Masked Idiopathic Intracranial Hypertension

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Background: 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.

Case description: 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 impaired vibration and proprioception in the bilateral lower extremities. CT imaging also showed bilateral papilledema and vitals were stable apart from tachycardia. Notably, her BMI was 24.1. The workup is complicated by a history of autoimmune disorder and elevated ANA. 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. 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. She was started on high-dose intravenous solumedrol IV as well as acetazolamide and a stent was placed in the right transverse sinus.

Conclusion: 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.