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CASE REPORT

Importance of Ophthalmological Evaluation in the Conservative Management of Increased Intracranial Pressure from a Ruptured Arachnoid Cyst

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ABSTRACT

Arachnoid cysts (ACs) are cerebrospinal fluid collections between the two layers of the normal arachnoid membrane. Although they are often asymptomatic with a stationary course, eventual complications may occur. Herein, we report the case of a 9-year-old boy who developed bilateral papilloedema secondary to spontaneous rupture of an AC in the left middle cranial fossa. Although the papilloedema worsened during follow-up, his visual field remained bilaterally stable, supporting the expectant management and obviating the potential morbidity associated with neurosurgical intervention. This case report highlights the importance of a multidisciplinary approach to patients with secondary intracranial hypertension, including serial ophthalmological examinations, which provide a useful guide to surgical decision-making.

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Introduction

Arachnoid cysts (ACs) are cerebrospinal fluid collections between the two layers of the normal arachnoid membrane. They are usually congenital, resulting from meningeal maldevelopment and account for 1–2% of intracranial mass lesions.[1] Acquired ACs have also been described, associated with head trauma, intracranial inflammation and neurosurgery. ACs may occur in any site of the arachnoid layer, including the middle cranial fossa (the most common site), the suprasellar and quadrigeminal cisterns, the posterior fossa, the cerebellopontine angle, the cisterna magna, the cerebral convexities and the interhemispheric fissure.[2]

Often asymptomatic, ACs are a relatively common incidental finding on intracranial imaging of children. They tend to be stationary or even disappear spontaneously, justifying a watch-and-wait approach.[3,4] However, occasional complications may occur, including subdural haematomas, subdural hygromas, intracystic haemorrhage and, rarely, cyst rupture, leading to subdural effusion and secondary intracranial hypertension.[5,6] These complications may require treatment such as shunt surgery.

In this report, we describe a paediatric patient with self-limiting intracranial hypertension (IH) causing papilloedema due to rupture of an arachnoid cyst. Absence of visual impairment on serial automated perimetry exams allowed safe follow-up without medical or surgical intervention

Case report

A 9-year-old boy was admitted to the Neuro-ophthalmology service of the State University of Campinas (Brazil) for investigation of bilateral papilloedema associated with sudden onset of a severe headache. He had a recent history of hospitalisation for spontaneous rupture of a congenital AC in the left middle cranial fossa, leading to a subdural hygroma and IH (Figure 1). The hypothesis of persistent IH was considered.

On admission, his visual acuity was 20/20 in each eye with unremarkable eye surface findings and mild bilateral optic disc oedema (Frisén grade 2) on fundoscopy (Figure 2). Automated perimetry showed an increase in the size of the blind spots in both eyes and mild peripheral

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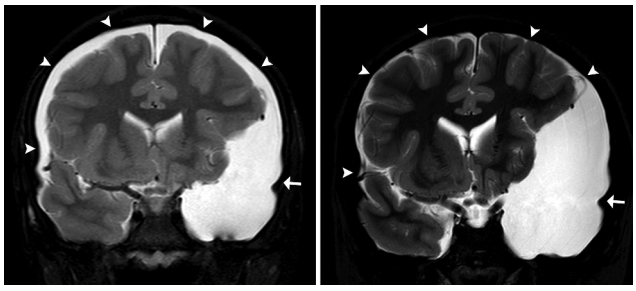

Figure 1. Neuro-imaging assessment. The left image shows T2-weighted coronal magnetic resonance imaging (MRI) at presentation, showing the arachnoid cyst in the left middle cranial fossa (white arrow), associated with a large subdural hygroma extending to the contralateral hemisphere (white arrowheads). On the right, subsequent T2-weighted coronal MRI after 1 year of follow-up showing a robust decrease of the subdural fluid collection (white arrowheads).

defects. When repeated 1 week later, no further progression was observed (Figure 3). Thus, he was submitted to close ophthalmological surveillance with regular perimetry and serial retinal photography. No medication was prescribed.

After 3 weeks, despite disappearance of the headache, a fundoscopic examination revealed worsening of the papilloedema, indicating the continuing presence of IH. Initially, a shunt was proposed. However, considering the potential surgical risk involved and, based on a prompt multidisciplinary evaluation by the neuro-ophthalmology and neurosurgery teams, a decision was made to maintain the expectant management approach, with perimetry and retinal photography repeated at 1 or 2-weekly intervals.

As an alternative to surgery, we considered prescribing acetazolamide to reduce intracranial pressure. However, we also considered the potential side effects of acetazolamide, especially metabolic acidosis, and the fact that our patient did not show worsening of visual function (visual acuity and visual fields). Therefore, we decided not to start acetazolamide and instead monitored him closely over a short period of time. The papilloedema progressively improved, the visual field remained stable over time, and the subdural collection spontaneously recovered (Figures 1–3).

