the typical criteria, studies have shown that adolescents with IIH are more commonly associated with higher BMIs than the patient.
- Additionally, the patient's history of hypothyroidism, elevated ANA titers, and moderately elevated myelin protein made it difficult to diagnose because together they inferred an autoimmune etiology such as Multiple Sclerosis. The atypical neurologic symptoms of the extremities in addition to the expected findings such as headaches, diplopia, rapidly progressive vision loss, and pulsating tinnitus further confounded the work up.
- Initially, medication alone was able to provide relief for the patient and decreased her blurry vision. However, within days the patient's vision continued to deteriorate despite optic nerve fenestration and CSF drainage via lumbar puncture.
- Current data shows that transverse sinus stenting has proved efficacious in reducing CSF pressure and resolving patient symptoms when IIH is refractory to first-line treatments (5). Ultimately, stent placement resolved the patient's visual symptoms and after extensive work up the etiology of her increased ICP is determined to be IIH.
- More common etiologies of acute vision changes and papilledema must be ruled out before making the diagnosis of IIH, but it should always be included in the differential. IIH has overlapping features with well-known autoimmune diseases and mass effect, so adverse anatomical changes of the brain and opening CSF pressure must be evaluated early on as to prevent irreversible vision loss and nerve damage

References

- Ahmed RM, Wilkinson M, Parker GD, et al. Transverse sinus stenting for idiopathic intracranial hypertension: a review of 52 patients and of model predictions. AJNR Am J Neuroradiol. 2011;32(8):1408-1414. doi:10.3174/ajnr.A2575
- Ahmad SR, Moss HE. Update on the Diagnosis and Treatment of Idiopathic Intracranial Hypertension. Semin Neurol. 2019;39(6):682-691. doi:10.1055/s-0039-1698744
- Balcer LJ, Liu GT, Forman S, et al. Idiopathic intracranial hypertension: relation of age and obesity in children. Neurology. 1999;52(4):870-872. doi:10.1212/wnl.52.4.870
- Chen BS, Newman NJ, Biousse V. Atypical presentations of idiopathic intracranial hypertension. Taiwan J Ophthalmol. 2020;11(1):25-38. Published 2020 Dec 2. doi:10.4103/tjo.tjo_69_20
- Riggeal BD, Bruce BB, Saindane AM, et al. Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis. Neurology. 2013;80(3):289-295. doi:10.1212/WNL.0b013e31827debd6
- Sheldon CA, Paley GL, Beres SJ, McCormack SE, Liu GT. Pediatric Pseudotumor Cerebri Syndrome: Diagnosis, Classification, and Underlying Pathophysiology. Semin Pediatr Neurol. 2017;24(2):110-115. doi:10.1016/j.spen.2017.04.002