Online Case Report

Non-classical presentation of congenital cholesteatoma

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Congenital cholesteatoma is a rare entity. It arises from aberrant epithelial remnants left at the time of closure of the neural groove, between the 3rd and 5th week of embryonic life. Otoscopy typically shows a pearly white mass, medial to the intact tympanic membrane, primarily in the anterosuperior quadrant, without a preceding history of otorrhoea, tympanic membrane perforation or surgery. It can present at any age from birth to early adulthood. The most common presenting symptom is hearing loss or facial nerve palsy. We present a non-classical presentation of congenital cholesteatoma with loss of taste sensation and numbness on the anterior two-thirds of the tongue associated with a bulging posterosuperior quadrant of the ipsilateral tympanic membrane in a 52-year-old male. After investigation, the patient underwent surgical management resulting in improvement of his symptoms.

Keywords: Congenital cholesteatoma – Chorda tympani – Taste sensation – Tympanic membrane – Temporal bone

Congenital cholesteatoma (CC) is thought to occur secondary to failure of normal involution of epidermoid formation within the middle ear cleft. A collection of stratified squamous epidermoid cells appears during embryonic development (weeks 3–5) and are instrumental in the development of middle ear mucosa. These remaining squamous cells grow slowly and become clinically apparent as a cholesteatoma usually during childhood. This lesion is usually located intradurally; less frequently (20%), an extradural location is noted. There are five general sites of extradural occurrence: the middle ear, external auditory meatus, the mastoid, squamous portion of temporal bone and the petrous apex.¹

CC may be asymptomatic or present as a conductive hearing loss. Other presentations include otalgia, vertigo and facial palsy. The later symptoms indicate erosion into the semicircular canals or the Fallopian canal, respectively. Symptoms of headache and fever may herald impending intracranial complication. Loss of taste sensation (chorda

tympani dysfunction) has not previously been reported as a presenting complaint.

Clinically, CC is identified as a white round mass medial to an intact tympanic membrane, usually at the anterosuperior quadrant.² CC is diagnosed on the basis of a specific set of criteria: (i) cholesteatoma sac medial to an intact tympanic membrane; (ii) a normal tympanic membrane; and (iii) no history of tympanic membrane perforation, otorrhoea, trauma or surgery.³ Early diagnosis of CC is important to prevent extensive surgery and complications. Computerised tomography (CT) demonstrates an isolated, non-enhancing, hypodense and, possibly, invasive lesion. Treatment is by surgical excision.

Case report

A 52-year-old man presented with a 2-week history of right-sided tongue numbness and loss of taste sensation.

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Figure 1 Otoscopic and intra-operative findings of congenital cholesteatoma. (a) Otoscopy revealing a bulge in the posterosuperior quadrant of the right intact tympanic membrane. (b) Cholesteatoma in the attic, eroding the incus and lying on the chorda tympani. (c) Further epithelial pearls overlying the facial nerve. (d) Complete removal of cholesteatoma. LSC, lateral semicircular canal; FN, facial nerve.

There was no history of otorrhoea or previous otological procedure. Otoscopy revealed a bulge in the posterosuperior quadrant of the right intact tympanic membrane (Fig. 1a). Facial nerve function, balance and flexible nasolaryngoscopy were normal. Pure tone audiometry demonstrated a 40 dB conductive hearing loss in this ear, with normal hearing contralaterally. CT scans of the temporal bone showed an isolated soft tissue

Figure 2 CT scan of the temporal bone shows an isolated soft tissue density lesion in the right middle ear cleft.

density lesion in the middle ear cleft (Fig. 2). Ear exploration, via an endaural incision, demonstrated cholesteatoma in the attic, eroding the incus and lying on the chorda tympani (Fig. 1b) with extension into the facial recess and sinus tympani. The facial canal was intact. The incus remnant was removed to facilitate cholesteatoma removal. Chorda tympani integrity was preserved. Further epithelial pearls overlying the facial nerve (Fig. 1c) were removed (Fig. 1d). A temporalis fascia graft was placed over the mobile stapes and a type-III tympanoplasty fashioned.

The patient made an uneventful postoperative recovery and his facial nerve function remained normal. At review (3 weeks), his taste sensation had subjectively improved with a 15 dB improvement in air conduction threshold. The patient's taste sensation remained normal at 12 months. There was no further change in the audiometric threshold. Histology confirmed the tissue to be a cholesteatoma.

Discussion

CC develops from a congenital epithelial cell rest in the temporal bone and may present at any age from infancy to adulthood with a mean age of 16.7 years.⁴ Typically, presentation is with a discrete white lesion behind an

intact tympanic membrane.⁵ The majority of cases involve the anterosuperior quadrant although they can arise from other parts of the tympanic membrane. If located in the posterosuperior quadrant, CC can involve the ossicular chain even in the absence of a conductive hearing loss.⁶ As the histology is similar in congenital and acquired cholesteatoma, the distinction is made by the presence of an intact tympanic membrane, and the absence of previous otological disease, tympanic membrane perforation or surgery.³ CC is usually asymptomatic in the initial stages of growth and micro-otoscopy cannot reliably predict the extent of disease; therefore, radiological evaluation with a CT scan should be considered.

This case confirms that CC may present non-classically in an older age, at an uncommon location without the appearance of a white mass behind the tympanic membrane. Although a conductive hearing loss is the most common presenting symptom, other symptoms in the elderly such as otalgia, vertigo and headache have been reported.²

This report illustrates a previously unreported symptom of loss of taste sensation. Unilateral loss of taste sensation at any age should prompt a thorough search for pathology in the middle ear. Although benign in nature, CC can be locally destructive. Late presentation often results in extensive disease and increases the potential for serious complications.

Conclusions

- Congenital cholesteatoma is a rare entity.
- We present a non-classical presentation in an older age group, in an uncommon posterosuperior location, with a previously unreported symptom of loss of taste sensation.
- Symptomatic improvement occurs after surgical removal of the disease.

References

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