



Epidermoid cyst of the posterior fossa: A case report of a combined microscopic and endoscopic transcranial approach



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Epidermoid cyst of the posterior fossa: a case report of a combined microscopic and endoscopic transcranial approach

AIM: To show a case of a giant epidermoid cyst of the posterior fossa treated with a combined microscopic and endoscopic transcranial approach.

MATERIAL OF STUDY: A 13-years-old girl presented recurrent episodes of convulsions and vision impairment for years. CT scan and MRI revealed a suprasellar cystic lesion. The patient was operated with a combined microscopic and endoscopic transcranial approach.

RESULTS: Postoperative MRI demonstrated complete resection of the mass and pathology confirmed the preoperative suspicion of epidermoid cyst. Post procedure, the patient recovered well and was discharged. At the 4 week follow up her symptoms had resolved completely.

DISCUSSION: Epidermoid tumors are congenital lesions with a benign nature. They are the most frequent congenital intracranial lesions. Surgical treatment of epidermoid and dermoid cysts may be challenging. In the recent decades endoscopic endonasal approach (EEA) has been commonly used for the treatment of cranial base lesions because it has several advantages compared to open transcranial approach. We decided to do a transcranial approach combining the use of the microscope with the endoscope because EEA was not possible due to the location of the tumor.

CONCLUSIONS: Despite the development of innovative techniques such as the EEA, the transcranial approach remains a viable option, especially when more techniques are combined. In fact, the combination of microscope and endoscope grants the advantages of both tools being very useful to assure a complete excision of intracranial tumors.

KEY WORDS: Combined Transcranial Approach, Epidermoid Cyst, Posterior Fossa

Introduction

Epidermoid tumors are congenital lesions that arise from ectodermal cells misplaced during the closure of the neural tube in the process of embryogenesis. They have

a benign nature, hardly ever undergoing malignant transformation and are characterized by a white pearly capsule filled with lamellated keratin debris¹⁻³. The peak age of occurrence of epidermoid cyst is between the third and fourth decades of life, rare in the pediatric population, with a male to female ratio of 3:2. The incidence of epidermoid tumors is between 1% and 2% of all primary intracranial tumors. The cerebellopontine angle (CPA) is involved in 40% to 60% of the cases. Other reported locations are in the suprasellar cistern, prepontine cistern, temporal lobe and the pineal region¹⁻⁶. Pure suprasellar and sellar tumors are rare findings. Besides their insidious development and infiltration into vital neurovascular structures, they have friable consis-

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tency which particularly facilitates the surgical resection. The most common symptoms are visual disturbances, cranial nerve deficits and endocrine disorders. It is thus necessary to preserve cranial nerve functions and brain vasculature, what frequently determines the extent of the resection ⁷.

Here, we present a case of a 13-years-old girl with a giant epidermoid cyst of the posterior cranial fossa, extending to the suprasellar region that was treated with a combined microscopic and endoscopic transcranial approach.

Case Report

A 13-years-old girl was admitted to our hospital complaining recurrent episodes of convulsions under medical treatment and vision problems for years. Laboratory investigations were within normal limits. She underwent neuroimage investigation. A skull computed tomography (CT) revealed a suprasellar cystic lesion. In magnetic resonance (MR) this lesion was better described by a posterior fossa extra-axial cystic mass lesion, measuring 50x40x35 mm in diameter, filling the prepontine and the interpeduncular cisterns, extending to the floor of the third ventricle partially encasing the left side of the optic chiasm and the optic tract, abutting the pituitary stalk, with signal intensity isointense to CSF on T2-weighted images, heterogeneous on T1 weighted images and FLAIR, with no enhancement after contrast, suggesting an epidermoid cyst. The patient was operated with a combined microscopic transcranial approach and endoscopic transcranial approach.

A standard pterional craniotomy was done, drilling the great lateral sphenoid wing, then the dura was incised, splitting of the Sylvian fissure and after reaching the optic nerve the mass was identified. The resection was done initially using the microscope, advocating a sharp arachnoid dissection and a mass excision. The Liliequist

membrane was sharply cut to access the posterior fossa. From here we accessed the posterior fossa utilizing 0 and 45-degree endoscope to visualize and remove the rest of the tumor via endoscope-assisted aspiration.

