Arachnoid cyst of the posterior fossa

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ABSTRACT

يعد ظهور الأكياس العنكبوتية في الحفرة الدماغية الخلفية أمراً غير شائع. نستعرض في هذا المقال حالة سريرية لرجل عمره 49 عاماً، كان يشكو من الصداع، والغثيان والدوار وعدم الاستقرار في المشي لمدة شهرين. وقد كشف التصوير بالرنين المغناطيسي (MRI) عن وجود كيس وسطي مُحاط إحاطة تامة داخل الحفرة الدماغية الخلفية، وكانت إشارات هذا الكيس تحمل نفس الصفات التي تحملها إشارات السائل النخاعي، ولم يكن هذا الكيس متصلاً بالبطين الرابع. مكنت هذه العلامات من اقتراح فرضيتي الكيس العنكبوتي أو الكيس العداري. وقد نجحت العملية الجراحية المباشرة في استئصال غشاء الكيس كلياً وكانت النتائج جيدة.

Arachnoid cysts of the posterior fossa are uncommon. Our case of a 49-year-old man presented with a 2 month history of headaches, nausea, and vertigo associated with walking instability. An MRI revealed a median well-circumscribed cystic lesion of the posterior fossa, with similar signal characteristics to CSF, and without connection to the fourth ventricle. This aspect suggested either arachnoid or hydatid cysts. Direct open surgery was performed allowing complete removal of the cyst wall, with a good outcome.

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Arachnoid cysts are developmental collections of CSF-like fluid covered by arachnoidal epithelium. They supposedly result from congenital malformations that might change during the postnatal stage of life. Most arachnoid cysts are located in the middle cranial fossa;[1] and posterior fossa localizations are uncommon. They are usually located in the cerebello-pontine angle, the cerebellum, and the fourth ventricle.[1,2] Herein, we report a clinical case of arachnoid cyst in the posterior cerebral fossa, and we discuss the clinical features, radiological findings, therapeutic options, and differential diagnosis from other cysts of the posterior fossa.

Case Report. A 49-year-old man was in good health until a year ago when he started having headaches, nausea, and vertigo during the 2 months prior to admission. He also reported slight walking instability. The neurological examination showed bilateral dysmetria and truncal ataxia. Ophthalmologic examination revealed bilateral papilledema. No other cranial nerve deficit was identified, and the otolaryngeal examination was normal. The MRI revealed slight dilatation of the lateral and third ventricles associated with a cystic well demarcated midline lesion in the posterior fossa without contrast enhancement, presenting with the same signal characteristics as CSF, especially in diffusion imaging sequences. The cyst was not connected to the fourth ventricle (**Figure 1**). Based on this MRI aspect, the diagnosis of arachnoid cyst was discussed as well as a hydatid cyst despite the absence of recent contact with dog in the history of the patient. A posterior neurosurgical approach was achieved via a midline suboccipital craniectomy with the patient in a prone position. The dura matter was carefully opened, and a translucent, smooth thin-walled cyst was found (**Figure 2**). Both the outer and inner membrane was easily dissected and completely excised. The fourth ventricle and the cisterna magna were opened, and CSF flow between the fourth ventricle and subarachnoid spaces was restored. The histological examination of the excised cyst wall confirmed the diagnosis of arachnoid cyst. Four weeks post-operatively, the headaches, vertigo, and ataxia completely disappeared, and he resumed daily activities. After one year of follow-up, he remained asymptomatic.

Discussion. Arachnoid cysts are benign developmental entities localized within the arachnoid membrane, and are mostly asymptomatic.[3] They are estimated to count for approximately 1% of all

