**Introduction**

Cholesteatomas are histologically benign but can behave in a locally aggressive manner. Most often cholesteatomas will remain confined to the middle ear and petrous mastoid bone. However, spread beyond this may occur due to the bone-eroding capacity of the disease process. The case study outlined below describes a very aggressive cholesteatoma with extensive skull base and C-spine involvement. A combined surgical approach between the Otology/Neurotology, Rhinology/Skull Base and Neurosurgical teams was utilized.

**Discussion**

Skull base cholesteatoma presenting outside of the petrous mastoid bone is extremely rare. In fact, to our knowledge, this is the first case report regarding a cholesteatoma involving the C-spine. While the principle for the treatment of cholesteatoma remains the same, the deep location of this cholesteatoma, abutting important structures such as the C-spine and lower cranial nerves gave clinicians a significant challenge for the complete removal of the disease with minimal morbidity.

During our first surgical approach, in the semi-urgent setting (occipital abscess) and without fully appreciating the extent of the disease, the traditional post-auricular incision was used. The incision was later extended into the neck for better exposure, however there was still limited visualization of the skull base and upper C-spine such that we were unsure if all of the epithelium had been completely removed. The revision surgery with endoscopic transnasal approach provided a clear view of the medial skull base and cranio-cervical junction. This case demonstrates how recent advances in endonasal surgical techniques and image guidance have significantly expanded our ability to safely operate on the skull base.

Advantages of newer endoscopic approaches to the skull base include avoidance of an open craniotomy, faster recovery and shorter hospitalization.

The pathogenesis of this aggressive skull base cholesteatoma was unclear. We did not have a detailed operative report from the first canal wall down procedure. The patient presented with normal hearing and a normal appearing tympanic membrane. Our revision tympanomastoidectomy revealed an uninvolved mastoid and middle ear. This does not follow the classic pattern for either primary or secondary cholesteatoma, and so a congenital cholesteatoma or a cholesteatoma from a remote trauma at the occipital area cannot be ruled out.

Image studies also played a significant role in the management of this case. Both CT scan and MRI were necessary to define the extent of the disease. With the extent of bony erosion on CT scan, a malignant lesion could not be ruled out. However, with no soft tissue invasion noted on MRI, a malignant lesion was less likely. The final diagnosis, of course, can be only confirmed by pathology.

**Results**

This case involves a 65 year-old male with a previous history of left sided canal wall down tympanomastoidectomy for cholesteatoma. He presented to the Loma Linda Emergency Department several years after his initial surgery with a two-week history of painful occipital swelling and left sided purulent otorrhea. Initial CT of the temporal bone (Figure 1) revealed left sided occipital bone erosion with overlying abscess, along with a soft tissue mass surrounding the left occipital condyle and associated bony erosion. The patient was taken for an I&D of the occipital abscess, biopsy of the lateral skull base lesion and revision tympanomastoidectomy. The intra-operative findings revealed just how extensive the cholesteatoma was, with gross involvement of the occipital bone extending to the occipital condyle. Interestingly, there was only a small amount of purulence noted in the inferior portion of the mastoid cavity, otherwise the middle ear and mastoid were uninvolved. A lateral skull base resection of the cholesteatoma was performed by Neurosurgery during the same general anesthetic.

The patient was followed with a CT scan of the temporal bone and MRI of the skull base eight months later; at which point his only symptom was persistent occipital pain. A repeat CT (Figure 2) revealed expansion of the disease well past midline and into the cranio-cervical junction, with extensive bony erosion of C1, the occipital condyle and clivus. MRI of the skull base (Figure 3) showed the lesion had increased signal at T2 and was neutral at T1 with no gross invasion of surrounding soft tissue or the spinal chord. He subsequently underwent a transnasal endoscopic resection of the skull base and C1/C2 cholesteatoma, as well as open occipital cervical fusion (performed by Neurosurgery).

The patient had a delayed occipital wound breakdown that required placement of a wound vac system, but did not develop any complications from the endonasal approach. He was discharged on post-operative day 8 following the endoscopic skull base resection. There was no recurrence of the skull base cholesteatoma noted on a 5 month follow up exam. The patient did however die of metastatic colon adenocarcinoma approximately 1 year after surgery.

**Conclusion**

Most often cholesteatoma will remain confined to the middle ear and petrous mastoid bone, however spread beyond this may occur due to the bone-eroding capacity of the disease process. This is believed to be the first case report regarding cholesteatoma extending to the skull base with C-spine erosion. Both CT scan and MRI are essential to define the extension of the disease and to help plan an adequate surgical approach. The addition of the endoscopic transnasal approach to the skull base rendered a safe and complete removal of this skull base cholesteatoma.

**Methods**

Chart review of a single patient treated from January 2009 to August 2010 by the Otology/Neurotology and Rhinology/Skull Base services of the Head & Neck department as well as the Neurosurgery department at Loma Linda University Hospital. In total the patient underwent 5 surgeries by the above-mentioned services as well as 1 tympanomastoidectomy by an outside surgeon. All radiographic images are from ‘in-house’ scanners.

**References**