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Case Report

Petrous apex epidermoid cyst: A rare case ☆

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ABSTRACT

Epidermoid cysts are rare intracranial lesions comprising approximately 1% of all brain tumors, with petrous apex involvement accounting for 4%-9% of cases. These congenital lesions arise from ectodermal remnants during neural tube closure, while acquired cases may result from trauma or chronic middle ear pathology. Clinical presentation is variable and depends on the lesion's location and impact on surrounding neurovascular structures, with cranial nerve dysfunction being the most common symptom. Imaging plays a crucial role in diagnosis, with diffusion-weighted MRI distinguishing epidermoid cysts from other lesions such as arachnoid cysts and cholesterol granulomas. Management remains challenging due to their proximity to critical structures; complete surgical excision minimizes recurrence but may increase morbidity, while subtotal resection requires long-term follow-up. We report the case of a 40-year-old female patient who presented with a history of progressive hearing loss and facial paralysis, in whom an epidermoid cyst of the petrous apex was diagnosed.

Introduction

Epidermoid cysts are rare intracranial tumors, comprising 1% of cases, with 4%-9% occurring in the petrous apex. These lesions, histologically identical to middle ear cholesteatomas, arise either from congenital ectodermal remnants during neural tube closure or as acquired lesions eroding medially through the otic capsule. Their clinical presentation varies, commonly involving cranial nerve dysfunction due to their proximity to critical neurovascular structures. Management is complex, as complete surgical excision minimizes recurrence but poses risks due to dense adherence to vital tissues. Subtotal excision may reduce morbidity, though it requires careful long-term monitoring for recurrence [1,2].

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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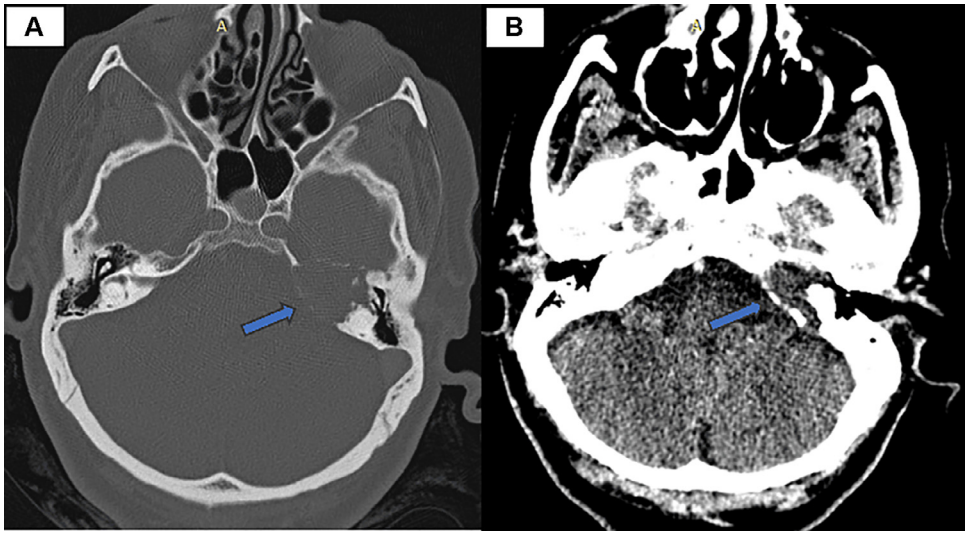

Fig. 1 – (A) Axial CT scan (Bone window) showing an expansile soft tissue mass the left petrous apex (Blue arrow). (B) Axial CT scan (Soft tissue window) Showing a hypodense soft tissue mass of the left petrous apex with no enhancement after contrast administration (Blue arrow).

Case report

We report the case of a 40-year-old woman with no prior medical history of recurrent otitis, who presented with progressive left-sided hearing loss, tinnitus, and left facial paralysis persisting over a five-year period without seeking medical consultation.

A computed tomography (CT) scan revealed an expansile, hypodense, soft tissue mass at the left petrous apex with scalloped bony edges and no contrast enhancement (Fig. 1). Magnetic resonance imaging (MRI) further characterized the lesion as an expansile mass at the left petrous apex, demonstrating T1 hypointensity, T2 hyperintensity, and mild peripheral enhancement with diffusion restriction. The mass exerted a slight mass effect on the ipsilateral temporal lobe and the left Meckel's cave. These imaging findings were consistent with a diagnosis of an epidermoid cyst of the left petrous apex (Figs. 2, 3, and 4).

The patient was referred to an otorhinolaryngology specialist at another facility for treatment management. The patient declined the proposed surgical intervention due to apprehension regarding potential complications, despite comprehensive counseling on its benefits and risks. However, no updates on her clinical progress are currently available.

