ECRONICON OPEN ACCESS

EC NEUROLOGY
Case Report

Intraclival Cholesteatoma or Epidermoid Cyst: A Rare Female Case Report

Abourak Chaimae*, Guennouni Asmae, Bahha Soukaina, Ech-Cherif El Kettani Najoua, Fikri Meriem, Jiddane Mohamed and Touarsa Firdaous

Department of Neuro-Radiology, Faculty of Medicine and Pharmacy of Rabat, Specialty Hospital, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco

***Corresponding Author:** Abourak Chaimae, Department of Neuro-Radiology, Faculty of Medicine and Pharmacy of Rabat, Specialty Hospital, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco.

Received: March 12, 2025; **Published:** March 27, 2025

Abstract

Introduction: Intracranial epidermoid cysts, also known as cholesteatomas or benign epithelial cysts, represent an uncommon subset of congenital lesions, marked by their slow-growing and non-malignant characteristics. These cysts are most commonly located in the cerebello-pontine angle cistern, which corresponds to their typical site of presentation. However, the appearance of such cysts within the intra-clival region, particularly along the midline, is exceedingly rare and deviates from the usual anatomical locations.

Case Report: We report a rare case of an intra-clival epidermoid cyst in a young woman in her third decade of life. The cyst's unusual localization within the clivus-a deep and less readily accessible region-highlights the unique nature and diagnostic challenge of this presentation. This case underlines the uncommon nature of epidermoid cysts and stresses the importance of maintaining a wide differential diagnosis for lesions in atypical anatomical sites.

Conclusion: This case holds significance not only due to its rarity but also because of its implications for clinical practice and diagnostic imaging. The intra-clival location of the cyst demonstrates that cholesteatomas may present in unexpected areas, expanding the differential diagnosis spectrum. Given the complexity of evaluating deep-seated regions, advanced imaging modalities, particularly magnetic resonance imaging (MRI), are essential for accurate identification and characterization of such lesions. This case emphasizes the critical role of comprehensive imaging in the diagnostic process and contributes to a broader understanding of the diverse manifestations of epidermoid cysts.

Keywords: *Medicine; Clivus; Cholesteatoma; Epidermoid Cyst; MRI*

Introduction

The clivus, originating from the Latin term meaning "slope" or "hill," is a key anatomical structure centrally located at the skull base [1-3]. It is also referred to as the Clivus of Blumenbach, named after the German physiologist and anthropologist Johann Friedrich Blumenbach (1752-1840) [1]. This structure is formed by the union of the occipital and sphenoid bones at the spheno-occipital synchondrosis, resulting in a wedge-like formation extending from the anterior foramen magnum to the dorsum sellae [1-3]. The clivus inclines forward and upward from the front of the foramen magnum to the posterior clinoid processes [2], measuring around 4 to 5.5 cm in length and approximately 3 cm wide at its center, with an average angle of 116° [2]. Due to its proximity to critical neurovascular elements, such as the brainstem and basilar artery, the clivus holds considerable importance in neurosurgical and radiological practices [2,4,5].

Anatomical variations of the clivus are highly relevant in clinical contexts. Certain studies point out the presence of rare structures, like the fossa navicularis magna, which can complicate surgical interventions and impact outcomes [6-8].

These variations may also be involved in rare conditions, such as transclival cerebrospinal fluid leaks, which further complicate surgical strategies [9].

Epidermoid cysts are benign, slow-growing congenital formations that result from ectodermal inclusions during the process of neural tube closure. They account for 0.5% to 1.8% of primary intracranial brain tumors [4-6]. These lesions, also known as congenital cholesteatomas due to their cholesterol-rich content, differ from acquired cholesteatomas, which arise from chronic middle ear infections and retraction pockets in the tympanic membrane's pars flaccida [4-6]. Histologically, epidermoid cysts are covered by stratified squamous epithelium and contain keratinous material, contributing to their distinct appearance on imaging modalities like MRI and CT scans [11-13].