Postoperative MRI demonstrated complete resection of the mass and pathology confirmed the preoperative suspicion of epidermoid cyst.

Post procedure, patient recovered well and was discharged. Her symptoms had resolved completely on her follow-up at 4 weeks (Figs. 1-3).

Discussion

Epidermoid cysts popularly known as “Tumeur perlees” (pearly tumors) were first described by Cruveilhier in 1835. They arise from rests of ectodermal cells misplaced during the division of the neuroectodermal and cutaneous ectoderm during the 3rd or 4th week of intrauterine development.

They are slow-growing, benign lesions however, they may rarely undergo malignant transformation. The mean age at presentation of these lesions is the fourth decade ^{8,9}.

The usual locations of epidermoid tumor are parasellar region and cerebellopontine angle. They are less commonly located in sylvian fissure, suprasellar region, cerebral and cerebellar hemispheres and lateral and fourth ventricles ^{1,4-6}.

Symptoms depend on their location and include hearing loss, dizziness, gait disturbance, trigeminal neuralgia, tinnitus, diplopia, visual impairment, apathy, headache, and gait ataxia ¹⁰.

Although epidermoid brain tumors can be visualized on computed tomography (CT) scans, they are best diagnosed by magnetic resonance imaging (MRI) ¹¹.

The surgical treatment of epidermoid and dermoid cysts may be challenging. Surgical resection with complete excision and preservation of the neural function should be the goal of their management. The surgical approach

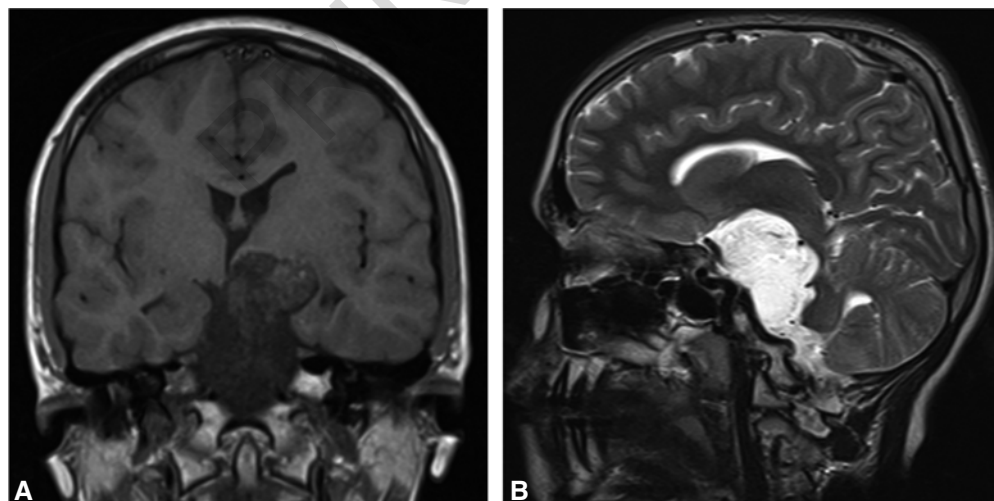


Fig. 1: (A) Preoperative T1-weighted coronal MR images; (B) T2-weighted sagittal images showing the epidermoid cystic lesion.

