Congenital Cholesteatomas Originating within the Facial Nerve Canal

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Abstract

Objectives: Congenital cholesteatoma (CC) is a squamous epithelial lesion within the temporal bone that can erode nearby structures and afflicts 1.2 per 1,000,000 individuals annually. Although the pathogenesis is unknown, the persistence of a squamous cell nest may underlie the condition. We present two cases of CC originating within the facial canal.
Study design: Case series.
Methods: Retrospective review of two CC patients presenting to our otology/neurotology clinic in a tertiary care academic center.
Results: Two patients, 16-months (patient 1) and 5-years (patient 2) of age, presented with grade VI facial paralysis. Hearing in both cases was normal. Computed tomography (CT) demonstrated lesions in the tympanic segment (patient 1) and the labyrinthine segment (patient 2) of the facial nerve. A tympanomastoidectomy with an extended facial recess was performed for the first patient, and a middle fossa approach was performed in patient 2. The epidermoid was medial to the nerve in patient 1, and accordingly a segmental nerve resection was required. Keratin debris was observed on both nerves, and the middle ear was otherwise normal. Nerve histopathology demonstrated cholesteatoma sacs. At 6- and 12-months postoperatively, patient 1 had grade III facial function while patient 2 had grade V facial function.
Conclusions: CC within the facial nerve canal may present with complete facial paralysis in otherwise healthy young patients. Complete CC resection is warranted and may improve facial function. Segmental resection and grafting may result in better nerve outcomes.

Introduction

- CC is typically found in the anterior mesotympanum or posterior superior quadrant.
- CCs are typically identified in early childhood (1-5 years of age).
- As they grow, CCs may obstruct the Eustachian tube, produce chronic middle ear effusions, and/or result in hearing loss.
- CCs are squamous epithelial lesions in the temporal bone that can erode nearby structures, with an incidence of 1.2 per 1,000,000.
- Although the pathogenesis is unknown, the persistence of a squamous cell nest may underlie the condition.
- We present two cases of CC originating within the facial canal.

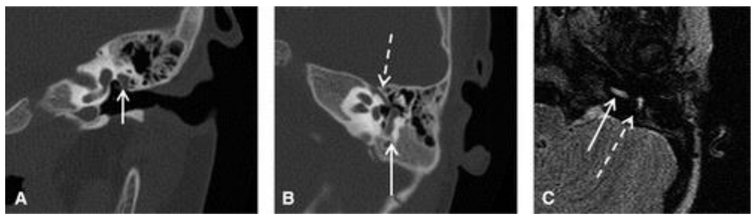

Figure 1. (**A**) Coronal noncontrast CT image of patient 1 at the level of the middle ear. The enlarged fallopian canal (*arrow*) can be seen in patient 1 (1.25-mm slice thickness). (**B**) Axial CT image at the level of the vestibule. The normal diameter of the proximal facial nerve (*dashed arrow*) can be contrasted with the enlarged fallopian canal (*solid arrow*) at the second genu. (**C**) Axial T2-fast spin echo MRI at the level of the basal turn of the cochlea (*solid arrow*). The facial nerve shows hyperintensity in the proximal mastoid portion (*dashed arrow*).

Methods and Materials

- Retrospective review of two CC patients presenting to our otology/neurotology clinic in a tertiary care academic center.

Results

- Two patients, 16-months (patient 1) and 5-years (patient 2) of age, presented with grade VI facial paralysis [**Table 1**].
- Hearing in both cases was normal.
- Magnetic resonance imaging (MRI) and computed tomography (CT) demonstrated lesions in the tympanic segment (patient 1; **Figure 1**) and the labyrinthine segment (patient 2; **Figures 2-3**) of the facial nerve.
- A tympanomastoidectomy with an extended facial recess was performed for the first patient, and a middle fossa approach was performed in patient 2.
- The epidermoid was medial to the nerve in patient 1, and accordingly a segmental nerve resection with greater auricular nerve interposition was required.
- Keratin debris was observed on both nerves, and the middle ear was otherwise normal.
- Nerve pathology demonstrated cholesteatoma sacs [**Figure 4**].
- At 6- and 12-months postoperatively, patient 1 had grade III facial function while patient 2 had grade V facial function.
- Patient 2 underwent a cross facial nerve graft with gracilis flap

Table 1. Tabulation of lesion characteristics.