The cerebellopontine angle cistern is the most common site for epidermoid cysts, accounting for 40% - 50% of cases [4]. Other frequent locations include the fourth ventricle (17%), the sellar and/or parasellar areas (10% - 15%), and, less commonly, the cerebral hemispheres or brainstem [4,5,6]. About 10% of cases involve extradural epidermoid cysts located in the skull or spine [4-6]. The clinical symptoms vary according to the cyst's location but typically involve compression of surrounding neural structures, leading to manifestations like cranial nerve deficits, headaches, or ataxia [4-6].

An epidermoid cyst within the clivus is an extremely rare occurrence, presenting a significant diagnostic and therapeutic challenge. A review of the available literature indicates the distinctiveness of these lesions, with only one case of an intra-clival epidermoid cyst reported by Fassett., *et al*. in 2006 [15]. This exceptional rarity highlights the necessity of recognizing atypical presentations of generally common lesions and underscores the importance of thorough imaging and meticulous surgical planning in such complex cases [10-12].

Case Report

A 39-year-old woman with an unremarkable medical history presented with chronic diffuse headaches, her only reported symptom. A comprehensive clinical examination, including an extensive neurological assessment, was normal, and her laboratory tests showed no abnormalities.

To further investigate her symptoms, a brain CT scan was performed, revealing a well-defined intra-clival lesion of liquid density (5 HU), with regular contours, as seen in figure A1-A3. The lesion showed no enhancement following contrast administration. A cerebral MRI was subsequently performed for more detailed imaging. The MRI revealed a cystic intra-clival formation exhibiting hypointensity on T1-weighted images, hyperintensity on T2-weighted images, and a heterogeneous hyperintensity on FLAIR sequences. The lesion also demonstrated diffusion restriction with a low ADC value and measured 15x21x10 mm (HxTxAP). Based on these imaging findings, an intra-clival epidermoid cyst was diagnosed, as further detailed in figure B, C1, C2, D1, D2, and E.

Discussion

Understanding the pathologies of the clival region, particularly the occurrence of epidermoid cysts, requires a solid grasp of clival embryology. Both mesodermal and ectodermal layers contribute to the development of the skull base, including the clivus [2].

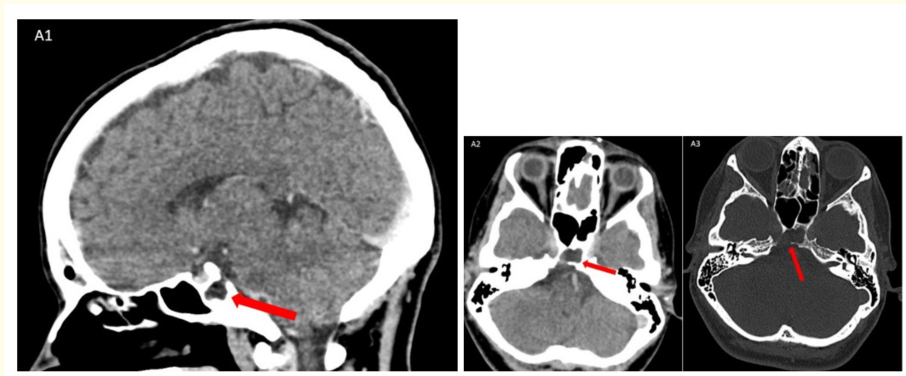

Figure A1-A3: *Brain CT with contrast injection (A1) sagittal plan and (A2) axial plan, in parenchymate window: intra-clival lesion of liquid density, not enhanced after injection of contrast agent (red arrow). (A3) axial plan, bone window: the expansive lytic lesion, responsible for an intra-clival gap with smooth edges (red arrow).*

