CASE REPORT

Headaches That Ring a Bell's Palsy

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ABSTRACT

Idiopathic intracranial hypertension is a rare and potentially sight-threatening condition that occurs in both pediatric and adult populations. Children, particularly those who are prepubertal, are more likely to exhibit atypical presentations of IIH as compared to their adult counterparts. We present a case of an obese eight-year-old girl with chronic headaches and a strong family history of IIH who presented to an outside hospital with several months of persistent occipital headaches acutely complicated by a left-facial nerve palsy and left upper extremity weakness. Computed tomography, magnetic resonance imaging, and magnetic resonance venography revealed no indication of an intracranial abnormality. A fundoscopic exam performed by an ophthalmologist was negative for papilledema. However, a sedated lumbar puncture revealed a significantly elevated opening pressure of 31 cm H20, indicating a diagnosis of idiopathic intracranial hypertension. The patient was started on maintenance acetazolamide and continues to follow outpatient with pediatric neurology. Diagnosing idiopathic intracranial hypertension in children poses a unique challenge, given the variability in clinical presentation. This case highlights that clinicians must have a high index of suspicion for IIH despite the absence of papilledema in patients with acute atypical neurologic features such as unilateral facial nerve palsy and radiculopathy in patients with chronic headaches refractory to medication.

BACKGROUND

Idiopathic intracranial hypertension (IIH), or pseudotumor cerebri, is a condition of elevated intracranial pressure (ICP) without a brain mass or other inciting cause. Patients typically present with symptoms of elevated ICP, such as headache, nausea, vomiting, visual disturbances, and papilledema.[1] IIH is diagnosed with the Modified Dandy Criteria[1]: 1) symptoms of elevated ICP (e.g., headache, nausea, vision loss), 2) lack of other neurologic abnormalities except for abducens (sixth) nerve palsy, 3) CSF opening pressure >25 cm H2O with normal CSF composition, 4) neuroimaging without evidence of other etiology for intracranial hypertension, 5) no other etiology for intracranial hypertension.

However, Friedman et al. proposed updated diagnostic criteria for IIH that additionally account for elevated ICP in the absence of papilledema.[2] These criteria factors in additional findings such as the presence of an abducens nerve palsy, empty sella, flattening of the posterior aspect of the globe, distention of the peri-optic subarachnoid space with or without a tortuous optic nerve, and transverse venous sinus stenosis.[2] Thus, the heterogeneous presentations associated with IIH can make the diagnosis challenging for the clinician. Though commonly seen in obese women of childbearing age, IIH can occur at any age and may be associated with a variety of symptoms. Some atypical presentations of IIH include the absence of papilledema, cranial nerve palsies beyond an abducens nerve palsy, olfactory or auditory dysfunction, and CSF leakage.[3]

PRIMARY OBJECTIVE

We report the case of an obese, prepubertal girl diagnosed with IIH in the absence of papilledema to serve as a reminder to consider this diagnosis in children with chronic headaches refractory to medications presenting with acute atypical neurologic findings.

SUBJECT PRESENTATION

An eight-year-old girl with a past medical history of obesity (BMI > 99 %ile, Z=2.23), chronic headaches, and a family history of neurologic pathology including migraines and pseudotumor cerebri in a younger sister (non-obese, pre-menarche) as well as a maternal aunt, presented to the emergency department with four days of unilateral left-sided facial and upper extremity weakness. She reported a pounding headache localized to her occipital region, difficulty closing her left eye, slight drooping of the left side of her mouth, and onset of weakness in her left arm. She was evaluated three days prior for similar complaints and discharged home on a 10-day steroid taper for idiopathic Bell's Palsy. The mother returned with the patient to the emergency department (ED), given her concern about the patient's continued neurologic symptoms despite steroid therapy.

Previous work-up from an earlier ED visit with hospitalization due to severe headache one month prior to this assessment included computed tomography (CT), magnetic resonance imaging (MRI), and magnetic resonance venography (MRV), all of which showed no signs of intracranial abnormalities. The fundoscopic exam was negative for papilledema. Her occipital pain was minimally responsive to over-the-counter anti-inflammatories, migraine cocktails, and abortive migraine treatment with dihydroergotamine. Her pain behaviors did not appear to correlate with her pain scale. Despite consistently elevated pain scores, she maintained functional activities, including tolerating loud noises, bright lights, and walks to the playroom. The patient was established with pediatric neurology for the management of her intractable headaches and was receiving treatment with amitriptyline.

Upon this admission, repeat MRI and MRV were obtained because of the progression of her neurologic symptoms, and the results were negative. A urinalysis, PT/INR, PTT, CBC, and CMP were unremarkable. Herpes simplex virus and Lyme serologies were negative. She was treated with intravenous fluids, placed on a migraine cocktail, and continued on her steroid taper.

