Dermoid cyst of the posterior fossa

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ABSTRACT

تمثل الأورام الجلدانية حوالي 0.1%–0.7% من مجموع أورام الجمجمة الداخلية، وتواجدها في الحفرة الخلفية يعد أمراً نادراً. نستعرض في هذا المقال حالة سريرية لمريض ذكر عمره 16 عاماً، وقد تم إدخاله إلى المستشفى بسبب ظهور علامات سريرية متعلقة بزيادة الضغط داخل الجمجمة وأعراض مخيخية. أظهر المسح بالأشعة المقطعية وجود كيس وسطي بالبطين الرابع داخل الحفرة الخلفية للدماغ تسبب في حدوث استسقاء ثلاثي في البطينات الدماغية. وأشار التصوير بالرنين المغناطيسي إلى وجود كيس وسطي مُحدد الشكل بقوة إشارة منخفضة مقارنة بالسائل النخاعي، ومحاط بغشاء تميز بارتفاع الإشارة بعد الحقن. خضع المريض لعملية فغر للبطين الثالث بالمنظار قبل الإزالة الجزئية للورم. لقد كانت نتائج العملية إيجابية ومن دون مضاعفات، كما أكد الفحص النسيجي وجود الكيس الجلداني. ومن خلال هذه الحالة السريرية، سوف نتطرق للعناصر السريرية، والجوانب الإشعاعية للأورام الكيسية الجلدانية في الحفرة الخلفية للدماغ، كما سنناقش الاستراتيجيات العلاجية.

Intracranial dermoid tumors represent a rare clinical entity accounting for 0.1-0.7% of all intracranial tumors. Their location in the posterior fossa is uncommon. We report a 16-year-old male patient who presented with clinical signs of increased intracranial pressure and cerebellar symptoms. The CT scan revealed a median cystic lesion of the fourth ventricle causing an active triventicular hydrocephalus. The MRI showed a median well shaped cystic lesion, of low signal intensity compared to the CSF, with capsular contrast enhancement. He underwent endoscopic third ventriculostomy before subtotal removal of the lesion. The postoperative course was uneventful, and the histological diagnosis was a dermoid cyst. Through this observation, we aim to discuss the clinical, and radiological aspects of the posterior fossa dermoid cyst, and to review the therapeutic strategies.

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Dermoid cysts are rare dysembryoplastic tumors accounting for 0.1-0.7% of all intracranial tumors.[1] They are mainly located in the sus-tentorial space, especially in the parasellar region. Their location in the posterior fossa remains uncommon.[2,3] Here, we report a case of dermoid cyst of the fourth ventricle diagnosed in a 16-year-old patient, and we discuss pathogenesis, clinical and radiologic features of this tumor, as well as treatment, and prognosis. Our objective in presenting this particular case is to highlight this rare localization of such tumors.

Case Report. A 16-year-old male, without significant medical history, was admitted to the neurosurgical emergency unit complaining of severe headaches, vomiting, diplopia, and moderate gait ataxia. Neurological examination revealed a conscious patient with statokinetic cerebellar syndrome associated with a bilateral horizontal nystagmus. The ophthalmological examination revealed a reduced visual acuity to 4/10 in the right eye, and 5/10 in the left eye with a bilateral stage III papillar edema at funduscopy. The CT scan revealed a median cystic lesion of the fourth ventricle causing an active triventicular hydrocephalus. Cerebral MRI showed a median well shaped cystic lesion, with a low signal intensity compared to the CSF and filling the fourth ventricular with a triventicular hydrocephalus (**Figure 1**). First, we performed an endoscopic third ventriculostomy to treat the hydrocephalus and to relieve the increased intracranial pressure. Then, the patient underwent a direct surgical approach through a median suboccipital craniectomy. At surgery, we discovered an intradural whitish-yellow soft lesion containing a hair with a typical macroscopic appearance demonstrating a dermoid cyst (**Figure 2**). Since the adherences were so close between the floor of the fourth ventricle and the tumor, the resection of the mass was subtotal. Histological study confirmed the diagnosis of dermoid cyst. The postoperative course was uneventful. He has been on follow-up for the last 2 years with no clinical or radiological evidence of recurrence.

Discussion. Dermoid cysts represent a rare clinical entity that accounts for 0.1-0.7% of all brain tumors,[1]

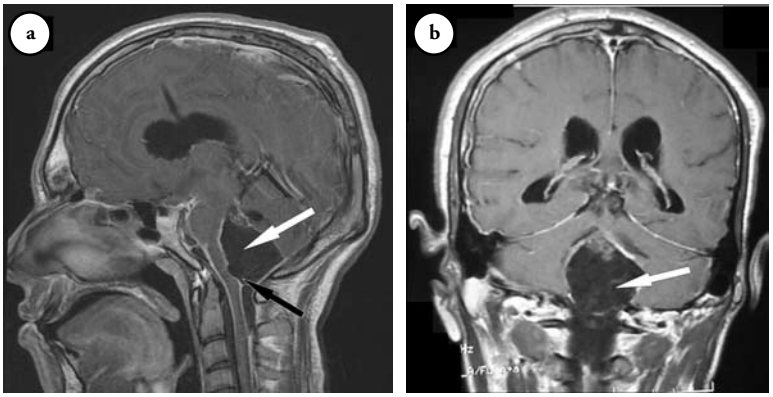

Figure 1 - Cerebral enhanced MRI in sagittal **a**) and coronal **b**) view showing a well defined midline cyst (white arrow) with discrete margin enhancement and cisternal extension (black arrow).

