Acquired cholesteatoma presenting as a large pars squamosa temporal bone mass

Eric M. Jaryszak MD PhD¹, Christopher Vanison MS¹, Amanda L. Yaun MD², and Diego A. Preciado MD PhD¹
Divisions of Otolaryngology¹ and Neurosurgery², Children’s National Medical Center
George Washington University School of Medicine, Washington, DC 20010

ABSTRACT

Educational Objective: At the conclusion of this presentation, the participants should be able to discuss the differential diagnosis of a pars squamosa temporal bone mass, discuss unusual locations in which cholesteatoma may appear, and explain the need for long term followup in canal wall down mastoidectomy patients.

Objectives: To present a case of an acquired cholesteatoma presenting as a large lateral pars squamosa temporal bone mass and review the literature on cholesteatoma presenting in unusual locations.

Study Design: Case report and review of the literature.

Methods: While congenital cholesteatomas have been reported to arise in atypical locations such as the maxillary sinus and occipitoparietotemporal junction, there have been very few reports of acquired cholesteatomas arising in unusual locations. A 16 year old female with an acquired cholesteatoma presenting as a large lateral squamous temporal bone mass with intracranial extension nine years after canal wall down (CWD) mastoidectomy and six years after revision surgery is presented. Her management and followup are discussed and a review of the literature for atypical locations of cholesteatoma is presented.

Results: Review of the English literature revealed only a single case of acquired cholesteatoma within the squamous temporal bone. Our patient underwent successful excision of the mass through a lateral approach, confirming the diagnosis.

Conclusions: Acquired cholesteatoma typically arises in the middle ear and mastoid. Rarely, it can present in atypical locations. The consequences of undiagnosed, untreated cholesteatoma can be significant. This case highlights the need for routine long term surveillance in children with CWD mastoidectomy cavities.

INTRODUCTION

While congenital cholesteatomas have been reported to arise in atypical locations such as the maxillary sinus (1) and occipitoparietotemporal junction (2), there have been very few reports of acquired cholesteatomas arising in unusual locations. This is particularly true in the pediatric otolaryngology literature. We present a case of a 16 year-old girl with an acquired cholesteatoma presenting as a large lateral squamous temporal bone mass with intracranial extension.

REFERENCES


CASE REPORT

A 16 year-old female with a past history of an isolated cleft palate with associated Eustachian tube dysfunction requiring tympanostomy tube placement presented to our emergency department nine years earlier with acute right facial paralysis. She was found at that time to have bilateral attic cholesteatomas. The complicated right cholesteatoma was treated with a modified radical mastoidectomy. Following healing, the more limited left cholesteatoma was eradicated by a straightforward tympanoplasty. She did require revision right tympanomastoidectomy 3 years after initial presentation for recurrent cholesteatoma.

In routine followup nearly 7 years after initial presentation, she was noted to have a left retraction pocket and a CT scan of the temporal bones was performed (Figure 1). While the left ear showed no evidence of extension beyond the retraction, a large right squamous temporal bone mass was identified. An MRI demonstrated a 2.1-cm x 1.9-cm x 1.8-cm mass that was slightly hyperintense on T1-weighted imaging (Figure 2a) and peripherally hypointense with central hypointensities on T2-weighted imaging (Figure 2b). The dura was medially displaced by the lesion but did not appear to be disrupted. No other intra- or extracranial lesions were noted. The lesion demonstrated no growth on followup MRI 2 months later.

Given the concern for invasive pathology with the intracranial extension, we proceeded to the operating suite for resection of the mass. An endaural incision was made at the incisure, brought up around the root of the helix into a preauricular crease and extended cranially. Lateral bony expansion from the mass was noted and exposure of the mass in the roof of the mastoid revealed squamous debris consistent with cholesteatoma. The cholesteatoma was circumferentially removed revealing a 2 cm area of exposed dura (Figure 3a). The matrix was densely adherent to the dura, but no dural defect was noted. The roof of the mastoid was reconstructed with a temporals muscle flap secured medially and the skull defect was reconstructed with OstioVation (OsteoMed, Addison, TX, USA) (Figure 3b). The patient was discharged on post-operative day 2 without complication.

DISCUSSION

Cholesteatomas arising from the lateral temporal bone are extremely rare, particularly when lesions are acquired. Only one other case has been reported in the English literature (6). Several factors associated with the case described in this report indicate that our patient’s lesion was most likely acquired. First, the patient suffered from cleft palate and chronic Eustachian tube dysfunction syndrome. She was found at that time to have bilateral attic cholesteatomas. Her original right-sided lesion required a canal wall down (CWD) mastoidectomy. While this procedure may be more successful in eradicating larger lesions compared to canal wall up (CWU) techniques, there is still a risk of leaving residual matrix behind in the surgical defect (3). Symm and Luxford (4) reported that residual cholesteatoma was found in 14.6% of patients who underwent CWD mastoidectomy. Our patient did not experience any of the typical symptoms of cholesteatoma and this lesion was not connected to the middle ear space.

One cannot completely rule out the possibility that our patient’s lesion represented a congenital cholesteatoma, unrelated to her acquired lesion. Cases of congenital cholesteatomas and congenital intradiploic epidermoid cysts arising from the calvarium have been described (2,5). While congenital calvarial lesions tend to remain intradiploic with only partial erosion, our patient’s lesion was more aggressive with erosion of both inner and outer bony tables and was not present on previous preoperative imaging.

This difference between our case and the one described previously (6) may relate to the commonly held theory that pediatric cholesteatomas behave more aggressively than similar lesions in adults (7, 8). This has also been supported by biochemical studies which have found higher keratinocyte proliferation rates in pediatric cholesteatomas (9). Furthermore, rates of residual and recurrent cholesteatoma have been found to be higher in children compared to adults, thus advocating especially close follow-up after surgical eradication in younger patients (7, 8).

CONCLUSION

Acquired cholesteatomas arising from the lateral temporal bone are extremely rare. Despite extensive surgical eradication via CWD mastoidectomy, a known subset of patients will experience residual and recurrent disease. Therefore, swelling of the lateral temporal bone after removal of cholesteatoma should be investigated thoroughly. This is particularly true for pediatric patients in whom cholesteatomas are thought to behave more aggressively and demonstrate higher rates of recurrent and residual disease.