

A rare case of suprasellar arachnoid cyst with giant perimesencephalic and mesial temporal extension - physiopathological mechanisms

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Abstract: The arachnoid cyst is a lesion commonly encountered in neurosurgery, especially in pediatric pathology. We are presenting the case of an adult patient with a suprasellar arachnoid cyst with giant perimesencephalic and mesial temporal extension discovered incidentally, where there is a discrepancy between the spectacular neuroimaging and the non-specific symptomatology. Some of the physiopathological mechanisms which led to the evolution of the cyst will also be presented.

Key words: giant arachnoid cyst, sellar cyst, enlargement basal cisterns

Background

The arachnoid cyst (AC) is a congenital collection of cerebrospinal fluid (CSF) contained within the arachnoidal membrane and the subarachnoid space (21, 27, 28). The first case of AC to be described was located in the middle cranial fossa and was published for the first time in 1831 by the English physician Richard Bright (1789-1858) (24), best known for his studies on kidney diseases (Bright's albuminuric nephritis). Bright defined AC as “serous cysts forming in connection with the arachnoid and apparently lying between its layers” (7).

The incidence of AC ranges between 0.2 and 1.7% (11, 20, 22, 38, 37), but is continuously growing due to the increasing use of magnetic resonance imaging (MRI) and computed tomography (CT) (2, 37, 38). Moreover, some studies report an incidence of 2.6% (3).

Most arachnoid cysts (ACs) are diagnosed incidentally after imaging explorations such as CT or MRI after a mild head injury (26). The predominance is greater in males, with a male - female ratio of 2:1. The multiple or bilateral forms of AC are unusual, and some studies even report a familial occurrence (15, 19, 24, 39).

Case presentation

We are presenting the case of a male patient, aged 34 years old, who sought medical attention for a right hemicrania-like migraine, anxiety and neurotic syndrome, as well as subjective neurovegetative symptoms. Further to a head CT scan, a sharply demarcated, non-enhancing, extra-axial cyst, with CSF density, located at the level of the basal cisterns and the medial temporal lobe was observed. The exploration was completed by a MRI scan, which highlighted large and relatively symmetrical cystic lesions in the middle cranial fossa, on the medial aspect of temporal lobes which follow CSF signal on all sequences (Figure 1). Given the fact that the symptoms were non-specific and very few compared with the neuroimaging, a conservative treatment was chosen, consisting in clinical and imaging follow-up of the patient.

Discussions

As mentioned, most ACs, regardless of their location, are an incidental finding, usually after a brain imaging performed for non-specific symptoms, as in our case. Other non-specific symptoms encountered in patients with AC are mild headache, dizziness or balance impairment (14, 34). If the ACs become symptomatic, the symptoms occur especially in early childhood, as 60-90% of patients with ACs being children (9, 24).

Several AC occurrence mechanisms were proposed, among which head injury in childhood (8, 10), the splitting or duplication of the arachnoid membrane (25, 27-29, 33), as well as some genetic factors (1, 4, 6, 16-18, 23).

In our patient's case, neuroimaging did not highlight any signs of intracranial

hypertension with cerebral compression, and the absence of abnormal brain development made us consider the existence of cystic lesions with progressive development. Moreover, in MRI imaging, even though a slight volume reduction of both hippocampal areas (Figure 2) was observed, the patient had no cognitive deficits.

In terms of the physiopathological mechanisms of AC, some studies (12, 31) suggest that the membrane of Liliequist plays an important role in the development of the suprasellar arachnoid cyst. Normally, it is perforated and separates the incomplete interpeduncular and chiasmatic parts of the suprasellar cistern, stretching between its attachments points at the dorsum sellae, hypothalamus, ventral midbrain and oculomotor nerves (13). If this membrane is imperforated, either as a result of congenital neurodevelopment or of an acquired adhesive arachnoiditis, the CSF obstruction from infratentorial to supratentorial subarachnoid spaces (12) occurs. The continuous flow of CSF from the fourth ventricle through the subarachnoid space in the posterior side of the spinal cord and then in the anterior side will produce upward expansion of the membrane in the suprasellar cistern with the appearance of a diverticulum. The progressive dilatation of the suprasellar cyst will lead to a widening of the suprasellar cistern extending towards the basal cisterns (Figure 3) and further into the crural and carotidian cisterns, widening the subarachnoid spaces on the inferior and medial region of the temporal lobe (13). (Figure 1).

