Case report

Management of Intrasellar Arachnoidocele

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ABSTRACT

Arachnoidocele in its sellar location is not common. The variability of its clinical signs and the lack of standard treatment make the management complex. The authors report a clinical case through which therapeutic modalities will be discussed. We report a case of a 42-year-old patient who complained of intermittent headaches of progressively increasing intensity over the past 2 years. The persistence of the headaches despite analgesic treatment had motivated the performance of a brain CT scan and then an MRI which had objectified the presence of an intrastellararachnoidocele. The endocrine laboratory function was normal. Short-term treatment with morphine and corticosteroids had achieved a rapid and long-lasting course after a 2-year follow-up. Endoscopic surgery is a treatment option for sellar arachnoidocele. The indication must take into account the clinical and radiological elements because some sellar arachnoidocele evolve favorably under drug treatment.

Keywords: Arachnoidocele, headache, neuroendoscopy, Turcic saddle, skull base.

INTRODUCTION

The sellar arachnoidocele is a collection of cerebrospinal fluid contained in the arachnoid leaflet at the level of the turcic saddle. It is one of the arachnoid cysts which are malformations related to a developmental abnormality in which splitting or duplication of the primary arachnoid leads to an intraarachnoid collection of cerebrospinal fluid.¹ Arachnoidocele was first
Intrasellar arachnoidocele is an extremely rare condition. Through clinical observation and data from the literature, the authors will discuss the different clinical presentations and discuss the therapeutic modalities of the sellar arachnoidocele.

CASE REPORT

The observation concerns a 42-year-old patient, an accounting officer, married and mother of two children. Her history includes an appendectomy 15 years ago; it has no connection to its current history. The patient complained of headaches that had been progressing for 2 years and gradually increasing in intensity. These headaches prompted the use of Dihydroergotamine. This painful symptomatology was associated with hot flashes without any particular schedule. There was no visual impairment or vomiting. Faced with these symptoms, she consulted a physician. After a normal clinical evaluation and laboratory assessment (complete blood count, C-reactive protein, blood glucose, urea, and creatinine), she was prescribed an analgesic treatment based on Tramadol. With this headache, she consulted an endocrinologist doctor where a brain-computed tomography (CT-scan) has been ordered. This revealed the presence of hypodensity similar to that of the cerebrospinal fluid which was located inside the turcic saddle. There were no bone wall abnormalities or other associated intracranial lesions (Figure 1A, 1B).

Brain magnetic resonance imaging (MRI) (Figure 2C, 2D) confirmed the intrasellar lesion that was hypointense in the T1 sequence without gadolinium and hyperintense in the T2 sequence. The diagnosis of arachnoidocele of the Turcic Saddle has been suggested.

The endocrine assessment carried out showed in the blood the prolactin (17 ng/mL); growth hormone (2.3 ng/mL); gonadotropic hormones (LH: 6.3IU/L; FSH: 5.9IU/L); thyroid hormone (ultrasensitive TSH 2.7 mIU/L) and in the urine, the urinary free cortisol 29ug/24h. This assessment has been interpreted without particularity. The patient was referred for neurosurgery. The presence of these headaches not relieved by level 1 and 2 analgesics with a strictly normal clinical examination required the initiation of morphine-based treatment 10 mg extended-release every 12 hours for 5 days combined with short-term corticosteroid therapy on 7 days with Methylprednisolone at a dose of 0.5 mg/kg administered in the morning. The course was marked by the regression of headaches and medication was given only in times of painful crisis. These seizures occurred once every 3 to 4 months but were controlled by...
the analgesic. Clinical monitoring was proposed with MRI in case of persistence or worsening of the symptom. After two years of follow-up, the headache attacks were spaced 6 to 8 months apart and the patient had resumed her activities.

