The Journal of Laryngology and Otology
March 1980. Vol. **94.** pp. 263–268

Congenital cholesteatoma

Preservation of cochlear function after extensive labyrinthine destruction

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IT has always been an established dictum that destruction of any part of the membranous labyrinth would lead to total destruction of the function of the inner ear. Exceptions to this rule exist (Sadé-Sadowsky, 1955; Shambaugh, 1967; Pedersen, 1969; Phelps, 1969; Bumsted *et al.*, 1977).

We have encountered a patient with a congenital cholesteatoma which invaded all three semicircular canals, as well as the cochlea, with normal hearing preoperatively, in whom sensori-neural hearing was preserved even after total removal of all the semicircular canals, including the ampullae. This case, which must be considered unique, will be reported below.

Case report

The patient was a 32-year-old woman, with no previous history of ear disease. She was admitted to our department because of a left facial paralysis, which had developed gradually over a period of 2½ months. She had no complaints of hearing loss, tinnitus or vertigo.

Physical examination revealed normal tympanic membranes, without signs of any chronic ear disease, and a partial but almost total left-sided facial paralysis of peripheral type involving all branches of the facial nerve. There were no signs of other cranial nerve involvement, and no other neurological deficits. The intermedius function, as assessed by testing the nasolacrimal reflex (Zilstorff-Pedersen, 1959), showed decreased function on the involved side. Audiological examination revealed completely normal hearing, while the vestibular function was abolished, even when stimulated with 20° water for 90 sec. CT scanning and pneumoencephalography were normal. Hypocycloidal tomography of the temporal bones showed the right side to be normal (Fig. 1); the left side is shown in Fig. 2. Extensive destruction of the labyrinth (arrow), with involvement of all semicircular canals, including the vestibule, was demonstrated. Large defects towards the middle cranial fossa and the basal coil of the cochlea can be seen. The basal coil of the cochlea is blurred, but the lumen is of normal caliber; the other coils were normal. The destruction also involves the internal auditory canal with destruction of the transverse crest. The ossicles, mastoid and external auditory meatus were normal in all projections. The radiological diagnosis was that of a congenital cholesteatoma.

The patient was operated upon through a postauricular incision and an ex-

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https://doi.org/10.1017/S0022215100088770 Published online by Cambridge University Press

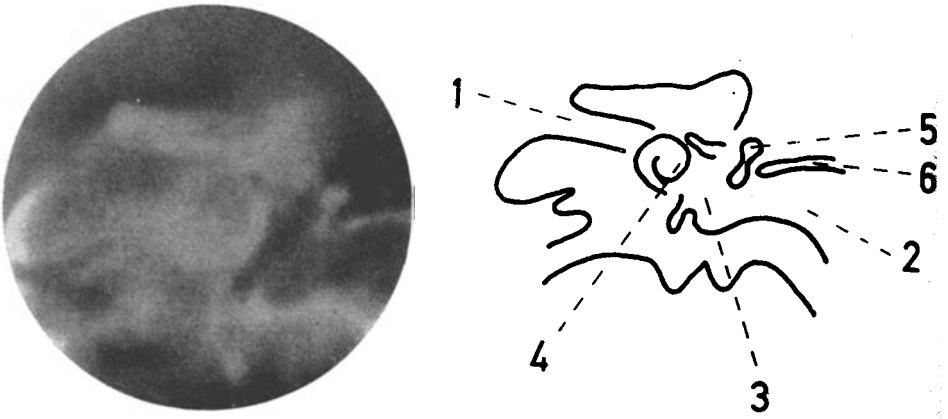


FIG. 1

Tomogram of right, normal temporal bone in 32-year-old female. 1. Internal acoustic meatus. 2. External acoustic meatus. 3. Tympanic cavity. 4. Cochlea. 5. Ossicles, 6. Spur.

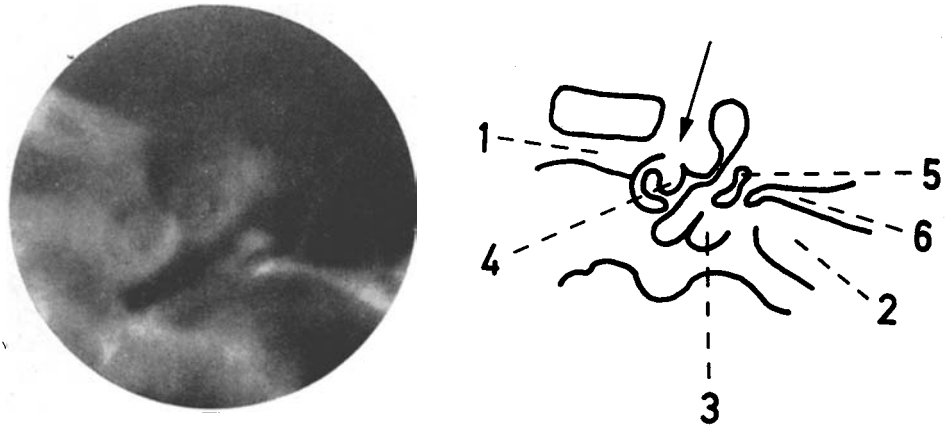


FIG. 2

Tomogram of left, diseased temporal bone in 32-year-old female. Extensive destruction of the labyrinth (Arrow), with involvement of all semicircular canals, including the vestibule. Extensive defects of the basal coil of the cochlea as well as of the bony lining towards the middle cranial fossa. The basal coil of the cochlea is relatively blurred, but its lumen is of normal caliber. The remaining cochlear coils are normal. The destruction involves the internal acoustic meatus, with destruction of the transverse crest. Numbering as in Fig. 1.