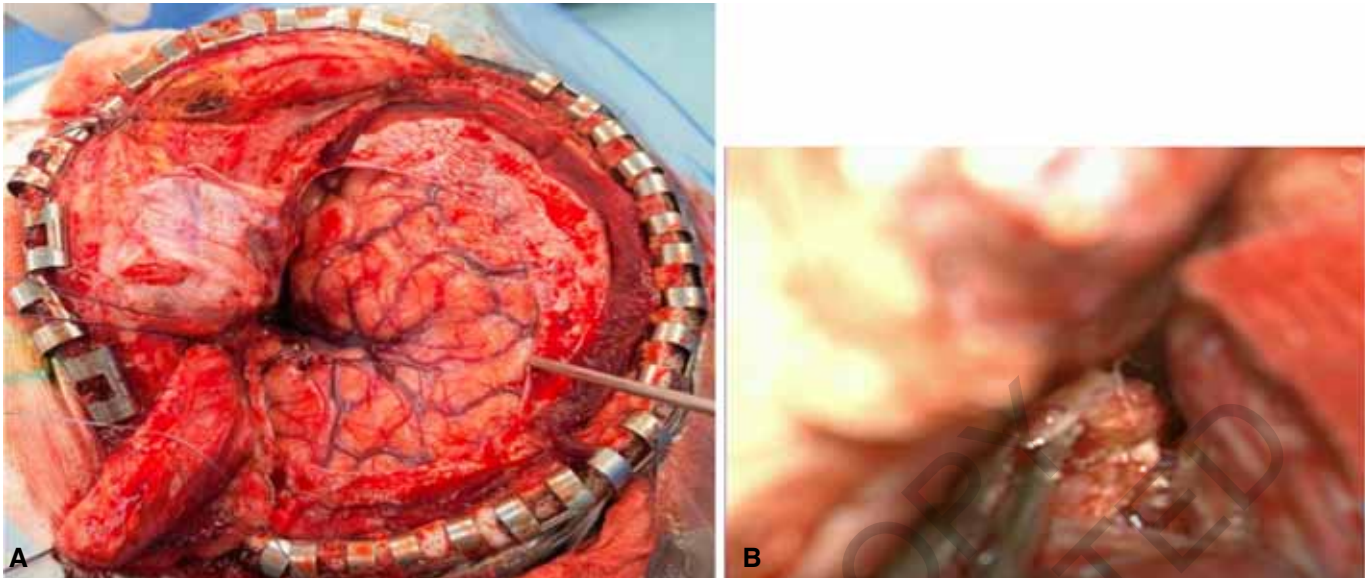


Fig. 2: Pterional transcranial approach: (A) intraoperative images showing access to reach the epidermoid cyst located in the suprasellar region. (B) Tumor resection using the microscope.

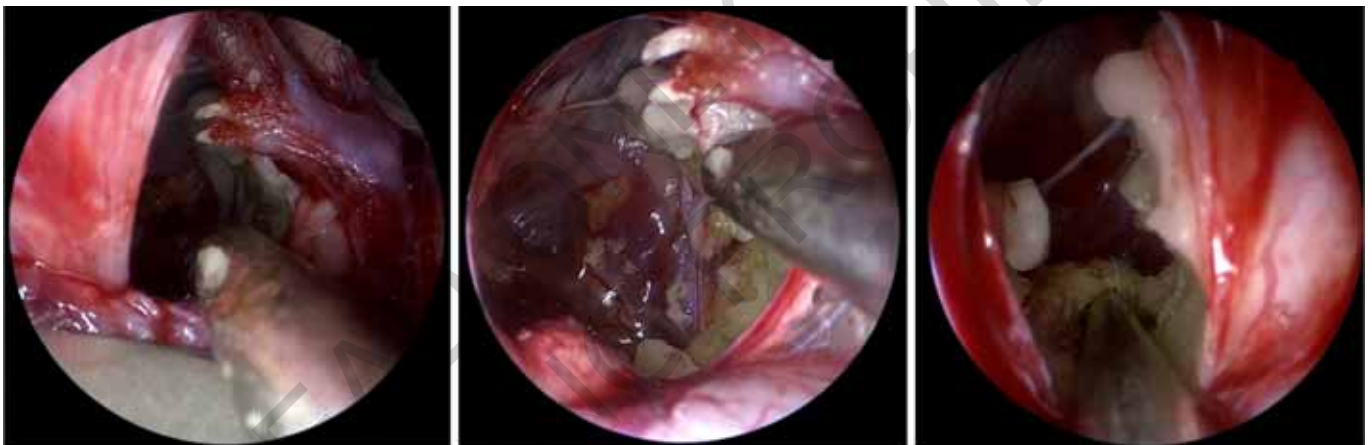


Fig. 3: Intraoperative images of the posterior fossa utilizing 0 and 45-degree endoscope to visualize and remove the rest of the tumor via endoscope-assisted aspiration.

depends upon the extent of the tumor, as determined by the preoperative diagnostic studies^{3,11-16}.