	Patient 1	Patient 2
Age (years)	1.3	5
Facial paralysis grade on presentation	VI	VI
Hearing on presentation	Normal	Normal
Lesion location	Tympanic segment of facial nerve	Labyrinthine segment of facial nerve
Surgical approach	Tympanomastoidectomy with extended facial recess	Middle fossa
Intraoperative findings	Keratin debris observed on nerve; epidermoid medial to nerve; normal middle ear	Cholesteatoma sac on nerve; normal middle ear
Intraoperative notes	Segmental nerve resection required	-
Histopathology	Cholesteatoma sac	Cholesteatoma sac
Facial paralysis grade 6-months postoperatively	III	V
Facial paralysis grade 12-months postoperatively	III	V

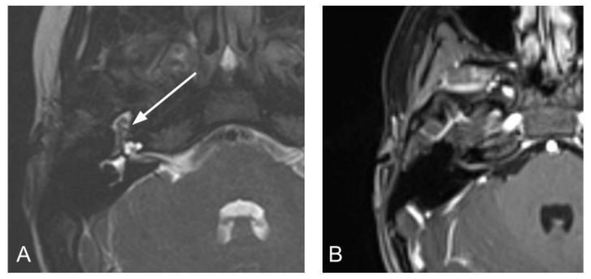

Figure 2. (**A**) Axial CISS sequence MRI of patient 2 demonstrating the lesion which is mostly hyperintense (arrow). (**B**) Axial post-gad fat-sat T1 MRI of showing lesion isointensity and some edema of the facial nerve in the IAC.

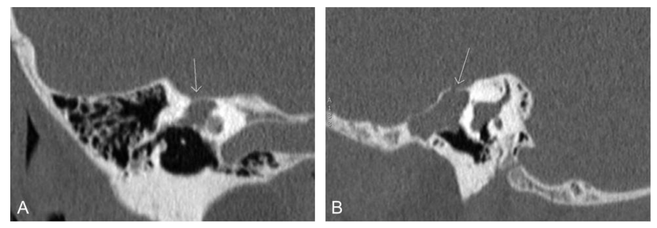

Figure 3. (**A**) Coronal noncontrast CT image of patient 2 demonstrating lesion in the labyrinthine segment of the facial nerve. (**B**) Sagittal noncontrast CT image.

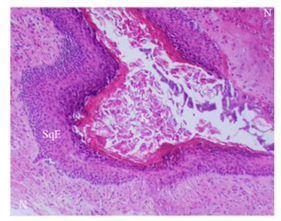

Figure 4. Hematoxylin and eosin-stained image of the resected specimen in patient 1 showing squamous epithelium (SqE) and keratin (K) surrounded by neural fibers (N). Original magnification is at ×20.

Discussion

- CCs can present as extradural or intradural, with the middle ear being the most prevalent site.
- CT and MRI are critical in the diagnosis of CC, and treatment is surgical.
- Clinical manifestations vary, including conductive hearing loss, disequilibrium, and neural manifestations.
- In a case of CC originating in the fallopian canal, we recommend excision and grafting of the involved segment of nerve if the cholesteatoma cannot be separated from the nerve.
- This is to be distinguished from a cholesteatoma that has secondarily involved the facial nerve, in which case the nerve should be left intact and the cholesteatoma should be dissected off the nerve or exteriorized.
- CCs originating in the nerve are distinguished by early facial paralysis and an enlarged fallopian canal on CT imaging.

Conclusions

- CC within the facial nerve canal may present with complete facial paralysis in otherwise healthy young patients.
- Complete CC resection is warranted and may improve facial function.
- Segmental resection and grafting may be required in some cases.

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8. Figure 1: http://www.adv-ent.com/congenital-cholesteatoma/
9. Figure 2: http://www.dallasear.com/conditions-cholesteatoma-cases-congenital2.html