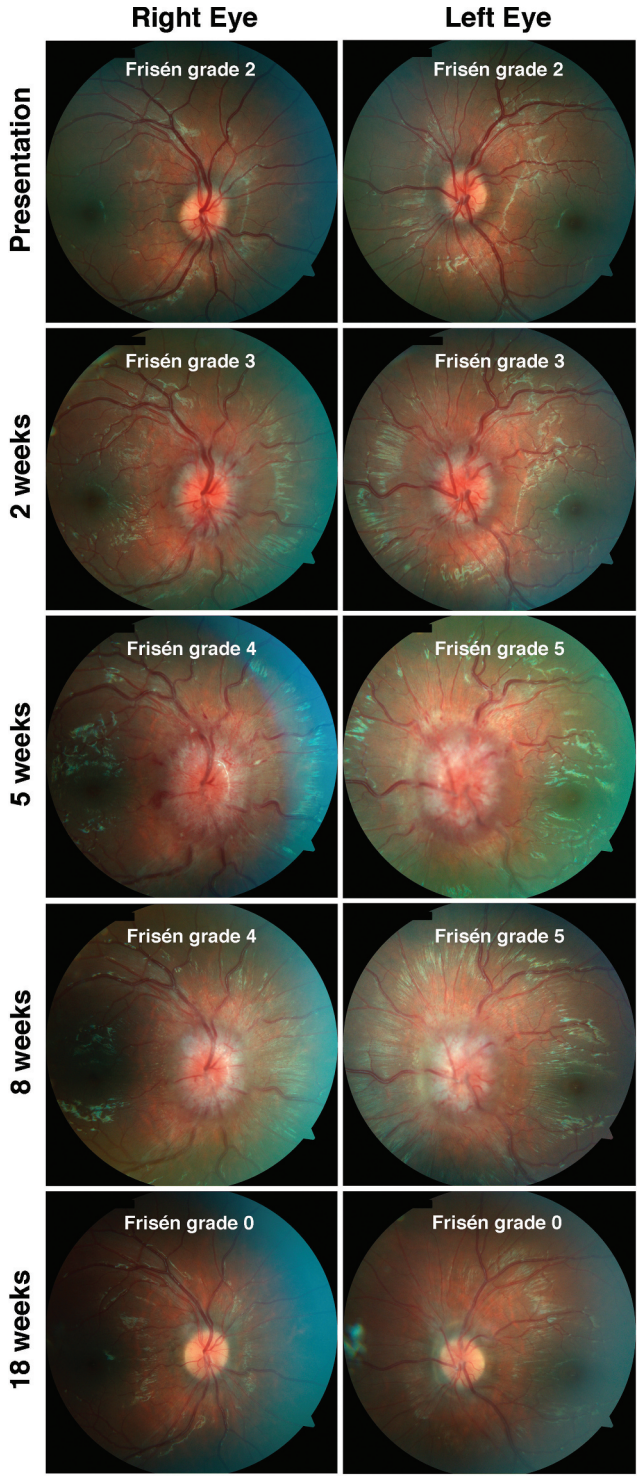

Figure 2. Series of coloured photographs of the optic nerve head showing mild papilloedema at presentation, that worsens over a few weeks, followed by spontaneous regression and normal appearances 18 weeks after presentation.

Discussion

The boy described in this report presented with asubtle intracranial pressure (ICP) increase after a left middle cranial fossa AC (the most prevalent lesion site) rupturing into the subdural space.[7] Although the worsening of the papilloedema suggested the persistence of IH (Figure 2), his overall