Supratentorial located tumors are usually excised by the pterional or frontotemporal approach, mesencephalic located tumors by either a supratentorial posterior interhemispheric transtentorial approach or an infratentorial/supracerebellar approach, and posterior fossa located tumors by either a medially or laterally positioned suboccipital craniotomy^{1,17}.

In the recent decades endoscopic endonasal approach (EEA) has been commonly used for the treatment of a myriad of cranial base lesions. The use of endoscopic endonasal approaches (EEAs) for resection of ventrally situated cysts (cysts in the frontonasal junction or suprasellar, interpeduncular, prepontine, or premedullary cisterns) has several advantages compared with open tran-

scranial approaches. The lack of brain, neural, or vascular retraction; excellent cosmetic results due to the avoidance of skin incisions; and the potential for shorter surgical time and length of hospitalization are among the most notable ones^{18,19}.

In our case, due to its sensitive location, endoscopic endonasal approach (EEA) was not considered since the lesion was behind the pituitary gland and stalk, and above the optic chiasm. In this case it can be difficult to reach the lesion without manipulating the pituitary gland using the endonasal endoscopic approach (EEA). For this reason we decided to do a transcranial approach combining the use of the microscope with the endoscope, which provides a clear and enlarged view of areas beyond the view of the microscope without additional brain retraction^{20,21,22}.

In fact the combination of microscopes and endoscopes utilizes the advantages of both techniques and can be very useful to assure a complete excision of intracranial tumors ²⁰.

Conclusions

Surgical treatment of suprasellar epidermoid cyst can be very challenging. Despite the development of innovative techniques such as the EEA, the transcranial approach remains a viable option.

This is one of the few documented cases of a suprasellar epidermoid cyst of the posterior fossa treated with a microscopic transcranial approach combined with an endoscope assisted aspiration. In fact endoscope can be very useful in adjunct to micro neurosurgical excision of intracranial tumors by enlarging the field of view allowing a complete resection of the tumor.

Riassunto

I tumori epidermoidi sono lesioni congenite che originano da cellule ectodermali. Hanno una natura benigna e presentano una capsula color bianco perla con all'interno frammenti di cheratina. I tumori epidermoidi rappresentano circa l'1-2% di tutti i tumori intracranici e si localizzano al livello dell'angolo pontocerebellare in circa il 40-60% dei casi mentre la localizzazione sellare e sovrasellare è più rara.

Il caso clinico portato in visione riguarda una ragazza di 13 anni che lamentava ricorrenti episodi di epilessia e problemi visivi da diversi anni. È stata sottoposta ad una Risonanza Magnetica Cerebrale che ha messo in evidenza una massa cistica in fossa cranica posteriore delle dimensioni di 5x4x3,5 cm che occupava le cisterne prepontina ed interpeduncolare e che si estendeva al pavimento del terzo ventricolo fino al chiasma ottico di sinistra e alla regione sellare.

La paziente è stata quindi sottoposta ad intervento chirurgico di rimozione della cisti mediante un approccio combinato: inizialmente attraverso una visione in microscopia è stata rimossa la parte più voluminosa della cisti e successivamente, attraverso visione endoscopica a 0 e 45 gradi accedendo in fossa cranica posteriore, è stato rimosso il resto del tumore. La Risonanza Magnetica post-operatoria confermava la completa rimozione della neoformazione.

Il trattamento delle cisti epidermoidi cerebrali, d'altronde, è sempre stata una sfida ardua per il chirurgo. Negli ultimi anni è stato sempre più utilizzato un approccio endoscopico nasale soprattutto per la resezione di tumori occupanti la giunzione frontonasale o le cisterne prepontina ed interpeduncolare.

In questo caso l'approccio endoscopico nasale non è stato considerato perché la cisti si localizzava dietro la

ghiandola pituitaria per cui è stato adottato un doppio approccio microscopico ed endoscopico transcranico. Quest'ultimo approccio in particolare, allargando il campo di visibilità, assicura una completa resezione del tumore.

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