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Papilledema and Chiari I in childhood: case report

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Introduction: Chiari malformation type I (CM-I) is a condition characterized by an abnormal craniocervical transition whose main finding is cerebellar tonsillar ectopia of > 5 mm below the foramen magnum. The main clinical manifestations are occipital headache, truncal ataxia, corticospinal tract dysfunctions, and scoliosis.
Objective: The scope of this article is to report a rare case of papilledema and visual impairment in a child diagnosed with CM-I.
Methods/case presentation: Presentation of the case of an 8-year-old child with difficulty in performing school activities due to visual blurring in the 3 days prior to hospitalization. She was taken by her parents to the ophthalmologist who identified bilateral papilledema and was urgently referred to the pediatric emergency unit. She underwent magnetic resonance imaging of the brain and CM-I was identified. She underwent urgent surgical posterior cranial fossa decompression and progressed to complete visual improvement, confirmed by ophthalmological examination.
Conclusion: Papilledema may be the initial clinical manifestation of CM-I in children and urgent surgical decompression of the posterior cranial fossa may prevent or even reverse the visual impairment.

Keywords: Arnold-Chiari malformation, papilledema, amaurosis, case report, neurosurgery

INTRODUCTION

Chiari malformation was initially described in the late 19th century by pathologists Julius Arnold (1835-1915) and Hans Chiari (1851-1916) [1] and can be classified in grades from 0 to IV, with Chiari malformation type I (CM-I) being the most prevalent [2,3,4,5,6]. The main clinical manifestations described in CM-I are occipital headache associated with cough or Valsalva maneuver, respiratory disorders, truncal ataxia, corticospinal tract dysfunctions, and scoliosis [3,5]. Papilledema and visual complaints are rarely reported, especially in the pediatric group, and their presence is an indication for urgent posterior fossa decompression (PFD) [7,8,9,10,11]. The scope of this article is to report a rare presentation of rapidly progressive visual loss associated with bilateral papilledema in a child diagnosed with CM-I.

CASE REPORT

Study approved by the institutional research ethics committee (CAAE: 70256723.5.0000.0048) after the consent and authorization of parents and guardians for the disclosure and reporting of the case and photos/images without identification of the child. The study was conducted ethically and in accordance with the Declaration of Helsinki.

An 8-year-old girl with a previous diagnosis of CM-I discovered incidentally after performing a computed tomography (CT) scan of the skull due to horse fall. Magnetic resonance imaging (MRI) of the brain performed after the CT scan confirmed the diagnosis of CM-I, showing a cerebellar tonsillar descent below the foramen magnum of > 5mm. There was no hydrocephalus or syringomyelia. Semiannual clinical follow-up and annual radiological follow-up were initiated, considering that the child was completely asymptomatic.

After 18 months without symptoms, she was admitted to the pediatric emergency with a complaint of visual blurring and rapidly progressive visual loss in the 3 days prior to hospitalization associated with difficulty in carrying out school activities (imprecise filling or coloring of paintings and drawings, difficulty in keeping writing on the line and differentiating colors), inability to play on the tablet and cell phone (games and applications available in these electronic devices). There was no report of accidental trauma. In view of the child's complaints, the parents took her to an ophthalmological evaluation in which bilateral papilledema was detected (Figure 1) and the child was referred to the pediatric emergency.

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Figure 1- Retinography showing papilledema due to Chiari I.

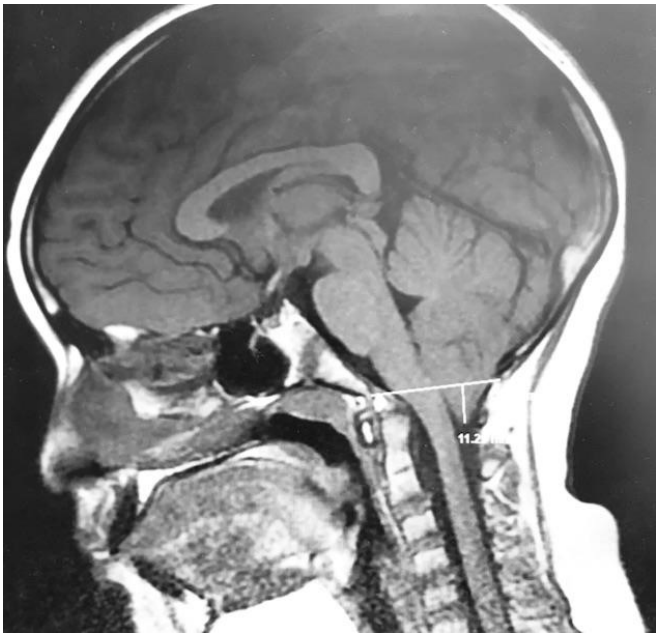

Figure 2- Magnetic resonance imaging of the brain showing the presence of Chiari malformation type I without hydrocephalus, with cerebellar tonsillar herniation of approximately 11 mm below the foramen magnum.

