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TEACHING FILES (GRAND ROUNDS)

FROM HEADACHE TO IIH: A PEDIATRIC CASE WITH AN UNEXPECTED TWIST

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Clinical Problem:
A previously healthy 11-year-old male from Venezuela presented to the emergency department with a two-week history of increasing headaches, nausea, vomiting, and new-onset visual disturbances, mimicking migraine headaches. Initially had a good response to ibuprofen but later worsened, accompanied by photophobia, blurry vision, and perception of "darkness" and "shadows." Diagnostic Workup Given worsening symptoms, the clinical team pursued imaging and laboratory workup to rule out intracranial pathology, including: CT Head without Contrast: Short segment of focal asymmetric expansion and hyperdensity of the middle portion of the superior sagittal sinus. Findings may represent intrinsic hyperdensity of the sinus secondary to elevated hematocrit, although dural venous sinus thrombosis would appear similar in the correct clinical setting. No areas of altered brain parenchymal density

CT Head with Contrast:

1. No CT venogram evidence of dural venous sinus thrombosis.
2. Incidentally noted, prominent bilateral optic nerve sheath complexes and optic discs. Correlate clinically for possible idiopathic intracranial hypertension with CSF opening pressure.
3. Partially visualized, moderate enlargement and striated enhancement of the nasopharyngeal soft tissues, likely secondary to an infectious/ inflammatory process.

MRI Brain with and without Contrast, MRI MRV Brain, MRI Orbit with and without Contrast:

1. Left frontal cerebral deep white matter small T2/ FLAIR hyperintense focus. Bilateral frontal and occipital periventricular cerebral white matter confluent subtle T2/FLAIR hyperintensities reflect gliosis, nonspecific. No focus on abnormal restricted diffusion or contrast enhancement within the brain to suggest an active demyelinating process.
2. Unremarkable brain MRV. No evidence of dural venous sinus thrombosis.
3. Redemonstrated prominent bilateral optic nerve sheath complexes with prominent subarachnoid spaces surrounding the optic nerves associated with mild enlargement and contrast enhancement of the optic discs, greater on the left. There is also partial empty sella. The constellation of imaging findings is concerning for idiopathic intracranial hypertension associated with bilateral papilledema vs bilateral optic papillitis.

Lumbar puncture: A lumbar puncture with manometry was done to directly measure the intracranial pressure. It revealed a significantly elevated opening pressure of 36+ cm H2O. The CSF analysis: cytology, cell count, and culture revealed no anomalies. There were no signs of infection or inflammation. A repeat LP showed improvement to 25+ cm H2O.

What is the differential diagnosis for pediatric patients presenting with headache and visual disturbances, and how can serious intracranial pathology be distinguished from benign conditions?

Discussion:

Headache is one of the most common complaints in pediatric patients, with a broad differential diagnosis.[1] While most pediatric headaches are benign and self-resolving, progressive or worsening headaches with associated neurological symptoms such as sudden vision changes, weakness, speech changes, or seizures require further investigation to rule out more serious intracranial pathologies.[1,3] This 11-year-old's presentation of progressive headache, nausea, vomiting, and visual disturbances with associated photophobia, initially raised suspicion for a migraine exacerbation, particularly given the family history of migraine. However, the failure to respond to initial treatment and the presence of new neurological symptoms warranted further investigation. The diagnosis of idiopathic intracranial hypertension (IIH) was ultimately confirmed with MRI imaging and Lumbar Puncture.

Idiopathic intracranial hypertension (IIH) typically presents in overweight females of childbearing age.[2] The condition is characterized by symptoms of increased intracranial pressure, including headache, often described as diffuse and pulsatile. Visual disturbances, such as blurred vision or transient visual obscurations, are common due to swelling of the optic

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disc. Patients may also experience nausea, vomiting, and tinnitus, with severe cases potentially leading to progressive vision loss if left untreated.

Although infections, such as viral or bacterial meningitis, are also common causes of headaches in children, the patient's negative CSF analysis and lack of fever made an infectious etiology less likely. Given the patient's history and the lack of response to antibiotics, the initial diagnosis of strep pharyngitis was unlikely to explain his persistent and worsening symptoms.

This case highlights the importance of considering serious intracranial pathology in pediatric patients presenting with progressive headaches, particularly when accompanied by worsening visual disturbances. In this case, the patient did not match the typical profile of IIH, however establishing a diagnosis hinged on delving into a further workup based on worsening symptoms. Early imaging and thorough evaluation are essential in ruling out life-threatening conditions such as brain tumors, even when the presentation initially suggests a more benign diagnosis like migraine. Clinicians must maintain a broad differential diagnosis and pursue appropriate diagnostic investigations when symptoms fail to improve with initial treatments.

Compliance with ethical standards

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Conflict of Interest: None

References:

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