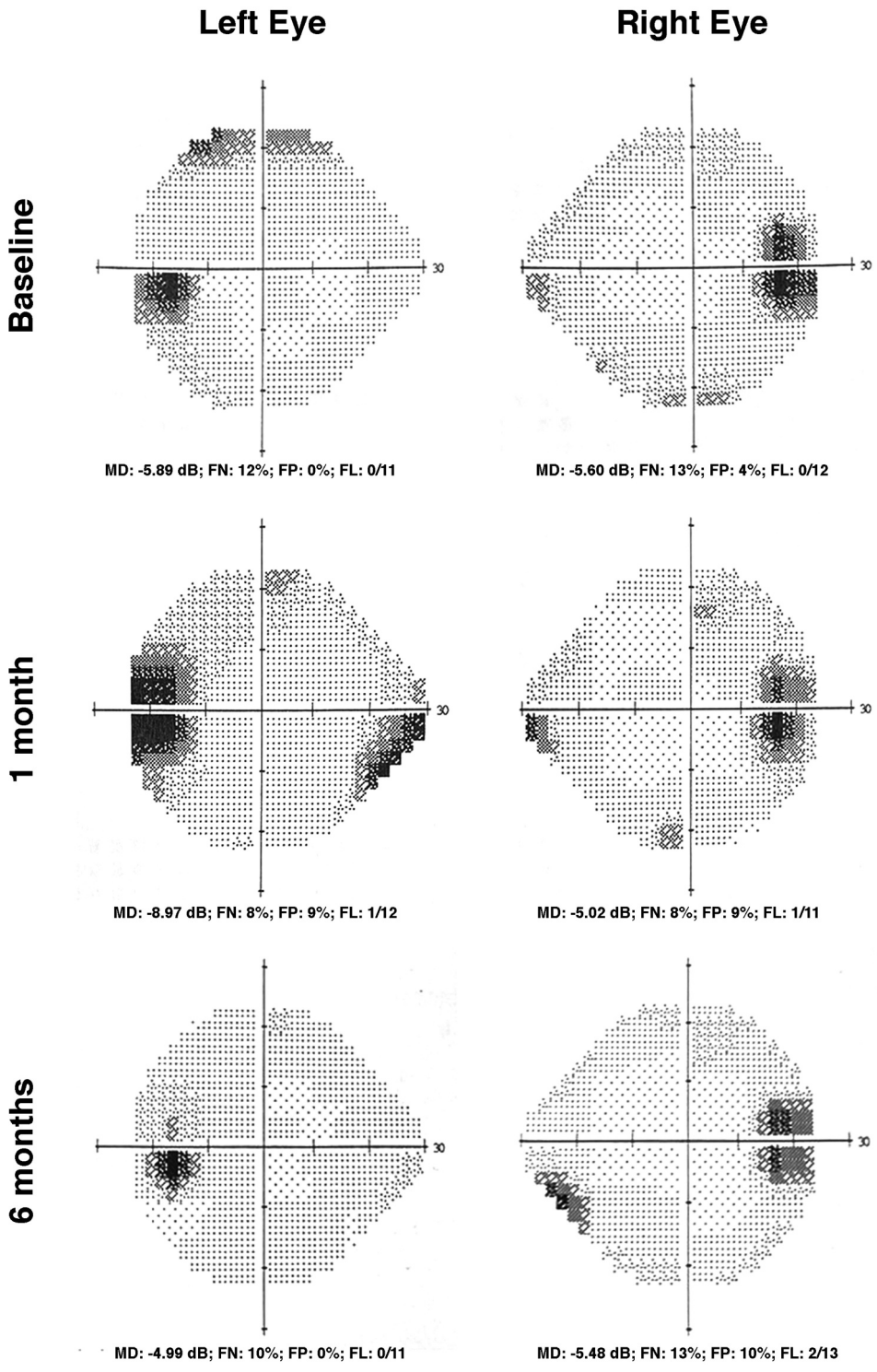

Figure 3. Visual field analysis. Automated perimetry was evaluated using the 24–2 Swedish interactive thresholding algorithm-fast strategy of the Humphrey® visual field analyzer (Carl Zeiss Meditec, Germany) showing stability or even mild improvement of the visual field throughout the follow-up period, supporting expectant management in this case. FL = fixation losses; FN = false negative; FP = false positive; MD = mean deviation.

good clinical condition, normal visual acuity and stable perimetry over time provided support for a conservative approach, avoiding the potential risk of surgical intervention.

The extra-axial fluid collection derived from a leaking or ruptured AC may be evacuated with different neurosurgical techniques, including endoscopic fenestration, craniotomy for fenestration, and

cystoperitoneal shunt placement. However, since all these techniques are associated with potential morbidity and occasionally require surgical revision,[8] surgery should be deferred for as long as possible, depending on disease progression.

In some cases, surgical intervention cannot be avoided. Deveney et al. recently described a 4-year-old boy who underwent expectant management with spontaneous resolution, whereas three other cases of varying ages required different modalities of neurosurgery.[9] In the latter cases, persistent papilloedema was the indication for intervention.

Serial ophthalmological monitoring is a useful guide for surgical decision-making in the management of patients with secondary IH. Nevertheless, the presence of papilloedema as the sole clinical sign of an optic neuropathy secondary to IH has been questioned by several authors in view of the well-documented lack of synchronicity between the dynamics of ICP and the appearance and resolution of papilloedema.[10] Thus, in patients with ruptured or leaking ACs, monitoring with serial visual field examinations should be adopted whenever possible, avoiding premature and, potentially, unnecessary interventions.

Standard automated perimetry may be unreliable in some children and so kinetic perimetry with, for example, the Goldmann visual field test, can be used as an alternative when available. Fortunately, our patient was able to perform automated visual field tests satisfactorily, making it a reliable way of monitoring his visual function.

Serial measurements of the thickness of the retinal ganglion cell/inner plexiform layers on optical coherence tomography can be used to assess neuronal integrity in patients with papilloedema, since loss of ganglion cell thickness correlates early with visual field defects.[11] In addition, IH causes anterior displacement of Bruch's membrane and therefore it can be used to monitor ICP levels.[12]

Disclosure statement

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