In the neurological examination at admission, during confrontation visual field testing, the child needed to move closer to the examiner to distinguish the number of fingers shown. She was unable to differentiate the images, figures and colors of children's drawings and stickers of the pediatric emergency unit. There were no motor deficits or difficulties in the cerebellar static or dynamic balance tests. The finger-nose and heel-knee tests were also normal. Urgent MRI of the brain was performed, which corroborated the diagnosis of CM-I without hydrocephalus and identified the descent of the cerebellar tonsils approximately 11 mm below the foramen magnum (Figure 2). In view of the diagnosis of CM-I, and despite the papilledema and the possibility of intracranial hypertension, the medical team decided not to perform the measurement of cerebrospinal fluid (CSF) pressure through lumbar puncture manometry. The child was submitted to urgent PFD.

PFD included the removal of the posterior arch of C1, opening of the dura mater and duraplasty. Because it was an 8-year-old child, the limits defined for PFD were 2 x 2.5 cm with the foramen magnum as reference (Figure 3). A Y-shaped opening of the dura mater was performed and surgical decompression was completed after the removal of the posterior arch of C1. The closure of the dura mater was performed with an autologous graft (pericranium), and a small area of the dura mater required the use of a

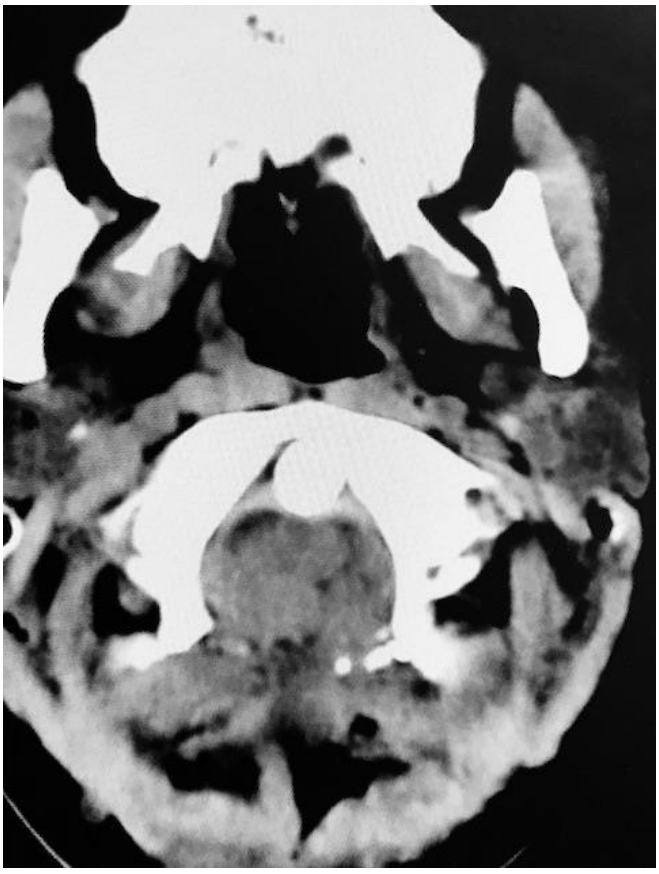

Figure 3- Postoperative computed tomography scan of the skull after decompression of the posterior cranial fossa and removal of the posterior arch of C1.

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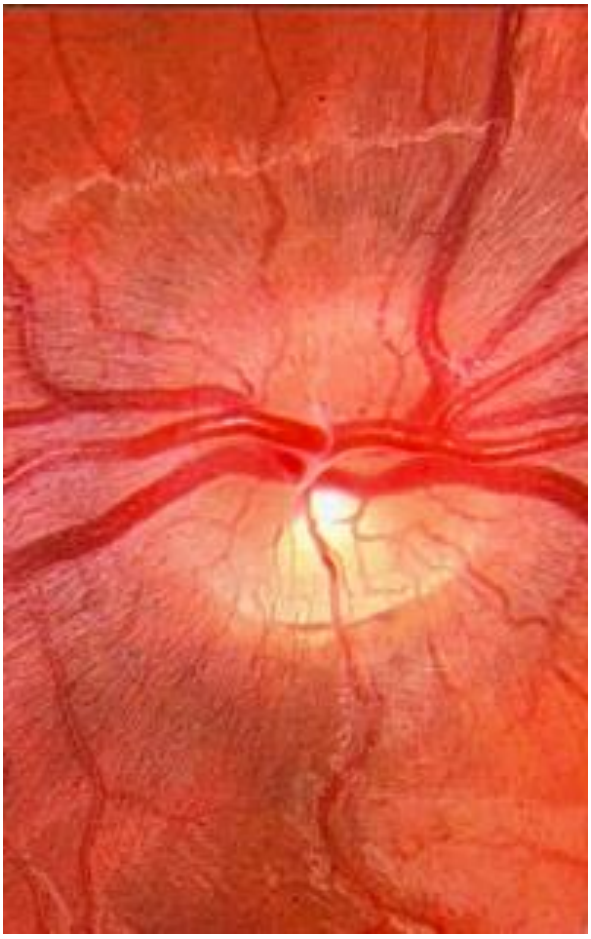

Figure 4- Retinography showing improvement of papilledema on the seventh day after decompression of the posterior cranial fossa.

heterologous substitute to complete the closure. There were no surgical complications.