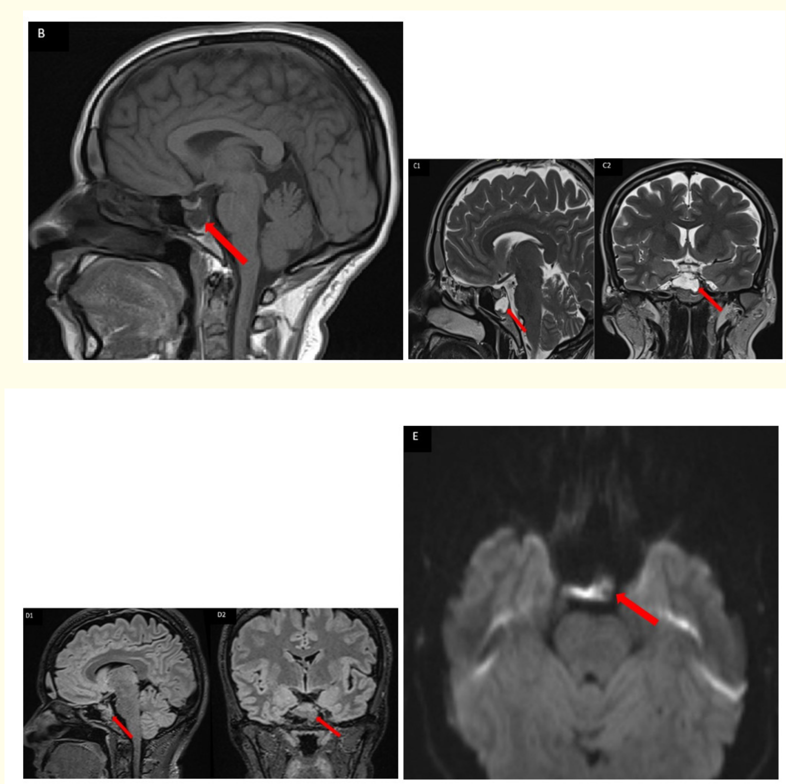

Figure B, C1, C2, D1, D2, and E: *MRI of the brain (B) Sagittal plan T1: intra-clival hyposignal lesion. (C1 and C2) sagittal and coronal plans T2: hyper-signal T2 lesion (D1 and D2) sagittal and coronal plans FLAIR: heterogeneous lesion. (E) Axial plan diffusion (DWI): restriction of diffusion (Red arrows).*

Epidermoid cysts originate from ectodermal inclusions during neural tube closure, which takes place between the third and fifth weeks of embryogenesis [11,13,16]. These cysts are defined by a thin wall of squamous epithelium, rarely containing calcifications. Microscopically, the cyst's lining consists of stratified squamous epithelium, surrounded by an outer collagenous connective tissue layer. The contents of the cyst include desquamated debris from the squamous epithelium, primarily composed of keratin and cholesterol arranged in lamellar patterns. Owing to their appearance, epidermoids are often described as "naked tumors" due to their shiny, waxy surface resembling "dry keratin" upon inspection [11,13].

Cholesteatomas, or epidermoid cysts within the central nervous system, can be found both intradurally and extradurally [11,13,15]. Intradural cysts typically occur in the cerebellopontine angle cistern, whereas extradural ones are frequently located in the petrous bone, either as congenital or acquired lesions from chronic ear infections. Other extradural sites include the skull's diploic layer, though a clival cholesteatoma has only been described once in the literature [11,13,15]. Proposing an acquired origin for a clival cholesteatoma remains speculative [15].

Epidermoid cysts are slow-growing and tend to manifest symptomatically between the third and fifth decades of life [11,13,16] as seen in our 39-year-old patient. Some studies suggest a slight male predominance [11], while others report no gender preference [16].

The growth dynamics of epidermoid cysts are comparable to those of skin cells, shedding within a closed cavity. As keratin, cholesterol, and exfoliated cells accumulate, the lesion enlarges, causing symptoms from either mass effect or rupture, leading to headaches or seizures. Diplopia may arise from compression of the abducens nerve in Dorello's canal, located at the posterior clival surface [11,13,15].

In some cases, rupture of an epidermoid cyst can trigger granulomatous meningitis due to the inflammatory effects of cholesterol-rich debris [11,13]. In our patient, the epidermoid cyst had likely been symptomatic for some time, presenting as chronic headaches.