The occurrence of clinical symptoms is closely related to the intracystic pressure (17, 18). For this reason, in case of increased intra-

cystic pressure (e.g. hemorrhage, cyst expansion), the most common clinical symptoms are headache, seizures and dizziness (5). Moreover, perfusion studies showed that increased intra-cystic pressure will eventually compromise the function of the adjacent cortex by reducing blood perfusion and neuronal metabolism (5, 32, 35, 36).

The therapeutic options consist of monitoring the patient or surgical treatment: cyst excision, fenestration, endoscopic fenestration or stereotactic suction (9, 30). Many authors recommend AC follow-up in case there are no symptoms or imaging signs of intracranial hypertension, regardless of the size or location of the cyst (9, 30).

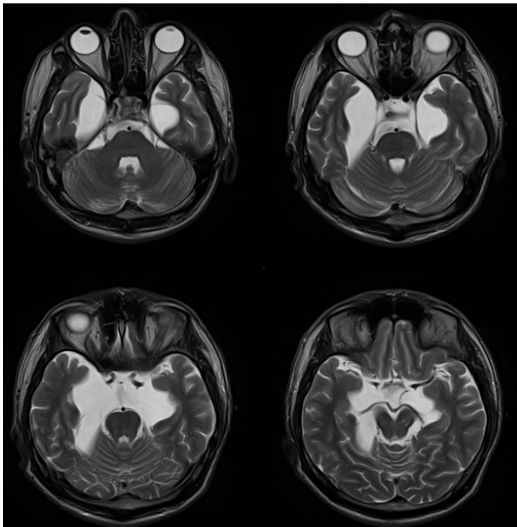


Figure 1 - The middle cranial fossa contains large and relatively symmetrical cystic lesions, on the medial aspect of temporal lobes; the lesions follow CSF signal on all sequences

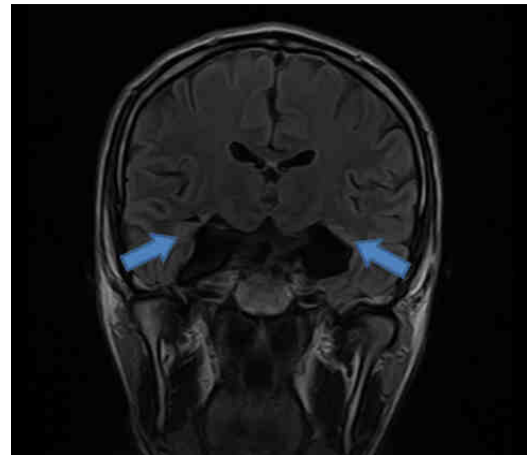


Figure 2 - There is a slight volume reduction of both hippocampal areas due to the mass effect of the cystic lesions (blue arrows)

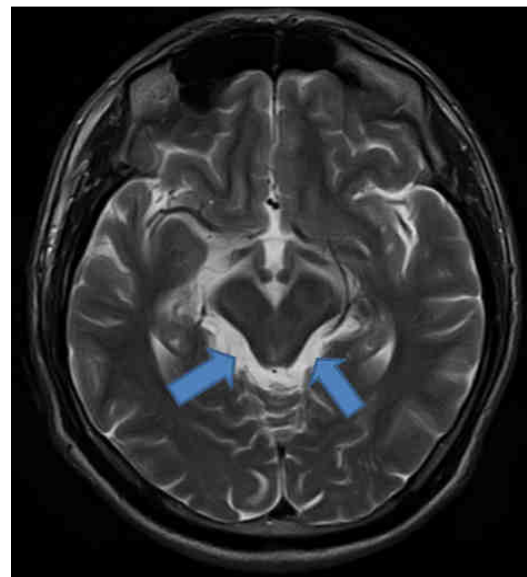


Figure 3 - Symmetrical enlargement of ambiens and quadrigeminal cisterns (blue arrows)

Conclusions

To the best of our knowledge, this case of AC with giant perimesencephalic and mesial temporal extension is the first of its kind to be reported in the literature, given its discrepancy between the spectacular neuroimaging and the nonspecific clinical image. In the absence of neurological symptoms, the clinical and imaging follow-up is recommended, due to the fact that an ill-timed surgery could produce an imbalance between the multiloculated spaces of AC maintaining an intracranial hypertension syndrome.

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