# Cholesteatoma of the Clivus

## Case Report

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**Abstract**

*Objective:* Cholesteatomas (CNS epidermoids) can be found intradurally or extradurally in the central nervous system. Extradural intraosseous lesions are most commonly found in the petrous bone.

*Design:* Case report

*Clinical Presentation:* The authors describe a unique case of a clival cholesteatoma occurring in a 64-year-old woman who presented with headaches. No other neurological complaints or physical examination findings were noted. Magnetic resonance imaging showed an expansile lesion centered in the middle portion of clivus with erosion of a large portion of the clivus.

*Results:* This lesion was explored via a transnasal transsphenoidal approach and granular debris was evacuated. The cystic lining was stripped from the surrounding bone and the bone opening was widely fenestrated. Pathological examination showed keratinous debris with macrophages and an outer lining of benign epithelial tissue consistent with a cholesteatoma (epidermoid cyst).

*Conclusions:* The authors review the radiographic, surgical, and pathological findings of this unique case.

**Running Title**: Cholesteatoma of the clivus

**Key Words**: cholesteatoma, clivus, epidermoid



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## Introduction

Cholesteatoma, also known as epidermoid, epidermoid cyst, or benign epithelial cyst, is a tumor-like mass of keratinized desquamated debris filling an epithelial-lined cyst. The squamous epithelium lining the cyst continuously sheds dead keratinized epithelial cells that gradually build up, causing the tumor to grow with a linear growth rate. The term cholesteatoma is most commonly used to describe these tumors when they occur in the middle ear, but others have used a broader definition to include all epidermoid cysts occurring in the central nervous system.[1,2] When the tumors are intradural, they most commonly occur in the posterior fossa at the cerebellopontine angle. Extradural tumors are most commonly found in the petrous bone, where they may be congenital or acquired in association with chronic ear infections.[1,3] Other common extradural locations include the diploic layer of the calvarium, but cholesteatoma has never been reported in midline clivus.

### *Case Report*

A 64-year-old woman presented to her primary physician with complaints of headache over the preceding 5 days. She reported no other neurological symptoms and physical examination was normal. A computed tomography scan was performed; it showed a lesion in the clivus, which prompted neurosurgical consultation and a magnetic resonance imaging (MRI) scan. The MRI scan showed a mildly enhancing, 2-cm diameter, expansile cystic lesion in the middle of the clivus (Fig. 1).

The lesion was explored via a transnasal transsphenoidal approach. A discoloration was noted on the clivus at the site of the apparent lesion. A thin shell of bone overlying the cystic lesion was removed and the cystic lesion was entered. Yellow-white soft debris was noted in the lesion. The debris was sent to pathology for analysis, along with the cystic lining that was stripped at the margins of the lesion. The posterior fossa dura was visualized as the lesion had eroded through the clivus. As a final step, the bone opening was widely fenestrated into the nasopharynx.

Histological examination showed keratinized anucleate debris with associated macrophages, scattered cholesterol crystals, and a cyst lining composed of benign epithelial tissue (Fig. 2). The patient's headaches resolved postoperatively and she did not have any complications from surgery.

## Discussion

*Cholesteatoma location*

This case illustrates the potential for cholesteatomas to occur in atypical locations such as the clivus. We suspect that the origin of this tumor was a congenital rest of ectodermal epithelium that slowly grew as a result of shedding of desquamated cells from the epithelial lining. Acquired cholesteatomas can occur in the middle ear as a result of a vacuum phenomenon in the middle ear caused by chronic ear infections, but it is difficult to speculate on an acquired etiology for the clival cholesteatoma described.


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*Imaging characteristics*

In reviewing the MRI characteristics of the case, the lesion did not have signal characteristics typical of an epidermoid cyst, and also in consideration of the location, a cholesteatoma was not considered a likely initial diagnosis. Cholesteatomas are typically hypointense (or isointense) on T1-weighted images and hyperintense on T2-weighted MRI images. In this case, the lesion was hyperintense on T1 and isointense on T2 in comparison with brain. These tumors are typically non-enhancing, but slight peripheral enhancement can occur, as in the case we describe, in theory because of infiltration of granulation tissue or infections. A superinfection could be one reason for the atypical MRI signal in the case we describe, but this was not confirmed on histological analysis. On computed tomography, these lesions are typically of low attenuation and there is often bone erosion, which may have smooth or sclerotic borders.[1,3,4]