Clinical presentation is typically non-specific, making imaging essential for diagnosis. The advent of MRI, along with modern imaging sequences, allows for preoperative diagnosis of epidermoid cysts in nearly all cases [16].

In this case, the intraosseous epidermoid cyst appeared as a hypodense lesion on brain CT, with limited enhancement after contrast administration. It was associated with expansive bone erosion, characterized by smooth or sclerotic edges [11,13,16]. Calcifications or a parietal bulge are present in 10% to 25% of cases [11,13,16].

Most epidermoid cysts are either isointense or slightly hyperintense relative to CSF on T1- and T2-weighted MRI images. They show incomplete suppression on FLAIR images, where they appear heterogeneous and hyperintense compared to CSF, with restricted diffusion on DWI. These findings were observed in our patient. Epidermoid cysts typically do not enhance after gadolinium administration, though mild peripheral enhancement is occasionally seen [11,13,16].

Rare "white epidermoids," containing high protein levels, may appear hyperdense on CT. Unlike typical epidermoid cysts, these "dense" epidermoids show reversed signal characteristics on MRI [13].

FLAIR and diffusion-weighted imaging (DWI) are vital for distinguishing an intra-clival epidermoid cyst from other cystic lesions, such as dermoid cysts (which show fat suppression), arachnoid cysts, neurenteric cysts, *Ecchordosis physaliphora*, sphenoid sinus mucoceles, neurocysticercosis, and cystic tumors (Table 1) [2-4].

Treatment typically involves observation and surgical decompression when indicated. Given their benign nature, there is no role for chemotherapy or radiotherapy in managing epidermoid cysts. When possible, extensive local excision is advised to prevent recurrence. In cases where complete excision is not feasible, marsupialization may be considered [7].

	MRI
Dermoid cyst	Dermoid cysts are typically located along the midline and resemble fat, not CSF.
Arachnoid cyst	Arachnoid cysts are isointense to CSF at all sequences, including FLAIR. They displace rather than invade structures such as the epidermoid. Finally, arachnoid cysts do not restrict on diffusion-weighted images.
Neurenteric cyst	sharply demarcated cystic mass with usually low intensity on T1-weighted and high intensity on T2-weighted MR images.
Ecchordosis physaliphora	It is located in the retroclival region and projects toward the intradural prepontine cistern. The tissue appears hypotense on T1-weighted MRI and hypertense on T2-weighted MRI with a lack of contrast enhancement.
Mucocele of the sphenoid sinus	Signal intensities at MRI depending on their protein content and presence of associated infection. Rim enhancement may be seen as a marker of encapsulation.
Cyst of neurocysticercose	Neurocysticercosis cysts often enhance and demonstrate surrounding edema or gliosis.
Cystic tumors	Cystic neoplasms often enhance and do not resemble CSF.

Table 1: *MRI characteristics of the main differential diagnostics of an epidermoid cyst of the clivus.*

Conclusion

This case demonstrates the possibility of an epidermoid cyst arising in rare locations like the clivus, a complex structure capable of hosting various uncommon lesions. Therefore, epidermoid cysts should be included in the differential diagnosis when assessing cystic lesions in this area. The diagnosis of an intra-clival epidermoid cyst poses significant clinical challenges, where imaging, particularly MRI, is essential for both accurate identification and long-term monitoring.

This study has several strengths. First, it provides a comprehensive analysis of intra-clival epidermoid cysts, incorporating various imaging techniques to differentiate them from other cystic lesions. The findings are clinically relevant, enhancing the understanding of rare cystic formations and aiding in accurate diagnosis and treatment planning. Additionally, the inclusion of multiple case studies allows for a broader perspective on the variations and presentations of epidermoid cysts, thereby contributing to the existing literature. However, the study also has limitations. It may be constrained by a small sample size, affecting the generalizability of the findings to a broader population. As a retrospective study, it is subject to biases in data collection and may not account for all relevant variables influencing outcomes. Furthermore, the reliance on specific imaging modalities might overlook other diagnostic methods, potentially limiting the understanding of differential diagnoses.

Acknowledgment

We thank our patient for his cooperation and willingness to participate in physician education.

