Cholesteatoma of the Clivus

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ABSTRACT

Cholesteatomas (central nervous system epidermoids) can be found intradurally or extradurally in the central nervous system. Extradural intraosseous lesions are most commonly found in the petrous bone. The authors describe a unique case of a clival cholesteatoma 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 the clivus. A large portion of the clivus was eroded. The lesion was explored via a transnasal trans-sphenoidal 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). When surgically accessible, these lesions should be excised to prevent a recurrence. If inaccessible, marsupialization may be considered.

KEYWORDS: Cholesteatoma, clivus, epidermoid

Cholesteatomas, also known as epidermoids, epidermoid cysts, or benign epithelial cysts, are tumor-like masses 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 at a linear 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 in the central nervous system.^{1,2} When the tumors are intradural, they typically occur in the posterior fossa at the cerebellopontine angle. Extradural tumors are most often 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 a cholesteatoma in the midline clivus has never been reported.

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

CASE REPORT

A 64-year-old woman sought treatment from her primary physician with complaints of headache over the preceding 5 days. She reported no other neurological symptoms and her physical examination was normal. Computed tomography showed a lesion in the clivus, which prompted neurosurgical consultation and a magnetic resonance imaging (MRI) scan. The MRI 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 trans-sphenoidal approach. The clivus was discolored 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 present in the lesion. The debris, along with the cystic lining that was stripped at the margins of the lesion, was sent for pathological analysis. The lesion had eroded through the clivus, exposing the dura of the posterior fossa. 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). Post-operatively, the patient's headaches resolved and she had no complications related to surgery.

Figure 2 (A) Material removed from within the cyst is largely composed 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. Hematoxylin and eosin stain.

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 desquamated cells were shed from the epithelial lining. Acquired cholesteatomas can occur in the middle ear as a result of a vacuum phenomenon caused there by chronic ear infections. However, it is difficult to speculate on an acquired etiology for the clival cholesteatoma described.

Imaging Characteristics

In reviewing the MRI characteristics of the case, the lesion lacked signal characteristics typical of an epidermoid cyst. Given its 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 MRIs. In this case, the lesion was hyperintense on T1-weighted images and isointense on T2-weighted images compared with brain tissue. These tumors are typically non-enhancing, but slight peripheral enhancement can occur, as in our case, in theory because of infiltration of granulation tissue or infections. A superinfection could be one reason for the atypical MRI signal in our case, but this was not confirmed on histological analysis. On computed tomography, these lesions are typically of low attenuation, and bone erosion, which may have smooth or sclerotic borders, is often present.^{1,3,4}

Treatment

Typically, these benign tumors are treated with close observation and surgical decompression as necessary. Given the benign characteristics of epidermoid tumors, there appears to be no 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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Commentary

The authors have described the case of a cholesteatoma that presented primarily in the clivus. The lesion could possibly have been a “cholesterol granuloma.” A cholesterol granuloma and a cholesteatoma have similar origins in that they occur from obstructed petrous apex air cells, but their appearance on MRI is different. This cystic collection then fills up with cellular debris and cholesterol crystals. It grows by expansion and may erode bone to push the dura. The treatment is complete excision, if possible, by an extradural route. This may be limited

if there is extensive dural involvement or if the cholesteatoma is very large. If complete resection is not possible, it may recur. Wide drainage into a sinus cavity is preferred if complete resection is not

possible; thus, it is very important to avoid violating the cerebrospinal fluid space.

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