Discussion

Epidermoid cysts are rare congenital intracranial lesions, accounting for approximately 1% of all brain tumors. These cysts develop from ectodermal remnants trapped during neural tube closure between the third and fifth week of gestation. While the cerebellopontine angle (CPA) is the most common site for intracranial epidermoids—affecting nearly 40% of cases—they can also occur in less common locations such as the mastoid, intradiploic bone, zygomatic root, and petrous apex. Primary epidermoids of the petrous apex are particularly rare, comprising only 4%-9% of all petrous apex lesions. Histologically, these lesions are indistinguishable from middle ear cholesteatomas, with their location medial to the otic capsule being the primary differentiating feature. This shared histology underscores their similar etiopathogenesis, with congenital cysts arising from epithelial rests and acquired ones typically originating in the tympanomastoid region, eroding medially into the petrous apex [1].

Clinically, the presentation of epidermoid cysts is highly variable and depends on the lesion's size, location, and impact on adjacent neurovascular structures. Cranial nerve dysfunction is the most common symptom, particularly involving the vestibulocochlear nerve (CN VIII), trigeminal nerve (CN V), and facial nerve (CN VII). Additional presentations may include headaches, cerebellar dysfunction, hydrocephalus, or, less commonly, aseptic meningitis due to irritation caused by cyst contents. Patients with petrous apex epidermoids may present differently from those with CPA lesions, as the location impacts which cranial nerves are predominantly affected. Hearing loss and facial nerve dysfunction are more frequently observed in petrous apex lesions, while trigeminal neuralgia and neuropathy are more characteristic of CPA lesions [1,2].

The diagnosis of epidermoid cysts relies heavily on imaging. On CT scans, these lesions often appear as well-defined, hypodense masses, occasionally with calcification of the cyst wall. MRI provides superior diagnostic accuracy, revealing low signal intensity on T1-weighted images and high intensity on T2-weighted images. Diffusion-weighted imaging is particu-

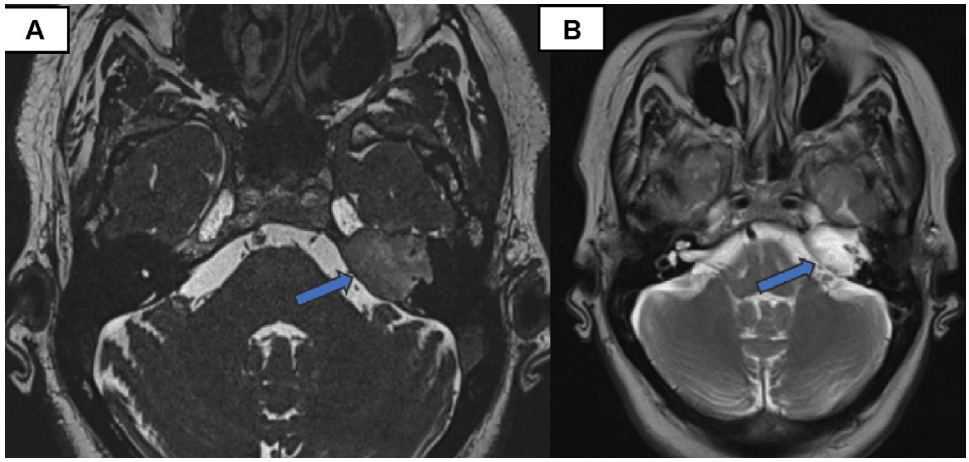

Fig. 2 – (A) MRI 3D CISS sequence showing an intermediate signal mass of the left petrous apex (Blue arrow). (b) MRI Axial T2 showing a mass of the left petrous apex exhibiting a high signal (Blue arrow).

