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Case Report

Huge intraventricular epidermoid cyst: A case report ☆,☆☆

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ARTICLE INFO

Article history:
Received 7 January 2025
Revised 3 March 2025
Accepted 11 March 2025

Keywords:
Epidermoid cyst
Intraventricular
Neurosurgery

ABSTRACT

Epidermoid cysts, originating from ectopic embryonic epithelial cells, are a common type of benign intracranial tumor. However, intraventricular epidermoid cysts are rare, with those occupying the lateral ventricles being exceptionally uncommon. This report details the case of a 59-year-old male presenting with recurrent headaches over an 8-month period, accompanied by behavioral changes, memory impairment, and seizures. Magnetic resonance imaging of the brain revealed a large intraventricular mass consistent with an epidermoid cyst. The patient underwent subtotal resection via a transfrontal transventricular approach, resulting in good postoperative recovery.

Introduction

Epidermoid cysts are rare congenital lesions arising from ectodermal remnants during neural tube closure in embryogenesis [1]. These benign, slow-growing tumors constitute less than 2% of all intracranial neoplasms and exhibit a long, indolent course before becoming symptomatic [1,2]. While they most frequently localize in the cerebellopontine angle and sellar/parasellar regions, intraventricular epidermoid cysts are exceedingly rare, with only sporadic cases reported in the lateral ventricles [1].

The rarity of lateral ventricular epidermoids poses significant diagnostic and therapeutic challenges. This distinction is particularly relevant for neurosurgeons, as preoperative suspicion of an epidermoid cyst can influence surgical planning. Unlike solid neoplasms, epidermoid cysts require delicate microsurgical techniques to avoid spillage of cyst contents, which can lead to chemical meningitis and other complications [2,3].

Clinical presentations vary widely, ranging from incidental findings to symptoms caused by mass effect, including headaches, seizures, and cognitive or psychiatric disturbances. However, many lateral ventricular epidermoids remain asymptomatic due to the pliable nature of the cyst, which allows slow molding around adjacent neural structures [4–6].

This article presents the case of a 59-year-old male with an unusual clinical presentation of a large lateral intraventricular epidermoid cyst, emphasizing the diagnostic complexity and

☆ Competing Interests: The authors do not have any conflicts of interest to declare concerning this article.
☆☆ Acknowledgments: None.

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https://doi.org/10.1016/j.radcr.2025.03.046

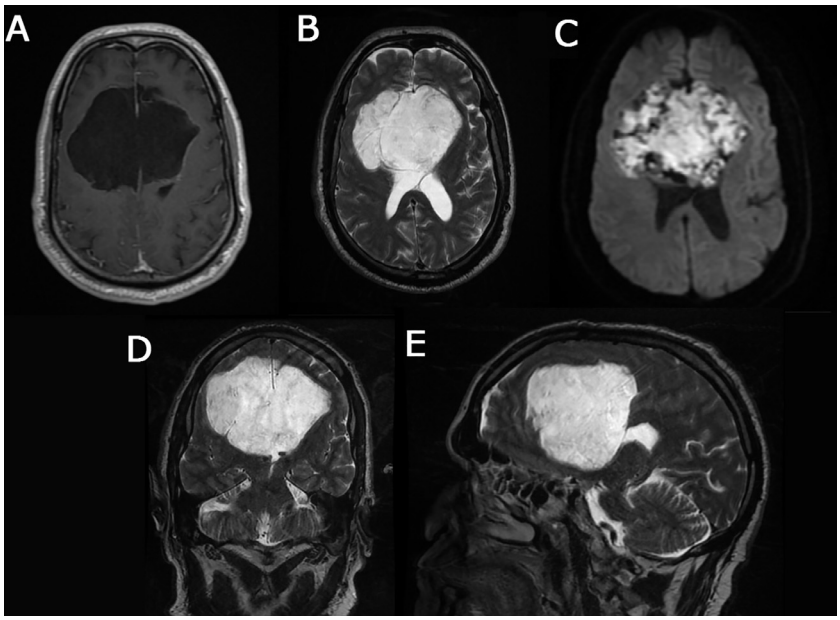

Fig. 1 – Axial (A, B and C), coronal (D) and sagittal (E) sections of a brain MRI in T1-WI with Gadolinium injection (A), T2-WI (B, D and E) and diffusion-WI (C) showing a large intraventricular mass which is hypointense on T1-WI, hyperintense on T2-WI. Diffusion MRI shows the typical appearance of a geographic map pattern suggestive of an epidermoid cyst.

surgical considerations of this rare entity. Through a comprehensive review of the literature, we aim to contextualize this case within the broader framework of intracranial epidermoid cysts, highlighting the role of advanced imaging, surgical techniques, and factors influencing prognosis.

Illustrative Case

A 59-year-old male patient with no significant medical history presented with an 8-month history of moderate, gradually progressive headaches, which he had neglected. These symptoms were followed by behavioral changes and forgetfulness, as observed by his family. The patient presented after experiencing a generalized tonic-clonic seizure. Neurological examination revealed no abnormalities.