Conflict of Interest

The authors have no conflicts of interest related to special relationships during the data collection and writing of this research manuscript.

Funding Support

This case report was not supported by any specific grant.

Author Contribution

C.A, I.B, S.B and A.G. contributed to the conception, acquisition, analysis, interpretation of data, and drafted the manuscript.

N.E, M.F, F.T, and M.J critically revised the manuscript and approved it.

Bibliography

1. Murjani B., *et al*. "Anatomical variations of clivus: a descriptive anatomical study". *Surgical and Radiologic Anatomy* 43.6 (2021): 945-951.
2. Rai R., *et al*. "A comprehensive review of the clivus: anatomy, embryology, variants, pathology, and surgical approaches". *Child's Nervous System* 34.8 (2018): 1451-1458.
3. Hofmann E and Prescher A. "The clivus anatomy, normal variants, and imaging pathology". *Clinical Neuroradiology* 22.2 (2012): 123-139.
4. Akkoca Kaplan F., *et al*. "Incidence of anomalous canals in the base of the skull: a retrospective radio-anatomical study using cone-beam computed tomography". *Surgical and Radiologic Anatomy* 42.2 (2020): 171-177.
5. Altafulla JJ., *et al*. "Transclival venous circulation: anatomic study". *World Neurosurgery* 121 (2019): e136-e139.
6. Bayrak S., *et al*. "Prevalence of anatomical variants in the clivus: fossa navicularis magna, canalis basilaris medianus, and craniopharyngeal canal". *Surgical and Radiologic Anatomy* 41.4 (2019): 477-483.
7. Kaplan FA., *et al*. "Evaluation of the relationship between age and gender of fossa navicularis magna with cone-beam computed tomography in orthodontic subpopulation". *Journal of the Anatomical Society of India* 68.3 (2019): 201-204.
8. Khairy S., *et al*. "Canalis basalis medianus with cerebrospinal fluid leak: rare presentation and literature review". *British Journal of Neurosurgery* 33.4 (2019): 432-433.
9. Vinayagamani S., *et al*. "Bipartite craniopharyngeal canal with a lipoma and cephalocele: a previously unreported entity". *Acta Neurochirurgica (Wien)* 161.2 (2019): 355-359.
10. Ray B., *et al*. "Morphological variations in the basioccipital region of the South Indian skull". *Nepal Journal of Medical Sciences* 3.2 (2015): 124-128.
11. Smirniotopoulos JG and Chiechi MV. "Teratomas, dermoids, and epidermoids of the head and neck". *Radiographics* 15.6 (1995): 1437-1455.
12. Fisahn C., *et al*. "Adult ligament of the dens lacks notochordal tissue: application to better understanding the origins of skull base chordomas". *World Neurosurgery* 101 (2017): 42-46.
13. Osborn AG and Preece MT. "Intracranial cysts: Radiologic-pathologic correlation and imaging approach". *Radiology* 239.3 (2006): 650-664.

14. Kasim N., *et al*. "Craniopharyngeal canal, morning glory disc anomaly, and hypopituitarism: what do they have in common?" *Oxford Medical Case Reports* 6 (2018): omy018.

15. Fassett DR., *et al*. "Cholesteatoma of the Clivus". *Skull Base* 16.1 (2006): 39-44.

16. Bhat DI., *et al*. "Interhemispheric epidermoids - An uncommon lesion in an uncommon location: A report of 15 cases". *Neurology India* 59.1 (2011): 82-85.

17. Neelakantan A and Rana AK. "Benign and malignant diseases of the clivus". *Clinical Radiology* 69.12 (2014): e533-e540.

18. Alalade AF., *et al*. "Fossa navicularis in a pediatric patient: anatomical skull base variant with clinical implications". *Journal of Neurosurgery: Pediatrics* 22.5 (2018): 523-527.

19. Tucker A., *et al*. "Neurenteric cyst of the lower clivus". *Neurosurgery* 66.1 (2010): E224-E225.

Volume 17 Issue 4 April 2025