



IDIOPATHIC INTRACRANIAL HYPERTENSION PRESENTING WITH UNILATERAL LOWER MOTOR FACIAL NERVE PALSY: A RARE CASE REPORT

Neurology

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ABSTRACT

Idiopathic intracranial hypertension, is a disorder characterized by increased intracranial pressure in the absence of other structural and obstructive lesions that is predominantly seen in obese women of childbearing age. Patients with idiopathic intracranial hypertension commonly present with a headache, transient visual obscuration, diminution of vision and tinnitus with some cranial nerves occasionally involved, most commonly CN VI. We report idiopathic intracranial hypertension (IIH) presenting with isolated complete lower motor unilateral facial nerve palsy which is a presentation rarely reported in the literature.

KEYWORDS

idiopathic intracranial hypertension, lower motor unilateral facial nerve , VII palsy

INTRODUCTION

Idiopathic intracranial hypertension (IIH), is a disorder characterized by increased intracranial pressure (ICP) of unclear pathogenesis, in the absence of intracranial mass lesions or clear cerebrospinal fluid (CSF) outflow obstruction [1]. IIH is predominantly seen in overweight and obese women of childbearing age and has also been associated with high-dose vitamin A derivatives, tetracycline, and oestrogen-progestin oral contraceptives. Patients affected by IIH commonly present with headache (92%), transient visual obscurations (72%), and tinnitus (60%). IIH can also be associated with single or multiple cranial nerve (CN) palsies, with 39–59% of the patients having some sort of CNs deficit. The most common CN palsy is that of CN VI, documented in 12% of adults and 9–48% of children. Less frequently, palsies of other CNs can be encountered, including CN III, IV, VII, IX, and XII. In a limited number of cases, CN VII (facial nerve) palsy has been reported in association with IIH and other CN and/or CNs involvement [2] and, in even fewer cases, as isolated unilateral CN VII palsy.

Case presentation

A 19 year old unmarried college going overweight girl with body mass index (BMI) of 26, presented to our outpatient department complaining of bilateral frontal headache for 7 days. Her headache started insidiously, was associated with vomiting, transient visual obscuration in her right eye, and ringing in both ears. A day prior to the presentation, she started to feel numbness and weakness of the left side of her face, along with an inability to close her left eye properly. She also complained of diminution of vision in both eyes for last 3 days. She denied having diplopia, photophobia, or any feeling of weakness, numbness, or tingling in other locations of her body. She had no history of migraine, tick bite, or any recent illness or fever. She was not taking oral contraceptive pills at the time. On clinical evaluation, she was hemodynamically stable and afebrile. On neurological examination, she was fully alert and oriented, and had fluent speech and intact comprehensive abilities. There were no signs of meningeal irritation. Cranial Nerve testing revealed normal size, bilaterally equal and reactive pupils with intact extraocular movements with no nystagmus, saccadic movement or skew. Visual acuity was diminished in both eyes (RE- 6/60, LE- 6-24). Visual field assessment revealed enlargement of blind spot and loss of visual fields. No signs of abducens nerve palsy were present. However, there was facial asymmetry evident by left lower facial droop, flat nasolabial fold, weaker left eye closure, and limited ability to raise the left eyebrow with angle of mouth deviating to right (Fig.1) Facial sensation was equal on both sides, with a strong jaw opening and a midline tongue of good power. Fundus examination revealed bilateral grade IV papilledema (Fig 2) The rest of her neurological examination, including motor function, sensation, reflexes, coordination, and gait analysis, was within normal limits.

Brain magnetic resonance imaging (MRI) with CT venography revealed a stenosis in the lateral transverse sinus, an empty sella turcica, flat globe and diffusion restriction of both ON heads, findings consistent with raised ICP (Fig. 3). Lumbar puncture (LP) was done. CSF opening pressure was 65 cm in lateral decubitus position, which is above the limit of the reference interval. The cytological and chemical findings of the LP were otherwise within normal limits: white blood cells (WBCs) 2, lymphocytes 100%, protein 22, and glucose 68. She was also started on 250 mg of acetazolamide twice daily, increased to 500 mg twice daily next day. Topiramate 50 mg twice a day was also added and ophthalmology opinion for optic nerve fenestration was also sought in view of grade IV papilledema and severe diminution of vision but attendants refused for the procedure. Two days later, she reported a dramatic improvement in both the headache and the facial nerve palsy but poor vision persisted. Her vision improved gradually on medical therapy in follow up over 2 weeks and currently she is on acetazolamide 500 mg twice daily and topiramate 100 mg twice daily with no active symptoms



Figure 1-Left LMN facial palsy

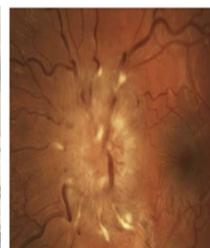


Figure 2 -Grade IV papilledema



Figure 3- Flat globe and empty sella

DISCUSSION:

IIH predominantly affects obese women with an incidence of 11.9 per 100,000 per year in this group compared to its incidence in the general population at 1.56 per 100,000 per year. IIH fits our patient's diagnosis, however, the unusual unilateral facial nerve involvement is atypical and warrants consideration of other differentials, namely brainstem stroke and other structural lesions. These disorders were eventually ruled out by the absence of supporting features on the imaging studies and the clinical presentation as well. The pathophysiology of facial nerve palsy in IIH is not entirely understood and no satisfactory hypotheses seem to exist. Most studies have described it as a false localizing sign, possibly resulting from increased ICP. The extra pressure is believed to exert traction forces on the extra-axial facial nerve [3]. The theory is quite acceptable in cases of combined sixth and seventh nerves dysfunction, due to the well-known fact that the abducens nerve is the most susceptible of all CNs to high ICP

explained by its long course. In contrast, the anatomy of the seventh nerve makes it less prone to such an effect. From the motor nucleus in the pons, facial nerve fibers wrap around the sixth nerve nucleus forming the facial colliculus and then run a relatively short course before reaching the petrous part of the temporal bone where it resides, somehow protected from pressure effects. Given these facts, it is extremely unusual to have an isolated facial nerve palsy from high ICP, with the abducens nerve usually spared. Many imaging findings have been traditionally described in IIH. These include perioptic nerve sheath distention, vertical buckling of optic nerve, globe flattening, optic nerve head protrusion, and an empty sellaturcica which were all present in this patient. Other conditions that were considered in the differential diagnosis of this case including Lyme disease, sarcoid, HIV affecting the nervous system were ruled out by appropriate investigations. Our patient fulfilled all criteria of IIH as described in its latest revision by Friedman et al. in 2013 [4], with the lack of enough diagnostic evidence to support the other differential diagnoses. In cases in which the medical therapy fails, certain surgical procedures can be sought out in order to stop the progression of visual loss. These procedures include optic nerve sheath fenestration, stenting the transverse sinus, and placing a shunt for CSF diversion [1].

CONCLUSION-

IIH should be strongly suspected in obese young women presenting with headache and transient visual complaints. The diagnosis of IIH remains one of exclusion and imaging studies should always be performed to rule out other structural and obstructive lesions. Here we aimed to draw attention to the possibility of IIH presenting with isolated unilateral CN VII palsy as the only CN involved, which has very rarely been described. The association between isolated CN VII palsy and IIH needs high index of suspicion by clinicians and it is a diagnosis of exclusion.

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