Magnetic resonance imaging (MRI) of the brain (Fig. 1) revealed a mass occupying both lateral ventricles, with an appearance suggestive of an intraventricular lesion. The mass contained cystic material that appeared hypointense on T1-weighted sequences and heterogeneously hyperintense on T2-weighted and diffusion-weighted imaging. The lesion measured 9,5 × 7,6 × 7,1 cm, had a lobulated contour, and showed no contrast enhancement. It caused compression of the corpus callosum and thalami, with associated mild edema in the surrounding frontal cortex.

The patient underwent surgical resection of the mass. During the craniotomy, the brain appeared tense and swollen. An incision was made through the right middle frontal gyrus to access the wall of the right lateral ventricle. Upon opening the ependyma, the tumor was revealed at the same time as cerebrospinal fluid (CSF) escaped, confirming its intraventricular location. The mass was soft, avascular, and whitish with a pearl-like appearance, occupying the anterior horns of both lateral ventricles as the septum pellucidum was completely dehiscent (Fig. 2). It was tightly adherent to the ependymal walls, and the pericallosal arteries were embedded within the tumor folds, making complete excision challenging.

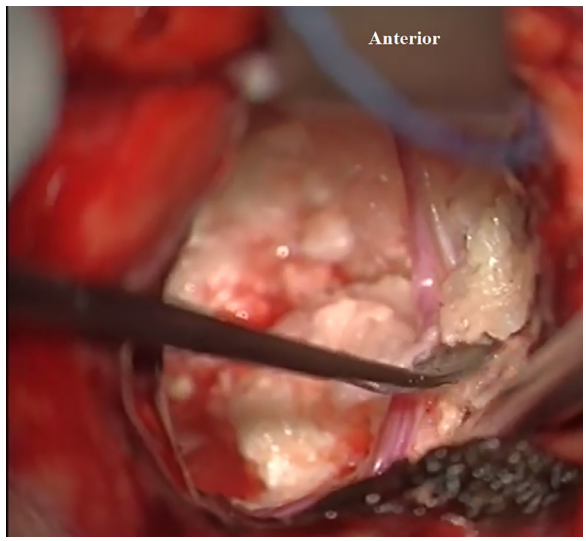

Fig. 2 – Intraoperative picture showing the epidermoid cyst and the pericallosal artery with a complete dehiscence of the septum pellucidum.

Microsurgical resection was performed, and the tumor was progressively debulked until the intraventricular foramina were exposed. The tumor had completely obstructed the

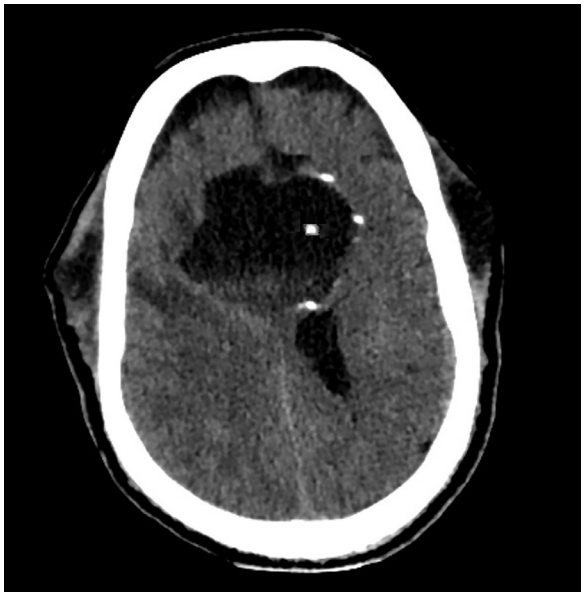

Fig. 3 – Axial section of a brain CT scan showing the remnant intraventricular cavity with the catheter of the external drainage system within it.

right foramen of Monro, which was successfully cleared, allowing unobstructed CSF flow. To prevent postoperative hydrocephalus, an external ventricular drain was placed at the conclusion of the procedure.

Postoperative period was uneventful. A contrast-enhanced computed tomography (CT) scan revealed near-total resection of the lesion (Fig. 3). The patient experienced a full neurological recovery, did not necessitate any further shunting (the external shunt was removed on the fourth postoperative day), and he was discharged on day 5 after surgery. Histopathological examination (Fig. 4) confirmed the diagnosis of an epidermoid cyst.

Discussion

Epidermoid cysts, also known as cholesteatomas or pearly tumors, arise from remnants of ectodermal cells during neural tube closure in the third to fifth week of fetal development [1,7,8]. These cysts are typically benign and account for 0.1% to 1.8% of all intracranial tumors [2,7]. Over two-thirds are localized to the cerebellopontine angle, followed by the cerebellopontine cistern, parasellar region, tentorium, and middle cranial fossa [6,8]. Intraventricular epidermoid cysts are extremely rare, with most cases reported in the third or fourth ventricles [3]. Localization within the lateral ventricles is extremely uncommon [1,2,6].

