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Facial palsy and diplopia revealing idiopathic intracranial hypertension in a child

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ABSTRACT

Idiopathic intracranial hypertension is rare in the pediatric population. It's characterized by increased intracranial pressure in the absence of any evident underlying neurologic disease. The most frequent cranial nerve deficit associated with that condition is abducens nerve palsy. The association of sixth and seventh cranial nerve damage is uncommon. In this report, we describe the case of an 8-year-old girl who presented with headache, diplopia, and peripheral facial palsy related to idiopathic intracranial hypertension.

KEY WORDS: idiopathic intracranial hypertension; abducens; facial; palsy; child

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INTRODUCTION

Idiopathic intracranial hypertension (IIH), also known as benign intracranial hypertension or pseudotumor cerebri, is an uncommon condition defined by elevated intracranial pressure without any evident underlying neurologic disease. Abducens nerve palsy is the most prevalent cranial nerve (CN) impairment associated with that disorder. The association of VIth and VIIth CN damage is extremely rare.

We report the case of an 8-year-old girl who presented with peripheral facial palsy and diplopia that later proved to be symptoms of IIH.

CASE PRESENTATION

An 8-year-old girl presented to the hospital with horizontal diplopia and bifrontal headaches for 2 days, with a feeling of numbness on the right side of her face.

On initial examination, the patient was alert, well oriented, and apyretic. Physical exam showed a facial asymmetry with right facial muscles weakness and an asymmetrical eye closure with a right Souques' eyelashes sign (Fig. 1AB). The rest of the otolaryngological exam and audiological evaluation were otherwise normal. The ophthalmologic examination found a preserved visual acuity at 6/6 in both eyes with horizontal diplopia in binocular vision and a discreet limitation of the external gaze of both eyes. The Hess-Lancaster test confirmed the right abducens paresis. At the slit lamp examination, the anterior segment was normal with no pupillary deficit. Examination of the fundus revealed bilateral stasis papillary edema (Fig. 2AB). The pa-

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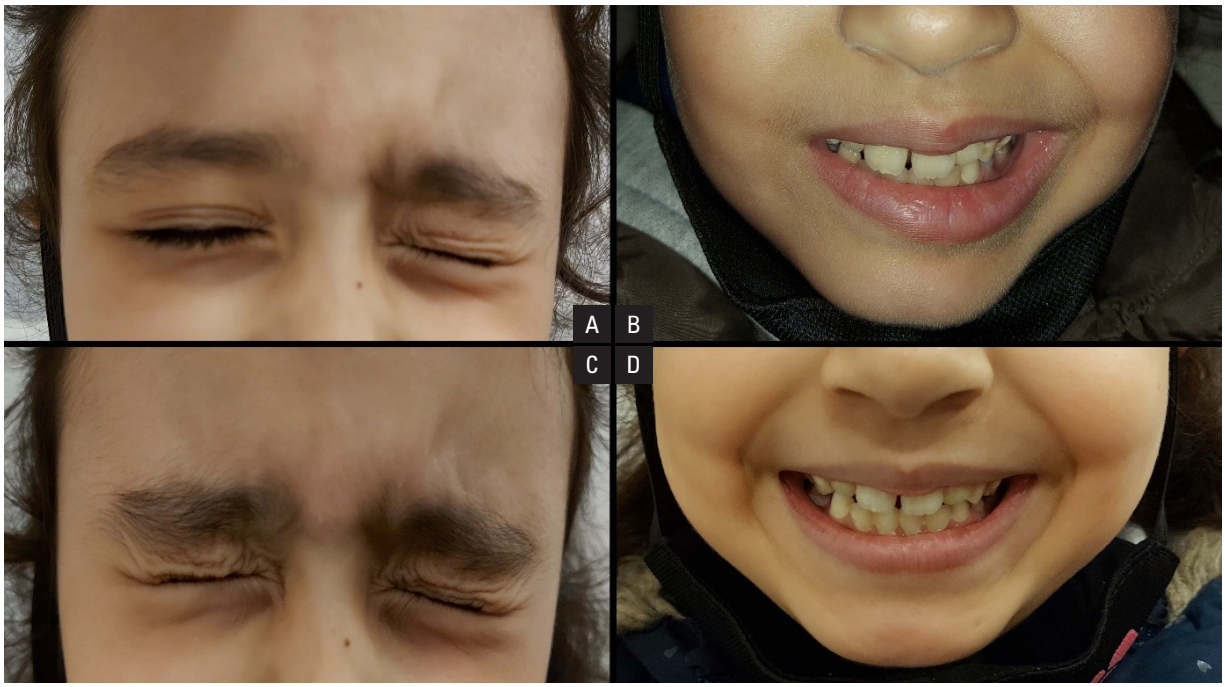

