FULMINANT IDIOPATHIC INTRACRANIAL HYPERTENSION IN A PEDIATRIC PATIENT FOLLOWING A MINOR HEAD TRAUMA

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Idiopathic intracranial hypertension (IIH) is a condition defined by elevated intracranial pressure but no clinical, laboratory, or radiographic evidence of responsible infection, vascular abnormality, space occupying lesion or hydrocephalus[1]. The incidence of IIH in the general population is 0,9 cases per 100,000 people annually. It is considered rare in children and exceedingly rare in infants and neonates. In pediatric population, there is no predilection toward females being more common in adolescents. The disease in post-pubertal children has a female predominance and association with obesity, similar to in adults[1,2]. Some conditions have been identified as causative agents of secondary pseudotumor cerebri, including certain medications (e.g. vitamin A/D, several antibiotics, steroids, retinoic acid, lithium), endocrine abnormalities, autoimmune disorders, anemias, and cranial venous outflow abnormalities. The role of associated diseases and medications seems to be stronger in children than in adults[1,2]. Atypical presentations include uncommon conditions leading to IIH or unusual clinical presentation other than the more typical progressive and insidious clinical course. The term fulminant idiopathic intracranial hypertension is reserved for those patients presenting with IIH with an acute presentation and rapidly progressive visual loss with very few cases reported so far[3].

We present on a definite case of fulminant IIH in a 11 years-old boy after a minor head trauma.

CASE

An 11-year-old male patient was admitted with diplopia of one week's duration. The patient was born to non-consanguineous parents, with an uneventful gestational history. He was born by normal delivery, with an uneventful neonatal period. His early development was normal and also presented normal neuropsychomotor acquisitions. There was also, no family history of neurological diseases.

The patient was previously healthy without clinical history of any medical conditions, receiving neither prescribed medications nor over the counter medications. The patient was not receiving any type of vitamin replacement.

Fifteen days before the admission, the patient presented a minor head trauma while he was swimming. The trauma was described as very mild, during leisure activities, without any immediate complaints or needing any medical attendance. Four days after the head trauma, the patient start with vomits, several times a day, bilateral periorbitary moderate pain. One day later, the patient started with visual loss and diplopia.

On physical examination, he was noted to be in good overall condition showing a normal physical examination without fever or any other signs of infection. His height was 147 centimeters, weighting 54 kilograms, and with a body mass index of 25 (97 centile)

The neurological examination was noteworthy for diplopia, convergent strabismus, and bilateral sixth nerve paresis (Fig 1). Fundoscopi examination revealed bilateral papilledema (Fig 2).

Laboratory evaluations, including a complete blood count, blood electrolytes, urea, creatinin and liver enzymes gave results at normal range.

Ophthalmologic evaluation revealed acute papilledema and diminished visual acuity.

Cranial MRI revealed signs of intracranial hypertension (Fig 3) but no signs of venous sinus thrombosis in the MRI venography. After neuroimaging procedures, a lumbar puncture was performed revealing an opening pressure of 80 cm H_2O with normal CSF composition with no evidence of pleocytosis, elevated protein or low glucose.

Following the diagnosis of IIH acetazolamide treatment was prescribed associated with repetitive lumbar puncture in order