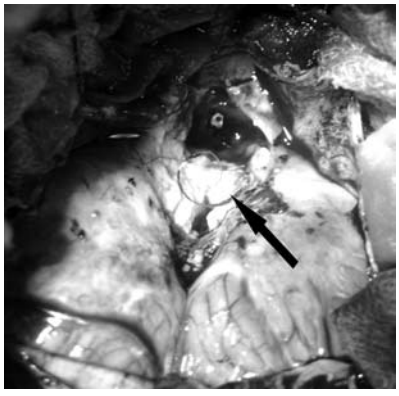

Figure 2 - Intraoperative view showing the dermoid tumor containing a hair (arrow).

and manifests mostly during early adolescence.[4,5] They are congenital benign tumors known to evolve gradually with slow growth as a result of progressive epithelial desquamation and gland secretion within the cyst. The cyst arises from inclusion of ectodermal elements within the neural tube during its closure between the third and fifth week of embryonic development.[6] Dermoid cysts are most commonly located in the cisternal spaces, mainly in the cerebellopontine angle and parasellar cisterns. Their location in the posterior fossa is uncommon.[2,3] Posterior fossa dermoid cysts are typically midline within the vermis, but can also occur in the fourth ventricle.[7] Lateral locations in the cerebellar hemispheres were also described.[8] The most remarkable characteristic of posterior fossa dermoid tumors in the posterior fossa is the their tendency to lie in the midline of the skull independently of their relationship to the intracranial structures, either outside or inside the dura mater. In the latter position, these tumors might expand asymmetrically over one or other cerebellar hemisphere, and the main bulk of the tumour overlies the vermis or penetrates the outlet of the fourth ventricle. Indeed, Logue and Till[2] classified posterior fossa dermoid cysts into 4 groups, depending on whether they are extradural or intradural, and on the degree of development of the dermal sinus, whether absent, partial, or complete: (1) extradural dermoid cyst with a complete dermal sinus; (2) intradural dermoid cyst (with no dermal sinus); (3) intradural dermoid cyst with an incomplete dermal sinus; and (4) intradural dermoid cyst with a complete dermal sinus. According to this classification, our case is of type 2.

Clinical manifestations of posterior fossa dermoid cysts are related to mass effect obstructive hydrocephalus, and raised intracranial pressure. Major symptoms and signs may include headache, nausea, vomiting, cerebellar signs such as ataxia and dysmetria, seizures, cranial nerve palsies (usually of nerves VI and VII), papilledema, bradycardia, and hypertension.[1,7,9-11] Aseptic meningitis secondary to the dissemination of the cyst content characterizes these tumors.[6,7,9] Radiologically, dermoid cysts are usually extremely hypodense on CT scan with a Hounsfield unit of -20 to -140, in keeping with their lipid content. Calcification is frequently present, and the tumor does not enhance after the administration of contrast medium.[11] Occasionally, they appear hyperdense mimicking a hemorrhage.[12] On MRI, they are typically hypointense on T1-weighted images and vary from hypo- to hyperintense, non-homogenous lesions on T2-weighted images. They typically have a high signal on FLAIR images and are moderately restricted on diffusion-weighted images.[13] The cyst may be round, oval, or bilocular, and varies from a few millimeters to several centimeters in diameter.

Posterior fossa dermoid tumors can be associated with other malformations such as Klippel-Feil syndrome, Dandy-Walker malformation, and agenesis of the corpus callosum.[4,5,7,9] None of these anomalies were found in our case. Pathologically, dermoid tumors are composed of a thick, stratified squamous epithelium cyst wall

containing dermal elements.[3] Macroscopically, the tumor has a yellow color and a calcified capsule. The content of the cyst comprises fat cells, sebaceous material, and cholesterol, with usually a number of long, coiled hairs lying separately or in clumps.[5]

The treatment of a posterior fossa dermoid cyst needs microsurgical excision.[1,3] Total removal of the tumor is preferred if the resection is ensured without damaging the adjacent structures. Nevertheless, it is not necessary to attempt complete removal of the cyst wall if it is firmly adherent to the brain structures. Careful bipolar coagulation is likely to prevent recurrence and reduce the risk of neurological impairment.[13] Extreme caution is advised when a dermal sinus is found to penetrate the occipital region, as a connection between the dermal sinus, dermoid cyst, and cranial venous confluence is possible, and unexpected penetration is associated with rapid and fatal exsanguinations. Mortality and morbidity increase if chemical or bacterial meningitis develops, or if there is associated cerebellar abscesses.[7] Otherwise, no recurrence after complete removal of the cyst has been reported in the literature.[1]

In conclusion, dermoid cysts of the posterior fossa are uncommon. They usually manifest by intracranial hypertension, cerebellar syndrome, and sometimes with meningitis. The MRI remains the key exam, and surgery remains the treatment of choice.

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CASE REPORTS

Case reports will only be considered for unusual topics that add something new to the literature. All Case Reports should include at least one figure. Written informed consent for publication must accompany any photograph in which the subject can be identified. Figures should be submitted with a 300 dpi resolution when submitting electronically or printed on high-contrast glossy paper when submitting print copies. The abstract should be unstructured, and the introductory section should always include the objective and reason why the author is presenting this particular case. References should be up to date, preferably not exceeding 15.