FIGURE 1. Evolution of facial paralysis. **A**. Asymmetrical eye closure with a right Souques' eyelashes sign; **B.** Asymmetrical mouth movement with a weakness on the right side; **CD.** 2 weeks follow-up: Resolution of the right facial palsy with symmetrical eye and mouth movements

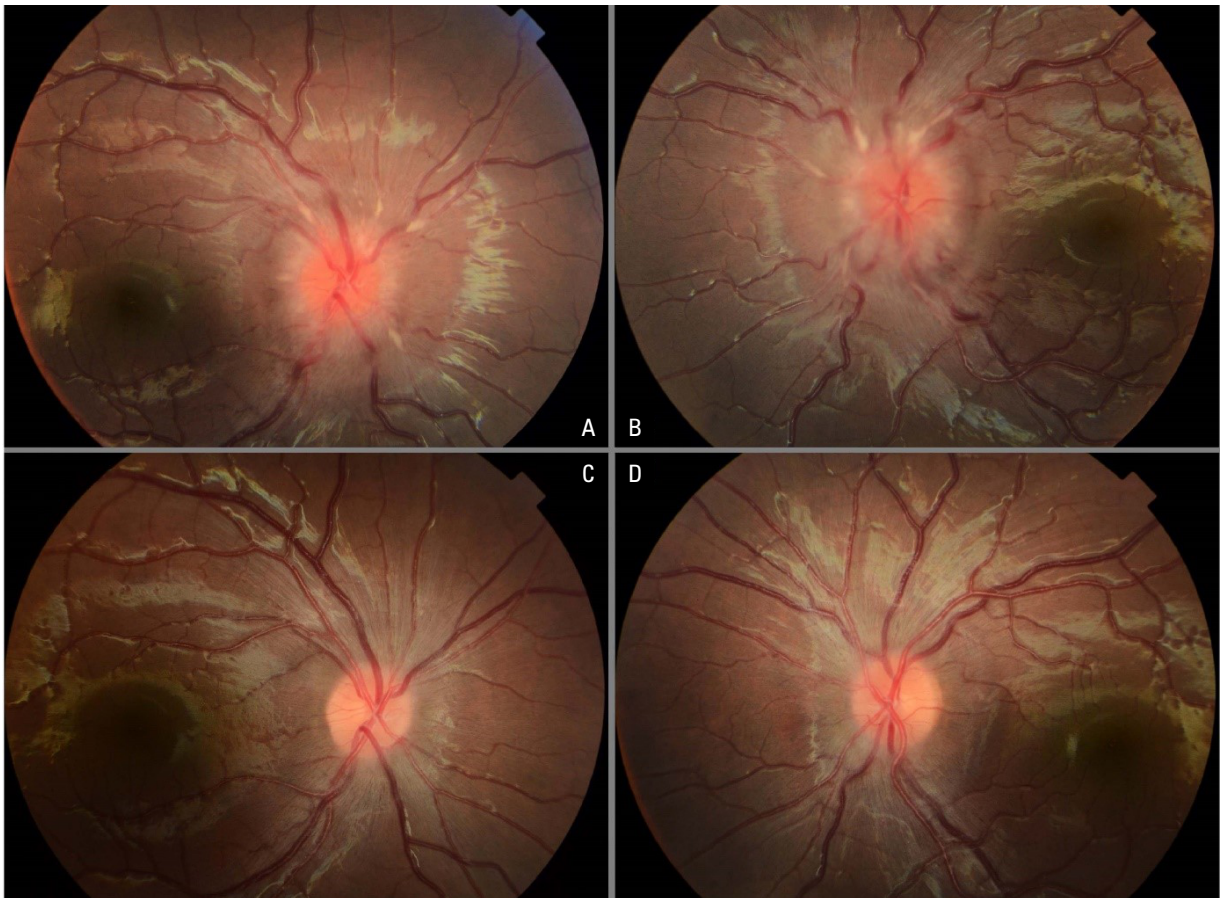

FIGURE 2. Evolution of the papilledema on fundoscopy. **AB.** Funduscopy at admission showing bilateral papilledema; **CD.** Follow-up fundoscopy after 1 month of treatment, showing a complete resolution of the papilledema in both eyes

tient had no meningeal signs. The results of the rest of her neurological examination, including motor function, sensation, reflexes, coordination, and gait analysis, were normal.