HIPERTENSÃO INTRACRANIANA IDIOPÁTICA FULMINANTE EM PACIENTE PEDIÁTRICO APÓS TRAUMATISMO CRANIANO LEVE

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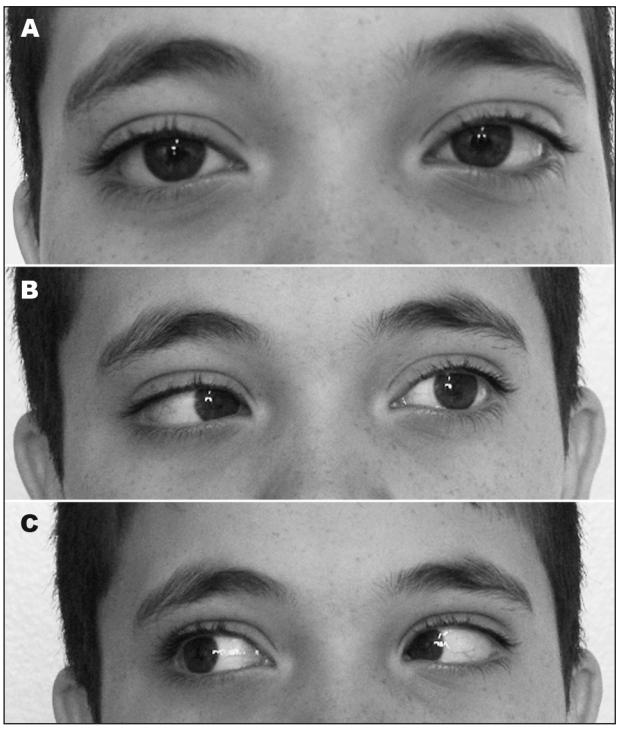

Fig 1. Neurological examination showing convergent strabismus [A] and bilateral sixth nerve paresis [B and C].

to decrease the CSF pressure. Within a week the visual acuity improved, and one month later the patient was symptom free. The patient repeated fundoscopic evaluations were normal and he is currently under regularly clinical follow-up.

The hospital ethics commission approved this case report and the parents gave informed consent for publication.

DISCUSSION

Although IIH can occur at any age, it is far more common in female adults, mainly obese individuals. In childhood, it now appears that there is an increase incidence of IIH among adolescents (12–15 years) as compared to children (2–12 years)[1]. When considering only cases of fulminant IIH the presented case seems very different from current descriptions[3].

Several theories have been proposed to explain the pathophysiology of IIH, including increased brain volume caused by increased water content, increase blood volume, increased rate of CSF formation and decrease rate of CSF absorption at the corionic villi[4]. It was also suggested that elevated intracranial venous pressure might be an universal mechanism in IIH leading to a rise in CSF and intracranial pressure by resisting CSF absorption[5].