*Treatment*

These benign tumors are typically treated with close observation and surgical decompression as necessary. Given the benign characteristics of epidermoid tumors, there does not appear to be a role for adjuvant chemotherapy or radiation therapy. Instead, when they are surgically accessible, wide local excision of the tumors is recommended to prevent recurrence. When lesions are not surgically amenable to local excision, marsupialization may be considered. A wide fenestrated opening of the epidermoid cyst into an area where the desquamated debris can pass can prevent the lesion from growing and causing more local destruction.[2,3]


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**Figure legends**

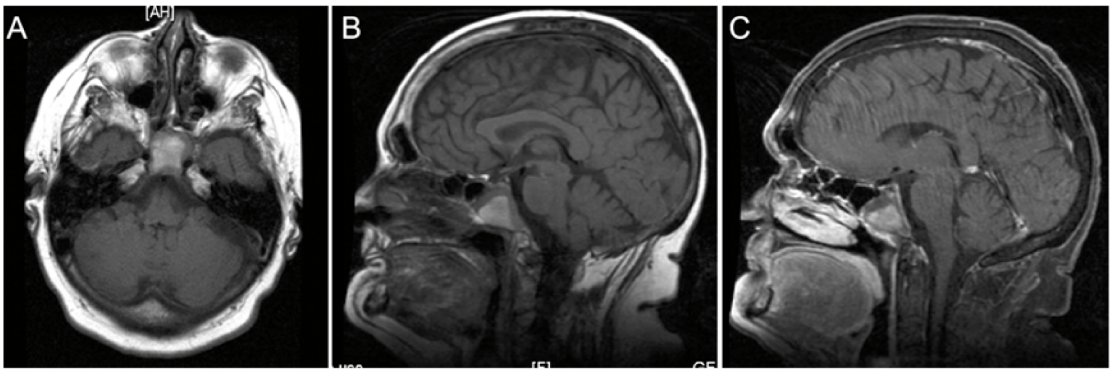


Figure 1: MRI from a 64 year-old woman with headaches showing a erosive lesion in the clivus. T1-weighted axial (A) and sagittal (B) images show a mixed signal intensity lesion in clivus with a hyperintense in central core surrounded by an area of hypointense signal. (C) Peripheral enhancement is shown on post-gadolinium T1-weighted sagittal image.



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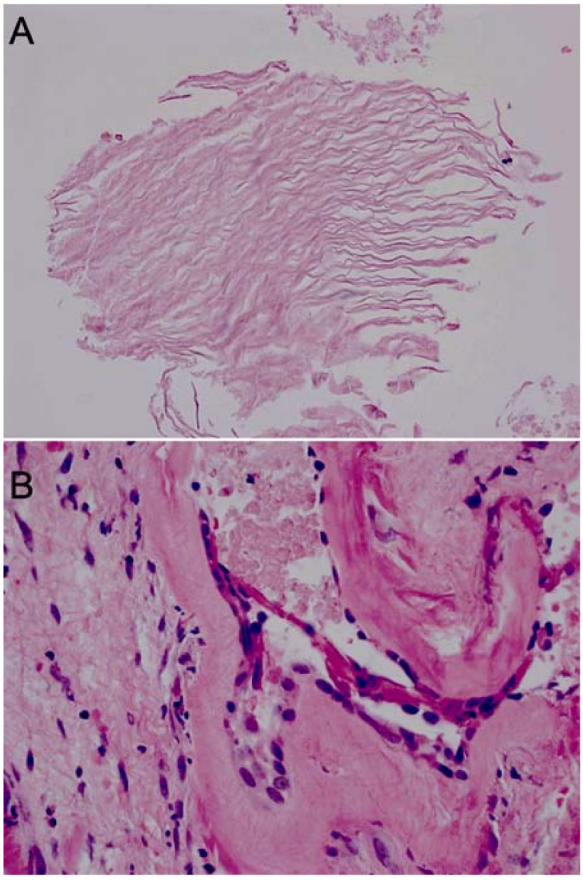


Figure 2. (A) Material removed from within the cyst is largely comprised of layers of anucleate squames. (B) A portion of thin benign epithelial tissue associated with overlying keratinizing squames and an underlying thickened basement membrane is evident. H&E stain.



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