Lumbar puncture (LP) revealed clear and colorless cerebrospinal fluid (CSF) with an opening pressure of 45 cm H_2O. The cytological and chemical findings of the LP were otherwise normal. Initial hematological, immunological, and biochemical tests were normal. Cerebral angio-MRI showed no abnormality.

The patient was diagnosed with idiopathic intracranial hypertension and was started on oral acetazolamide (5 mg/kg/d). The evolution was marked by rapid clinical improvement. The abducens paresis, along with the headaches and diplopia, resolved within the next three days. The facial paralysis fully recovered on day 15 (Fig. 1CD). Papilledema resolved gradually, and normal fundoscopic examination was obtained within 1 month of treatment (Fig. 2CD).

DISCUSSION

Idiopathic intracranial hypertension is uncommon in children, with an incidence of 0.1 to 0.9 per 100,000 children [1]. Prepubertal children are particularly less at risk for it, and pubertal adolescent girls experience it more frequently than boys [2]. Studies also confirmed a positive relationship between obesity and age at diagnosis of pediatric IIH [3].

Patients with IIH commonly present with headaches, vomiting and visual complaints. A papilledema and unilateral or bilateral VIth nerve palsy are frequently found during an ophthalmological examination. It affects 9 to 48% of children with IIH, making it the most prevalent cranial nerve deficiency [4]. Other cranial nerve palsies are rarely involved. To our knowledge, there have only been 5 pediatric case reports of the association of VIth and VIIth nerve palsies in the literature.

Diagnosis is based on the modified Dandy criteria: (1) signs of elevated intracranial pressure, (2) absence of localizing symptoms other than abducens (sixth) nerve palsy, (3) patient awareness, (4) normal computed tomography/magnetic resonance imaging (MRI) results, (5) lumber puncture opening pressure of > 25 cm H_2O, the normal biochemical and cytological composition of CSF fluid, and (6) absence of any other cause for the elevated intracranial pressure [5].

Although the pathogenesis is unclear, several explanations were suggested, including the restriction of cerebrospinal fluid exit through the arachnoid villi, accelerated production of cerebrospinal fluid at the choroidal plexus, and elevated cerebral venous sinus pressure [6]. Particularly vulnerable to increased intracranial pressure is the sixth cranial nerve. Due to its long intracranial course, resulting in reversible dysfunction of the nerve [7].

The concomitant facial palsy may be caused by pressure-related phenomena with traction on the extra axial facial nerve [1]. However, seventh CN fibers run a reasonably short course before they enter the petrous temporal bone of the skull. It is then less likely to be affected by elevated intracranial pressure [7]. Pressure effects of IIH on VIIth CN are perhaps felt mostly from the Pons, where the sixth nerve's nucleus is wrapped in the fibers of the seventh nerve. The lower motor fibers of the seventh nerve may be affected by any lesion to the sixth nerve nucleus [7].

As far as treatment is concerned, most authors believe that acetazolamide (carbonic anhydrase inhibitor) in appropriate doses, along with weight loss efforts, should continue to be the first-line therapy for IIH patients [9] and prohibit corticosteroids as it tends to aggravate the IIH. However, according to Tzoufi [6], the initial acetazolamide treatment can be supplemented with corticosteroids without worsening existing severe intracranial hypertension or ophthalmologic abnormalities [6].

On the other hand, 18–22% of IIH patients do not respond to medical treatments and request surgical procedures, which include bariatric surgery, venous sinus stenting, optic nerve sheath fenestration, and CSF diversion techniques [8].

The evolution of IIH patients is often positive. In most cases, a good prognosis with papilledema resolution occurs in 3 to 6 months [2]. IIH recurrence is not common, and it affects children less frequently than it does adults (between 6% to 22% *vs.* 40% of total patients) [2].

CONCLUSION

Many questions remain unanswered about idiopathic intracranial hypertension. The physiopathology of cranial nerve damage remains unclear. The case we present shows how IIH can manifest with unusual symptoms. Although facial paralysis can be frequently encountered among children, any other associated sign should lead to thorough investiga-

tions. Early management of IIH often offers good results and can avoid permanent visual disability.

Conflict of interest

The authors declare that they have no competing interest.

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