Abstract. We report here on a case of congenital cholesteatoma that extended into the internal auditory meatus and cochlea. A 17-year-old boy underwent surgery for a very large cholesteatoma, which was discovered behind an intact tympanic membrane. Pure tone audiometry revealed an unresponsive ear. High resolution temporal bone computed tomography showed perilabyrinthine extension with its absence in the vestibular area, and destruction of the bony cochlea at the basal turn, the tegmen and the posterior cranial fossa. Magnetic resonance imaging revealed no intracranial extension. Surgical exploration of the middle ear and the mastoid cavity revealed that both the vestibule and the basal turn of the cochlea were filled with a noninfected cholesteatoma. The cholesteatoma extended into the internal auditory meatus through translabyrinthine destruction; it extended into the basal turn of the cochlea through the infralabyrinthine route. The bony segment of the facial nerve canal demonstrated near total dehiscence. The cholesteatoma was removed by the transotic approach. Congenital cholesteatoma is characterized by no specific history. Therefore, early detection of this malady can be challenging, but it is important to prevent such complications as were observed in this reported case.

Congenital cholesteatoma was defined by Derlacki and Clemis (1) as embryonic nests of epithelial tissues in the ear without tympanic membrane perforation or a history of ear infection. Levenson et al. (2) modified the definition of a congenital cholesteatoma to include a normal pars flaccida and pars tensa, the absence of prior otorrhea and no history of prior otologic procedures. A congenital cholesteatoma that extends beyond the middle ear is rare. Cholesteatomas extending beyond the limits of the middle are not common; 15% of middle ear cholesteatomas involve the inner ear structures and they were reported to be extensive in a large series (3). Bone destruction with a labyrinthine fistula is commonly associated with cholesteatoma. The semicircular canals and the promontory of the cochlea are the common areas for a labyrinthine fistula which is complicated by cholesteatoma. Gravelli et al. (4) reported on 4 cases of extensive cholesteatoma with translabyrinthine destruction. However, extensive perilabyrinthine destruction extending into the internal auditory meatus is very rare in patients with congenital cholesteatoma.

We have treated a 17 year old boy who had a congenital cholesteatoma with a perilabyrinthine extension into the cochlear and the internal auditory meatus via a labyrinthine fistula.

Case Report

A 17-year-old patient underwent surgery for a very large cholesteatoma that was discovered behind an intact tympanic membrane. The patient had no history of otitis media. At about the age of 12, he had noted a rapid loss of hearing in the left ear along with intermittent headache. At our first examination, the symptoms indicated the total absence of left inner ear function; the boy was referred to us too late and the hearing loss could not be restored. Otomicroscopy revealed an intact tympanic membrane. Temporal bone computed tomography (CT) showed a soft tissue mass invading the labyrinth, the internal auditory meatus and the basal turn of the cochlea (Figure 1A, B). A bony defect at the tegmen and the posterior cranial fossa was noted (Figure 2A, B). The T1-weighted magnetic resonance imaging (MRI) (axial view) showed a low signal intensity mass and the T2-weighted MRI images (axial view) revealed a high signal intensity mass without intracranial extension (Figure 3A, B).

Left tympanomastoidectomy was performed under general anesthesia. A cholesteatoma was found filling the mastoid cavity and epitympanum and it had completely invaded into...
The facial nerve was found in the expected location, but the bony canal of the facial nerve from the mastoid segment to the geniculate ganglion revealed bony dehiscence. The cholesteatoma passed behind the pyramidal bend of the facial nerve and invaded into the basal turn of the cochlea. All the cholesteatoma matrix and granulation tissue were removed from the inner ear by the transotic approach. However, there was pinpoint tearing of the dura at the recently eroded internal auditory meatus. Leakage of the cerebrospinal fluid was treated using a small piece of fat and a temporalis muscle fascia graft with fibrin glue. The mastoid was partially obliterated using the temporalis muscle; this blocked the communication route between the inner ear and the cranial fossa which was caused by the inflammatory products. Lumbar drainage was performed immediately after surgery. The pathological examination of the mass confirmed a cholesteatoma.

No facial palsy or dizziness occurred after surgery. The lumbar drain was removed seven days later. There has been no recurrence of the cholesteatoma for over 24 months after surgery.

Discussion

Congenital cholesteatoma is a slowly progressive, destructive middle ear disorder that produces keratinizing stratified squamous epithelium in the middle ear cleft. In contrast to a
common cholesteatoma, most congenital patients present without otorrhea, and the tympanic membrane appears to be normal. The presenting signs of an extensive intratemporal congenital cholesteatoma may be misleading. The first symptom is usually hearing loss (5). Sheehy and Brackmann (3) reviewed their 1,024 cholesteatoma cases and they found that a labyrinthine fistula was present in 97 (9.5%). Involvement of both the cochlea and the internal auditory canal was observed in 2 ears (0.2%).

Facial palsy is frequently associated with extensive cholesteatoma (6, 7). This symptom is often related to the involvement of the geniculate ganglion and the tympanic segment of the nerve following the extension of the cholesteatoma to the supratubal recess from the attic compartment (8). In the present case, facial palsy was not associated with the ipsilateral hearing loss despite the presence of a near totally destroyed bony canal of the intratemporal facial nerve. The preoperative radiological examination of an extensive cholesteatoma is based on temporal bone high resolution CT (9, 10). MRI can provide important information in cases with intracranial extension, as well as suggesting the probable histological type of the lesion (11, 12). In the present case, the extension involved the translabyrinthine, supralaby-rinthine and infralabyrinthine areas. The cochlea basal turn may be involved by an infralabyrinthine extension (7). Involvement of the cochlea by a cholesteatoma has a poor prognosis with regard to the patient’s hearing. In our case, the translabyrinthine lesion was approached through a transotic route, and this approach carries a low risk of damage to the facial nerve. In the present case, we removed the matrix by the transotic route. The defect of the bony cochlea was obliterated using temporalis muscle fascia. Because congenital cholesteatoma is silent in nature with regard to the signs and symptoms, early detection of this malady is very challenging, but making the correct, timely diagnosis is important to prevent complications that can lead to permanent hearing loss.

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References


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