Despite a previously negative ophthalmic exam for papilledema, a lumbar puncture (LP) was deemed medically necessary to evaluate for IIH due to the patient's family history, continued headaches refractory to standard migraine therapies, and acute left facial nerve palsy. The sedated LP was diagnostic and therapeutic for IIH. The opening cerebrospinal fluid (CSF) pressure was 31 cm of H20. Routine CSF studies were all normal, and symptoms resolved following the procedure. Maintenance acetazolamide therapy was begun, and instructions were given for outpatient follow-up with pediatric neurology.

DISCUSSION

The initial diagnosis of a headache with associated neurological weakness in a child presenting to the ED is broad. Life-threatening etiologies explaining these symptoms must be excluded promptly. IIH is rare in children less than 18 years of age, with an incidence rate of 0.63 to 0.90 per 100,000 population.[4,5] If left untreated, IIH can lead to permanent vision loss and significant morbidity due to chronic headaches.[6] Diagnosing IIH in the pediatric population, particularly in patients <\less than 11 years of age poses a unique challenge due to the increased likelihood of atypical symptoms or asymptomatic presentation as compared to older children and the adult population.[4] Atypical presentations include IIH in the absence of papilledema, facial nerve palsy, hearing loss, vestibular dysfunction, and seizures, among other clinical features.[3]

Obesity and female gender are not risk factors for IIH in prepubertal children, while they represent the most significant risk factors for IIH in adults.[7] Aylward et al. analyzed 203 cases of IIH, 142 of which were diagnosed as IIH, and found a female-to-male ratio of 1:1.04 for prepubertal and 6:1 for post-pubertal primary intracranial hypertension patients.[8]

Our case of an obese 8-year-old female with chronic occipital headaches refractory to standard inpatient abortive headache therapies and features of acute unilateral facial nerve palsy and left extremity weakness. The refractory headache, atypical neurologic symptoms, and strong family history of IIH made IIH a plausible diagnosis. Her neurologic examination and imaging studies did not support obstructive and vascular intracranial abnormalities. The diagnosis of IIH was confirmed with the demonstration of increased CSF opening pressure and symptomatic improvement following the procedure.

Several observational studies describe the emergence of differing neurologic symptomatology based on the ages of patients presenting with IIH. In general, atypical disease presentations inherently increase the probability of misdiagnosis. Facial nerve palsy is an uncommon finding among patients with IIH, and abducens nerve palsy is much more prevalent. In two observational reports, one identified and diagnosed 30 children with IIH, of whom only one patient had facial nerve palsy, and the other described a case of a 13-year-old girl with both an abducens nerve palsy and contralateral facial nerve palsy due to IIH.[9,10] The pathophysiology and underlying disease mechanism of facial nerve palsy in IIH are poorly understood. However, it is speculated that IIH can cause compression of the intra-temporal segment of the facial nerve, which traverses the facial canal alongside a substantial venous complex.[3]

Our patient also presented with unilateral left upper extremity radiculopathy; the predominant complaint was weakness and loss of strength. Associated radicular symptoms in the setting of IIH have been documented in the literature in observational studies and are currently not well described due to their rarity. Several case reports describe adolescents and young adult females who were later diagnosed with IIH, but all initially presented with varying radicular symptoms in the form of acute-onset quadriparesis and bilateral limb paresthesias, raising concern for an acute inflammatory or infectious disease etiology.[10-12] These patients experienced symptom relief following diagnostic and therapeutic interventions that lowered their intracranial pressure, as occurred in our patient. Although the exact mechanism of these findings is unknown, it is suggested that high intracranial pressures result in mechanical compression of various nerve roots. This has been reported in a patient diagnosed with IIH with distended root pouches and enlargement of their spinal subarachnoid space, supporting this etiologic hypothesis.[10,12]

IIH may occur without papilledema, an atypical presentation more commonly seen in adults and rarely documented in the pediatric population.[8,13-15] Optic nerve dysfunction has been described as a loss of visual acuity, reported in 6-20% of children with IIH, and visual field loss occurring in up to 91% of cases.[16] Several studies describe children presenting with chronic headaches without papilledema who were diagnosed with IIH and had CSF opening pressures ranging from 32 to 44 cm of H_20.[17,18] Suggested mechanisms behind the absence of papilledema in IIH include congenital or acquired optic nerve sheath defects, resolution of papilledema in chronic IIH, and intermittent ICP elevations below the threshold required to facilitate papilledema.[18]

In conclusion, the presentation of IIH as an isolated facial nerve palsy is rare. Still, it should be considered even in the absence of papilledema or other forms of visual disturbances on physical examination. Our case serves as a reminder that IIH cannot definitively be such a scenario. It also highlights the importance of thorough history-taking and the probable influence of genetic components in the etiology, as demonstrated by a positive family history. A high index of suspicion for IIH should remain, particularly in younger children, as they are most likely to present with atypical symptoms and can have lifelong visual impairments or complete vision loss if left untreated.

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