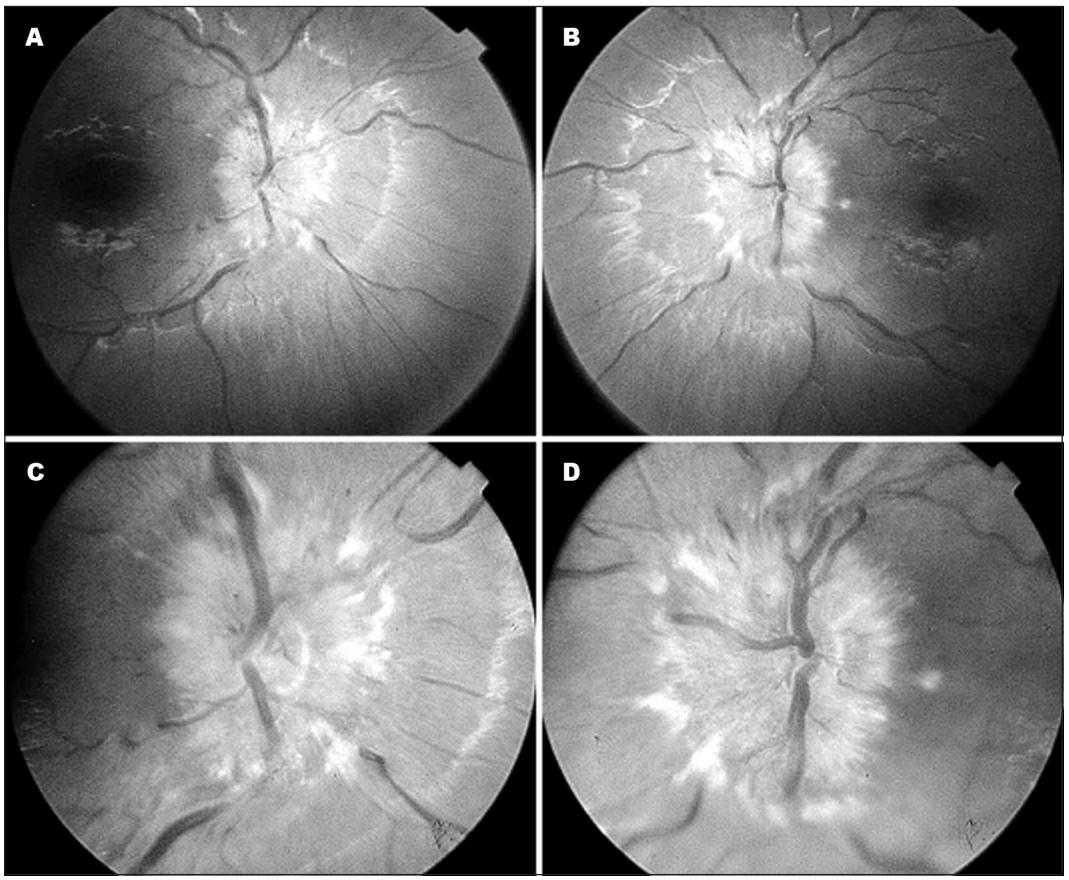

Fig 2. Initial optic nerve appearance (the right eye is on the left and the left eye is on the right) showing bilateral acute disc edema with dilated veins.

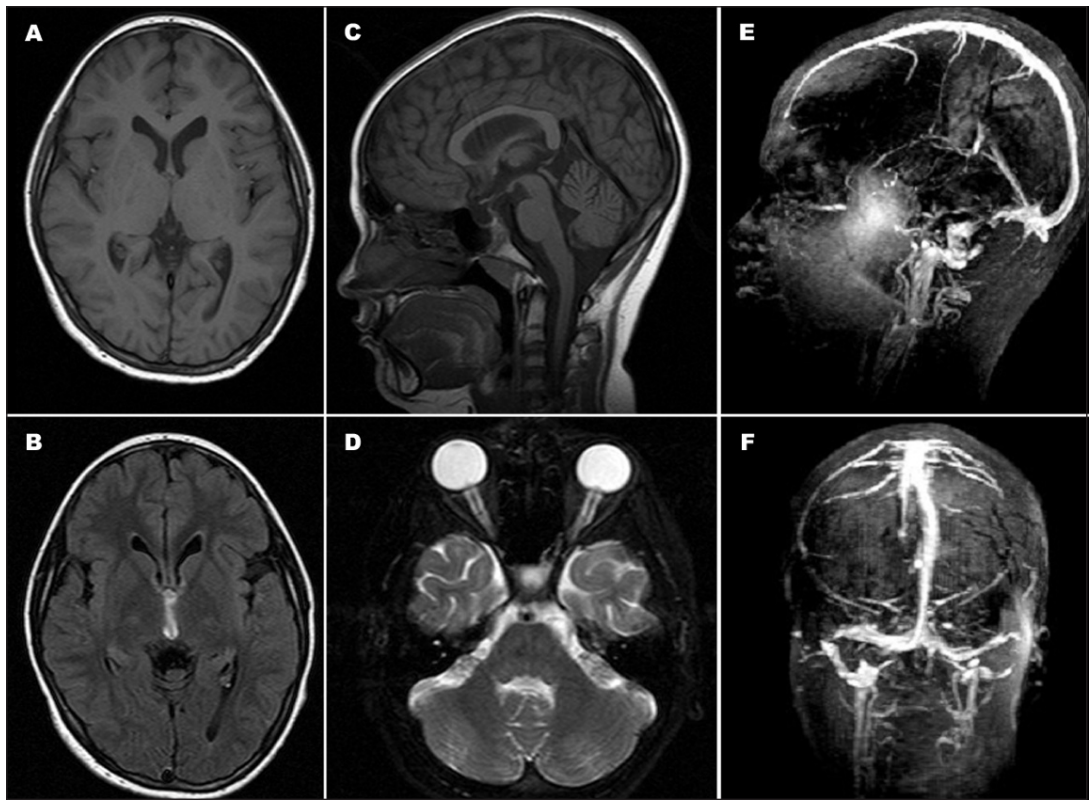

Fig 3. [A] T1-weighted and [B] FLAIR axial MR images with no evidence of hydrocephalus, mass, or structural lesions. [C] T1-weighted sagital MR image shows a partially empty sella. [D] T2-weighted axial MR image shows flattening of the posterior sclera, distension of the perioptic subarachnoid space, and tortuosity of the optic nerves. [E and F] Magnetic resonance venography showing dominant left transverse sinus and no signs of venous sinus thrombosis.

