Short communication

Idiopathic intracranial hypertension in a patient with Chiari I malformation

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ABSTRACT

Case report: The case involves a 22-year-old woman who presented with headache and decreased vision. She showed asymmetric papilledema, and a 6-mm tonsillar descent was observed in the image tests. She was diagnosed with secondary intracranial hypertension coinciding with the symptoms of a Chiari malformation (MC).

Discussion: Chiari malformation type I is the most common in this group of malformations, and is characterized by a greater than 5 mm descent of the tonsils, being able to cause increased intracranial pressure and papilledema by blocking the flow of the cerebrospinal fluid. In this case, the MC was not the responsible for triggering the secondary intracranial hypertension, but a mere coincidence of both processes.

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Hipertensión intracraneal idiopática en paciente con malformación de Chiari tipo I

RESUMEN

Caso clínico: Mujer de 22 años que consultó por cefalea y disminución de la visión. Presentaba papiledema asimétrico y, en las pruebas de imagen, un descenso amigdalar de 6 mm, siendo diagnosticada de hipertensión intracraneal idiopática coincidente con una malformación de Chiari (MC).


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Introduction

Chiari malformations (CM) are structural defects that affect the cerebellum and brain stem, which are displaced below the foramen magnum, blocking the flow of cerebrospinal fluid (CSF) giving rise to intracranial hypertension (ICH). Of unknown origin, this malformation can be genetic or acquired due to exposure to substances, and deficiency of vitamins and nutrients. CM are classified according to the structure that moves toward the spinal chord.\(^1\)–\(^3\) The most frequent form of CM is a type I which could even be asymptomatic. Diagnosis is usually in adolescence or adulthood, and imaging tests reveal an extension of the cerebellum amygdala toward the foramen magnum, without involving the brain stem, exceeding 5 mm.\(^1\)–\(^4\)

This paper presents the case of a patient with asymmetric papilledema and retrohyaloid hemorrhages with CM type I, the initial diagnostic of which was given as cause for ICH, even though the presence of both processes was unrelated. Finally, the diagnostic was idiopathic intracranial hypertension (IIH) in a patient with CM type I.

Case report

Female, 22, who visited the emergency section due to holocranial headache and diminished visual acuity as well as myodesopsia in the left eye (LE) with several weeks of evolution. The patient did not refer personal or familial history of relevance or known allergy to drugs. In addition, she did not refer intoxication habits or usual consumption of pharmaceutical drugs. The patient presented obesity type I with a body mass index of 31.89.

The ophthalmological exploration revealed visual acuity of 0.6 in both eyes (BE) with normal anterior pole and intraocular pressure in BE. Ocular fundus revealed asymmetric papilledema with retrohyaloid hemorrhages in LE (Fig. 1). Analysis of the peripapillary retinal fiber layer (RNFL) with optic coherence tomography (Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, California, USA) as well as of the ganglion cells and internal retina plexiform (GCL/IP) by means of macular segmentation revealed edema in the 4 quadrants of BE and GCL/IP unaffected in BE (Fig. 2). Campimetry (Humphrey 24-2) was normal (Fig. 3). Imaging tests exhibited CM type 1 with amygdala descent of 6 mm (Fig. 4). A conservative approach was decided after the patient refused to undergo lumbar puncture (LP), initiating medical treatment with high doses of acetazolamide, potassium and topiramate supplements, which improved clinic and partially improved papilledema and hemorrhages after 2 weeks.

After 4 months of medical and dietetic treatment (which produced weight loss of 18 kg and reduced body mass index to 27.64, considering the patient as overweight), the patient was free of symptoms, without papilledema (Fig. 5) in optic coherence tomography, showing slight inferior sector or edema in LE with normality of the GCL/IP (Fig. 6).

Discussion

The prevalence of CM is unknown\(^5\) although it was typically estimated to be of 1 out of every 1000 births. However, with current imaging techniques, the number of diagnosed cases has risen to 1%.\(^5\) In one-third of patients it can be a casual finding.
Fig. 2 – Optic coherence tomography on peripapillary retinal fiber layer edema and normality in the ganglion cell layer and internal retina plexiform by means of macular segmentation in both eyes.

Fig. 3 – Normal campimetry in the acute phase of the process.
CM type 1 affects predominantly women who are 3 times more predisposed, and is associated to syringomyelia in up to 40% of cases. Symptomatology could go unnoticed because patients consult for insidious and progressive symptoms such as headaches, cervical pain, vertigo or tinnitus. In the case presented herein, it was considered that the amygdala descent could have produced CSF blockage giving rise to ICH and asymmetric papilledema. As this cause was considered to be an objective cause for ICH, IIH was initially discarded. However, the evident response to medical and dietetic treatment modified the case report as well as the diagnostic.

**Fig. 4** – Magnetic resonance showing Chiari type I malformation with amygdala descent of 6 mm.

**Fig. 5** – Normal ocular fundus 4 months after the initial process.

**Fig. 6** – Optic coherence tomography at 4 months after the initial process with inferior residual edema in the peripapillary retinal fiber layer in the left eye, and normality in the retina ganglion cells and internal plexiform by means of macular segmentation in both eyes.
Even though CM type I is the most frequent one, this case features predominantly LE papilledema of high intensity, to the extent that it caused the rupture of the peripapillary blood vessels and retrohyaloid hemorrhages (Fig. 1). In addition, as the patient exhibited diminished vision and myodesopsia, the urgency of the consultation contributed to the diagnostic at the age of 22. Even though in this case both processes occurred simultaneously by pure chance, CM should be taken into account in young patients who debut with ICH. Lumbar puncture must be carried out to complete and confirm the study, as there is no contraindication in this type of malformation. Imaging tests, maybe magnetic resonance, are essential for adequately diagnosing this disease.

As conclusion, a possibly random relationship between processes such as IIH and CM should be taken into account, because this malformation and specifically type I, may not be considered as an objective cause responsible for CSF blockage and secondary ICH.

Conflict of interest

No conflict of interest was declared by the authors.

REFERENCES