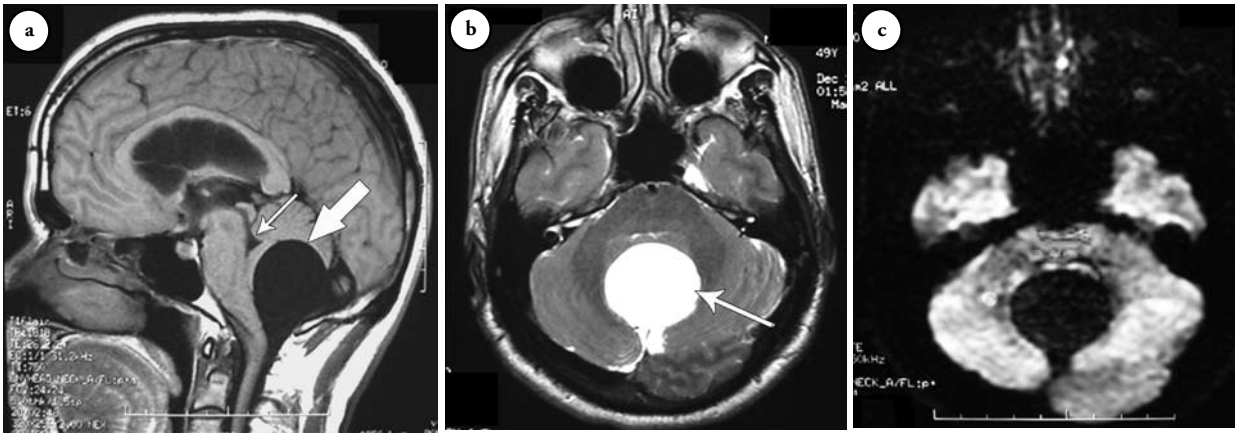

Figure 1 - Cerebral MRIs showing well-demarcated arachnoid cyst in the sagittal T1 image **a**) with contrast enhancement (thick arrow), **b**) axial T2 image (arrow) confirmed by the axial diffusion weighted image in **c**). A well-demarcated midline slight cyst of the posterior fossa, with the same signal characteristics as CSF (thick arrow) without connection to the fourth ventricle (thin arrow) is also demonstrated in the sagittal T1 image **a**).

Figure 2 - Operative view after opening of the dura showing a smooth thin-walled cyst containing a translucent liquid (arrow).

intracranial mass lesions.[4] Most arachnoid cysts are located in the middle cranial fossa.[1] Only rates of 5-10% of this type of cysts have been found to occur in the posterior cranial fossa.[5] There is no age predilection, and the male to female ratio is reported to be 3 for 1.1 The landmark of the arachnoid posterior fossa cyst is a variability of clinical signs and symptoms.[5,6] Posterior fossa arachnoid cysts are often unexpected, because they remain asymptomatic and might generate vague and non-specific symptoms such headache, dizziness, hearing loss, tinnitus, lower cranial nerve palsies, facial numbness, cerebellar and pyramidal signs, psychomotor retardation, seizures, and symptoms compatible with multiple sclerosis. No single symptom or group of symptoms would define posterior fossa arachnoid cysts. Hence, these lesions should be considered as a potential diagnosis when symptoms are longstanding, vague, fleeting or difficult to explain.[6]

The etiology of arachnoid cysts remains controversial, it is most probably congenital and the cyst supposedly results from splitting of the arachnoid membrane cells. However, acquired etiologies, such as inflammation and trauma have also been suggested. Several mechanisms could be responsible for gradual enlargement of an arachnoid cyst. This might include intracystic hemorrhage, an osmotic gradient allowing a passive fluid-diffusion into the cyst, a ball-valve mechanism, or an active secretion from the cyst wall.[2]

Magnetic resonance imaging (MRI) is the preferred method of investigation. Signal intensity is identical to that of CSF. The MRI is also supportive in differentiating between other cystic lesions of the posterior fossa. In fact, an arachnoid cyst can be differentiated from an epidermoid cyst by the abnormal higher signal intensity of the cyst in diffusion-weighted and FLAIR imaging acquisitions. The differential diagnoses includes the Dandy-Walker malformation, and its variant, mega cisterna magna,[2,3] and cerebellar hydatid cyst particularly in our context. The key features for distinguishing between these possibilities are the position of the fourth ventricle, the tentorium, the torcula, and its sinuses, the size of the posterior fossa, the confirmation of the vermis, and the presence of a communication between the cyst and the fourth ventricle. In our case, considering the normal position of the vermis and the torcula, the lack of communication between the cyst and the fourth ventricle, and the rostral deviation of the fourth ventricle on MR imaging, we retained the diagnoses of arachnoid and hydatid cysts of the posterior fossa, and the surgical precautions were taken. At surgery, the macroscopic aspect was in favor of an arachnoid cyst.

Various procedures of surgical treatment of the posterior fossa arachnoid cyst have been reported.[4,7,8] Cyst fenestration, stereotactic puncture, endoscopic cyst fenestration, cysto peritoneal shunt, cyst marsupialization into the subarachnoid space, and complete or partial resection of the cyst wall via a direct surgical approach was achieved. In our context, we believe that the direct approach is the surgical procedure of choice since a hydatid cyst can have similar radiological characteristics, and hydatidosis is still present in our country at an endemic level. The direct surgical approach allows complete resection of the arachnoid walls and communication of the cyst content with the subarachnoid spaces in order to prevent recurrences.

In conclusion, arachnoid cysts are benign cystic lesions. They usually occur in the supratentorial space and contain CSF. Their occurrence in the posterior fossa is rare. An MRI is the exam of choice, and complete cyst resection is the most reliable option for preventing recurrences. Long-term follow-up shows improvement in most of the preoperative manifestations and symptoms.

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