**DISCUSSION**

Arachnoidocele is a herniated meninges containing cerebrospinal fluid (CSF). The intrasellar localization of this anomaly is very rare, in the order of 3%. It was first described in 1831 by BRIGHT. It corresponds to a hernia of the optochiasmatic cistern through a dehiscence of the saddle diaphragm resulting in an empty turcic saddle. Articles on this pathology are clinical cases or series of cases reported in the literature due to its rarity. As in our observation, this pathology is more reported in women. Exceptionally, intrasellar arachnoidocele can be seen in children during radiological assessment for another disease, as was the case of the 12-year-old patient in Silvia’s series. This condition is thought to be related to a developmental abnormality in which splitting or duplication of the primary arachnoid leads to an intraarachnoid collection of cerebrospinal fluid. The presence of a large opening of the sellar diaphragm or the absence of a diaphragm, and the difference between the volume of the pituitary gland and the turcic saddle are factors that favor the penetration of the arachnoid into the turcic saddle. It has been shown that the subarachnoid space can penetrate the saddle under normal conditions. The mechanism of cyst extension is not known. The clinical presentation is variable. As in our observation, headache is the main and earliest symptom. It is very frequently associated with visual disturbances. These, as well as the signs of endocrine damage that can be seen in the lesions in the saddle and the sellar region, were absent in our patient. When they are present, endocrine signs are mostly manifested by disorders of the gonadotrophic axis. These include menstrual cycle irregularities, infertility, and decreased libido. We think that the headache could be due to the distension of the sellar dura mater under the pressure of the cyst inside the turcic saddle. Weber et al, asserted a lack of a link between headache and the presence of cysts in some cases, arguing that some cases of arachnoidocele were incidentally discovered. In its intrasellar location, the cyst may extend above the pituitary fossa and manifest as optochiasmatic compression, a decrease in the function of the anterior pituitary gland associated with headache. Several clinical varieties exist. A case of arachnoidocele revealed by Sheehan’s syndrome with adrenocorticotropic and thyrotropic insufficiency has recently been reported. While in the vast majority of the cases described, the disease occurs alone; In very rare cases, it can be associated with other pathologies. This complicates management as in the case described by Cherkaoui and al. The diagnosis of arachnoidocele is radiological. Brain MRI is the gold standard. It makes it possible to evoke it formally, by specifying the density of the cyst’s contents and the presence or absence of peripheral contrast. The diagnosis is evoked in the presence of any cystic formation whose densities and signals appear, at all points, identical to those of the cerebrospinal fluid. Sometimes, there may be a contrast at the level of the cyst wall. This may be related to compression of the pituitary shaft and displacement of the normal pituitary gland, which may be elevated. In our observation, a brain CT scan was the first-line examination. In the presence of headaches without neurological signs of focus, CT is warranted. Also, because of its availability and accessibility, CT-scan is a good unscrambling test. In the presence of diagnostic doubt, as was the case in our observation, an MRI will be necessary. From a therapeutic point of view, because of the variability of clinical signs, management remains a matter of school. Conservative treatment is not codified. The
gradual use of analgesics in stages is difficult to apply in this context. Conservative treatment becomes complex in cases of endocrine disruption associated with the clinical signs. Under these conditions, management will become multidisciplinary involving endocrinologists. As for surgical treatment, according to some authors, it should be indicated in symptomatic patients or when the cyst is progressing.4 From our point of view, surgery should be performed in patients who have not been improved by drug treatment and/or when there are signs of radiological evolution of the lesion. Patients may respond favorably to conservative treatment. This was the case in our observation. There were two opposing tendencies in the realization of fenestration by the rhinoseptal route. These were the microsurgical approach and the endoscopic surgery. If surgeons of a certain era opted for microsurgery, it was mostly related to their habits. Nowadays, the endoscopic surgical approach is the most practiced because of its simplicity, its speed of performance, and the low morbidity associated with it. The current trend for the surgical treatment of lesions of the sellar region is the endoscopic rhinoseptal technique. Guinto et al. had used the endoscopic route for the surgical treatment of 3 patients. They proceeded with the bone remodeling technique of the Turcic Saddle with very good results.7 According to Raffael and al, there are no reports defining the best surgical treatment in terms of outcomes.19 A meta-analysis may remove any doubt about the best therapeutic choice to offer patients in terms of good outcomes. The subfrontal, pterional, transventricular, or transcallous upper approach would be relatively invasive; due to the depth of the cyst.4 It should certainly address the voluminous cyst with suprasellar extension. This procedure will be supplanted shortly by minimally invasive surgery by endoscopic approach.

CONCLUSION

Sellar arachnoidocele is a benign condition whose management is uncodified. Surgery is reserved for cases not improved by conservative treatment. The addition of endoscopy has reduced surgery-related morbidity and length of hospital stay.

REFERENCES


Additional Information

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Conflicts of Interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:
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