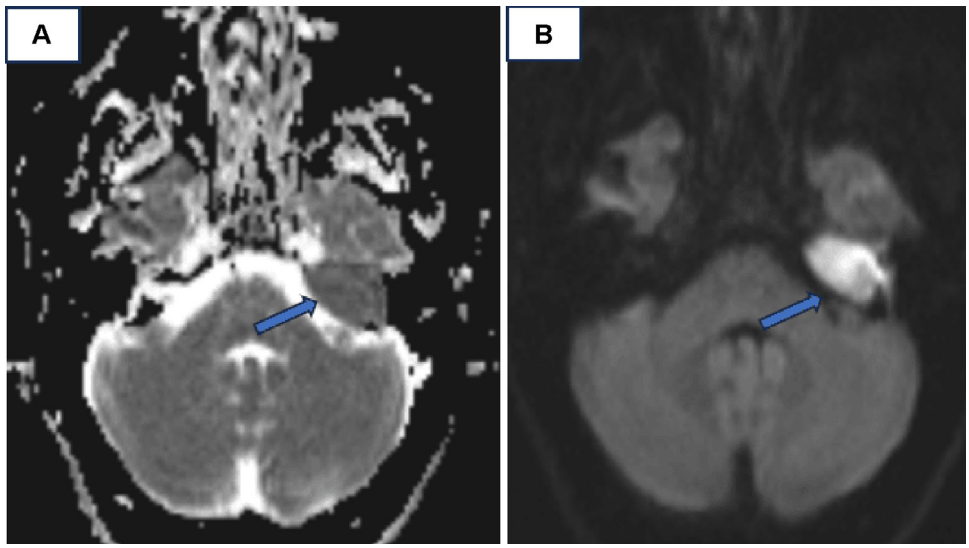

Fig. 3 – (A) DWI sequence showing a hyperintensity of the mass (Blue arrow). (B) ADC cartography showing restriction (Blue arrow).

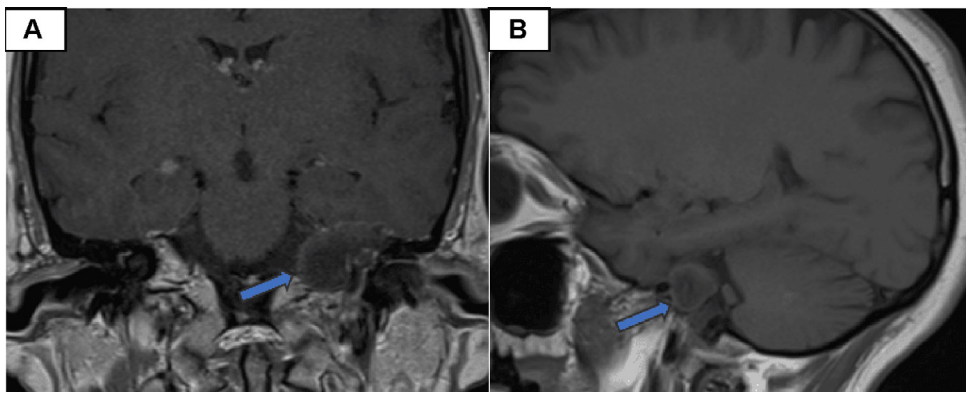

Fig. 4 – (A) Coronal T1 post Gadolinium administration showing a discreet peripheral enhancement. (B) No contrast Sagittal T1 showing a hyposignal mass of the left petrous apex.

larly useful in distinguishing epidermoid cysts from other cystic lesions, such as arachnoid cysts and dermoids, due to the restricted diffusion characteristic of their keratinous content [2,3].

The differential diagnoses for petrous apex epidermoid cysts include a variety of cystic and solid lesions with overlapping clinical and imaging features. Key differentials include cholesterol granulomas, arachnoid cysts, and dermoid cysts, which lack restricted diffusion on diffusion-weighted imaging (DWI), unlike epidermoids. Other potential diagnoses include schwannomas, meningiomas, and paragangliomas, which often show contrast enhancement, and bone pathologies like fibrous dysplasia or metastatic lesions. DWI is crucial in distinguishing epidermoid cysts due to their characteristic restricted diffusion, while clinical presentation and additional imaging features, such as vascularity or enhancement, help refine the diagnosis. Accurate differentiation requires a combination of imaging findings, clinical correlation, and occasionally biopsy or surgical exploration [3].

Management of petrous apex epidermoids remains complex due to their deep location and close proximity to critical neurovascular structures, including the internal carotid artery and cranial nerves. Surgical excision is the primary treatment modality, but there is no consensus on the extent of resection. Complete excision reduces the risk of recurrence but is often challenging due to the lesion's adherence to vital structures. Subtotal excision, while safer, carries a higher risk of recurrence and requires long-term follow-up. Advances in microsurgical and endoscopic techniques have improved outcomes by minimizing morbidity while allowing for effective lesion removal [4].

In rare cases, epidermoid cysts may exhibit unusual behavior, such as rapid growth, secondary infection, or complications like abscess formation. Trauma, including penetrating injuries, has been implicated in some cases as a trigger for cyst development or exacerbation. These unusual presentations necessitate a tailored approach, including surgical intervention and management of secondary complications. Overall, while epidermoid cysts are benign and slow-growing, their potential to cause significant neurological impairment and surgical challenges underscores the importance of early diagnosis and individualized management strategies [4].

Conclusion

Epidermoid cysts, particularly those in the petrous apex, are rare lesions that can cause cranial nerve dysfunction. Imaging plays a crucial role in diagnosis, with MRI being the primary tool. Imaging helps guide management, as these cysts are challenging to excise due to their location near vital structures.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Patient consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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