Currently, IIH is diagnosed as following: (1) symptoms and signs of generalized intracranial hypertension; (2) documented elevated ICP; (3) normal CSF composition; and (4) no evidence of hydrocephalus, mass, or structural or vascular lesions on brain MRI specifically, no evidence of cerebral venous thrombosis[6]. In order to identify cases of fulminant IIH, the following definitions are considered: (1) acute onset of symptoms and signs of intracranial hypertension; (2) less than 4 weeks between onset of initial symptoms and visual loss; (3) rapid worsening of visual loss; and (4) MRI and MRI venogram ruling out venous thrombosis[3]. Our patient, fulfilled all the diagnostic criteria for fulminant IIH.

Fulminant IIH usually, do not differ from those with typical IIH, except for the temporal profile and the visual loss. Typically, in cases of IIH, visual acuity is affected late in the course of IIH, in large series of patients, the prevalence of loss of visual acuity as a presenting feature of the illness is low, and usually, the deterioration of visual acuity is gradually experienced over a long period of time[7,8].

Our patient presented with severe visual loss associated with vomits and diplopia only five days after a minor head trauma. Because of this atypical presentation a throughout workup ruling out a secondary cause of isolated raised ICP was performed (specifically for meningeal process and cerebral venous thrombosis).

Similar reports of fulminant IIH are exceedingly rare in the literature and the pathophysiology remains unknown[9]. Identifiable conditions have been associated with IIH, especially in pediatric patients, the most common of which include drugs (such as tetracyclines, vitamin A, steroids, and chemotherapy agents); infections (acute sinusitis, varicella, measles); and endocrine conditions[1].

In adults, there is a well-established association between obesity and IIH. In children this association is usually found in older patients[1]. When taking it into account, our patient could not be considered obese, presenting a body mass index (IMC) of 25 (at 97 centile) classified as overweight. None of the other above-mentioned conditions was found in our patient.

In the clinical history, however, it was mentioned an episode of minor head trauma few days before the symptoms onset. Minor head trauma was previously reported in association with pediatric IIH but in most of the cases, later neuroimaging examinations demonstrated cerebral venous sinus thrombosis or cerebral edema[1]. In our patient, MRI and MRI venography ruled out any sings of cerebral edema or venous thrombosis.

In our patient, it was found in the MRI venography an asymmetry in the right transverse venous sinus. It is well known that the brain hemispheres are anatomically and functionally asymmetric and asymmetries are also usually find in the venous system[10]. In a study of 100 patients with normal MRI imaging of the brain, MRI venography revealed a dominant right transverse sinus in 59 patients, a dominant left transverse sinus in 30 and a co-dominant transverse sinus in ten[11]. In children, those asymmetries are also usually find, in a study of 50 children ranging from 3 months to 17 years old, MRI venography showed a dominant right transverse sinus in 27 (54%) patients, a dominant left transverse sinus in 18 (36%), 4 co-dominant transverse sinus and 1 absence of both transverse sinus[10]. Those asymmetries were not associated with clinical or neurological symptoms.

Kantarci et al. studied the correlation between venous sinus asymmetries through MRI venography and intraocular pressure measured through pneumotonometry in patients without neither neurological nor ophthalmologic diseases. As a general conclusion it stated that if the size of transverse sinus in one side is larger and its venous drainage is greater, intraocular pressure of this side is lower. It can be speculated that the transverse sinus sizes may be associated with pathogenesis of diseases with increased intraocular pressure such as glaucoma[12].

Stating that slightly differences in the venous drainage due to transverse sinus asymmetries could be responsive to an idiopathic intracranial hypertension would be highly speculative and further studies in this direction is needed.

The syndrome of IIH is already known for over a century since its early descriptions by Quincke in 1893, and until nowadays it remains perplexing medical professionals worldwide with its variable underlying conditions, clinical presentation, and outcome. Herein we presented a unique case of fulminant IIH in a pediatric patient.

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