On the second day after surgery, the child reported visual improvement, but continued with blurred vision and difficulty differentiating colors and objects. She was still unable to use the tablet or mobile phone and had difficulty seeing television. The ophthalmological evaluation on the second postoperative day showed stabilization of the papilledema and normal tonometry for age (12 x 12). There was a progressive improvement in visual acuity, reported by the child, and by the seventh day after surgery she was able to use the usual mobile phone applications, watch television and distinguish images and colors. The ophthalmological evaluation performed on the seventh postoperative day showed improvement of the papilledema (Figure 4). When the child returned for outpatient follow-up on the 21st day after surgery, she reported complete recover of normal vision, as corroborated by the parental observations.

DISCUSSION

CM-I consists of a congenital malformation of the posterior cranial fossa with cerebellar tonsillar herniation of > 5mm through the foramen magnum accompanied or not by syringomyelia [7,12]. Approximately 50% of children with CM-I are diagnosed incidentally and remain asymptomatic or with minimal clinical signs for a long time, without the need for neurosurgical intervention [13]. When patients develop clinical manifestations, this usually happens in adulthood and the main complaints are headache (mainly occipital headache, related to Valsalva maneuvers), cerebellar disorders, paresis, paresthesias, dysesthesias and impairment of low cranial nerves [7,12,13]. The initial option of non-surgical management in the case reported here was precisely due to the incidental discovery of CM-I, absence of complaints, symptoms, or changes identified in the patient's neurological examination, according to the literature on the subject [3,13].

Approximately 1.3% to 2.7% of patients with CM-I may develop a secondary rise of intracranial pressure possibly associated with impaired cerebrospinal fluid flow through the foramen magnum and sometimes a secondary increase in venous pressure, and the pathophysiology of the papilledema is similar to the theory applied to cases of idiopathic intracranial hypertension (IIH) [8,9,11]. The main complaints in IIH are headache and visual disorders (diplopia, visual loss, scotomas) and the main complication is visual loss, which can be irreversible [11,14]. The report of CM-I associated with papilledema is rare, described in approximately 2% of the cases [7,11]. Although the clinical complaint of the child described here was similar to the picture of IIH, which is a differential diagnosis to be considered, the medical team decided not to perform the manometric CSF pressure assessment at admission because of the risk of acute clinical worsening described in cases of CM-I following CSF sampling by lumbar puncture [8].

Several authors report PFD as the only effective treatment in cases of papilledema secondary to CM-I, suggesting the removal of the posterior arch of C1 and duraplasty [8,10]. The decision for PFD on an urgent basis described here, using a previously established surgical technique, was based on the risk of irreversible blindness [3,10]. During the postoperative follow-up to identify visual improvement after PFD, ophthalmological follow-up in addition to the neurological clinical evaluation is essential. Retinography and ocular tonometry are eye exams described in the literature as possible methods for monitoring and comparing papilledema regression and for indirectly measuring the intracranial pressure by measuring the intraorbital pressure, respectively [15]. The calm and cooperative attitude of the child was a factor that made us decide to perform the ophthalmological follow-up, associated with the reports of the girl and her parents, to identify the clinical improvement. The case described here brings two infrequent events: the development of symptoms of CM-I in the pediatric group and an initial clinical picture characterized by papilledema and visual loss.

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CONCLUSION

Although infrequent, papilledema and visual deficits should be recognized as associated with CM-I. Urgent PFD with the removal of the posterior arch of C1 and duraplasty is the recommended treatment. Multidisciplinary follow-up is important for the adequate definition of visual improvement in the pediatric group.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the Hospital São Rafael Ethics Committee, number: CAAE: 70256723.5.0000.0048

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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CONTRIBUTIONS

-**José Roberto Tude Melo**: Conceptualization, Data curation, Formal Analysis, Funding acquisition, Investigation, Methodology, Project administration, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing
-**Leonardo Conrado Silva Lima**: Funding acquisition, Resources, Validation, Visualization, Writing – review & editing
-**Igor Sandes Pessoa da Silva**: Funding acquisition, Investigation, Resources, Validation, Visualization, Writing – review & editing

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