Patients with such lesions are typically in their 50s or 60s and are often asymptomatic [4,5,7]. This prolonged indolent course is attributed to the gradual expansion of the cyst into the lateral ventricle cavity and the smooth, pliable nature of the cyst wall and contents, which allows slow molding of adjacent neural structures. Additionally, cerebrospinal fluid (CSF) can circulate around the lesion, preventing hydrocephalus in most cases [4]. Larger cysts, however, can compress adjacent structures, leading to symptoms such as headaches and seizures, as well as cognitive deficits and psychiatric manifestations [5].

From a diagnostic standpoint, computed tomography (CT) scans, often the initial imaging modality, may be inconclusive because the density of epidermoid cysts is similar to that of CSF, leading to possible misdiagnosis as an arachnoid cyst. MRI is superior, as it clearly differentiates the cyst from adjacent CSF-filled cisterns or arachnoid cysts.

On T1-weighted MRI, epidermoid cysts typically appear hypointense, while they are hyperintense on T2-weighted images. Fluid-attenuated inversion recovery (FLAIR) images reveal a heterogeneous lesion with a hyperintense center. Diffusion-weighted imaging (DWI) shows hyperintensity, with corresponding hypointensity on apparent diffusion coefficient (ADC) maps, confirming characteristically restricted diffusion. This feature helps distinguish epidermoid cysts from arachnoid cysts, which do not exhibit restricted diffusion. MRI is the gold standard for evaluating epidermoid cysts and distinguishing them from other space-occupying lesions, including dermoid cysts, teratomas, intraventricular meningiomas, ependymomas, and choroid plexus papillomas [7,9].

Surgical resection is the primary treatment for intraventricular epidermoid cysts [7]. The surgical approach depends on the cyst's location within the lateral ventricle (atrium, temporal horn, etc.) [7,8]. Lesions in the anterior horn are typically accessed via a transcortical-transventricular approach, while caudal lesions are approached transcallosally. A subtemporal route may be used for cysts extending into the temporal horn or choroid fissure [8].

Total removal is the ideal treatment. Most epidermoid cysts are resected microscopically. However, in recent years, endoscopic techniques have gained traction due to their superior illumination and visualization, which can facilitate complete or adjunctive removal of the tumor [1,10]. Despite this, total resection can be challenging because of the cyst's adherence to critical neurovascular structures.

The extent of tumor resection may be limited by its extension, but neither the preoperative size of the cyst nor the degree of excision significantly impacts prognosis, which is primarily determined by the patient's preoperative neurological status. Even after subtotal resection, patients with preserved neurological function generally have favorable outcomes. Poor outcomes are more likely in older patients or those with prolonged delays between symptom onset and diagnosis [3].

Surgical principles for epidermoid cysts include decompressing the putty-like contents, excising the cyst membrane, preserving choroidal vessels and ependymal tissue, and avoiding subependymal injury. Contamination of the intraventricular cavity with cyst contents must be prevented, as spillage can cause chemical ventriculitis and hydrocephalus [8].

Aseptic meningitis is a well-recognized complication of epidermoid cyst surgery and can significantly affect outcomes. If it occurs, it can usually be managed with corticosteroids and repeated lumbar punctures. However, complete tumor removal remains the best strategy for preventing aseptic meningitis [7,8].

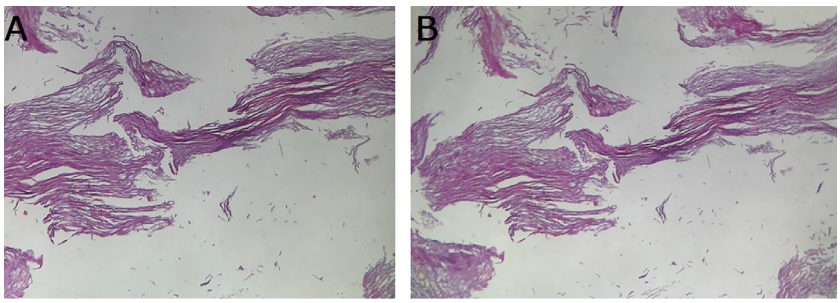

Fig. 4 – Histopathological sections of an epidermoid cyst showing concentric keratinized lamellae without nucleation (H&E stain; magnification × 100). Panel (A) illustrates homogeneous keratinized stratification, while Panel (B) highlights the dense lamellar organization characteristic of the lesion.

Conclusions

Intraventricular epidermoid cysts, though exceptionally rare, pose unique diagnostic and management challenges due to their slow growth and significant mass effect on adjacent neural structures. Early identification, especially in uncommon locations like the lateral ventricles, is crucial for timely intervention.

MRI plays a key role in distinguishing them from other cystic intracranial lesions, while Surgical resection remains the primary treatment. However, total excision can be difficult due to adherence to vital neurovascular structures. Notably, incomplete resection does not necessarily lead to poor outcomes, and patients often have favorable prognoses with appropriate surgical strategies.

Patient consent

A written consent has been obtained from the patient regarding this publication.

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