tensive mastoidectomy was performed. A large cholesteatoma was found to have destroyed the major part of the bony semicircular canals, and eroded away all the bone around the internal acoustic meatus, as well as the bone towards the middle cranial fossa. The horizontal part of the facial nerve was completely exposed by the cholesteatoma, including its superior and medial aspects, but there was no direct involvement of the middle ear. The incus was removed in order to secure total removal of the cholesteatoma. At the end of the operation

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the normal contents of the internal auditory meatus were seen through the dura, but there was no defect anywhere to indicate intracranial intrusion.

The postoperative course was uneventful and the patient was discharged in good condition. On the day of discharge her audiogram showed a conductive hearing loss, due to removal of the incus; but her sensori-neural hearing capacity was retained despite some high tone impairment (see Fig. 3). Discrimination score was 72 per cent. Tuning fork tests confirmed the audiometric findings, with a negative Rinne, and lateralization of the Weber response to the operated (left) side. Her facial function was unchanged, compared to the preoperative condition.

At a 3-month control examination she complained that something had happened just recently in her left ear, and she was afraid that she had lost the hearing in that ear. We found a normal tympanic membrane, but a dead ear. Facial function was slightly improved.

The patient was seen one year postoperatively; facial function was now normal, but the hearing and vestibular function in the left ear were abolished. Repeat tomography of the temporal bone (Fig. 4) revealed the extensive cavity following the operation, no signs of ossicles and some additional blurring of the cochlea, compared with preoperative tomography (Fig. 2).

Discussion

In the fenestration era, a total of 6 cases were recorded with preservation of hearing despite damage to the membranous labyrinth. These cases were reviewed by Sadé-Sadowsky (1955), who added 3 cases of his own. Other authors have presented cases with more extensive destruction

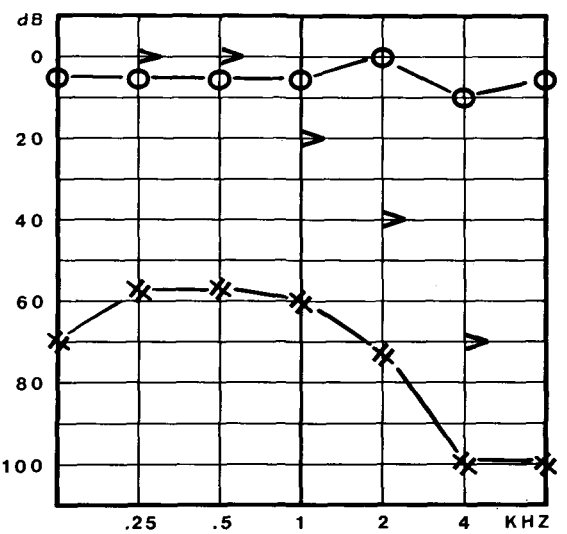


FIG. 3

Audiogram of 32-year-old female, who had large congenital cholesteatoma removed 7 days previously, with total removal of the superior part of the labyrinth. Left-sided conductive hearing impairment, with some high-tone sensori-neural impairment.

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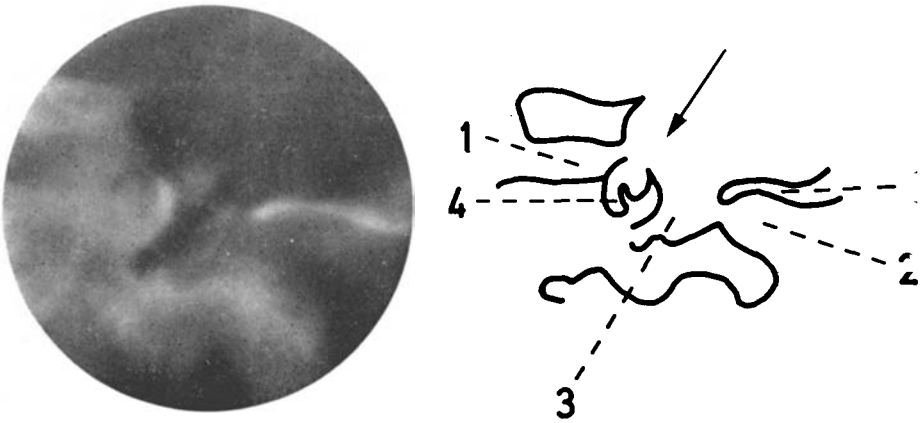


FIG. 4

Tomogram of left temporal bone in 32-year-old female, who had large congenital cholesteatoma as well as having the superior part of the labyrinth removed 1 year previously. The extensive cavity created at the operation is seen, and the cochlea is additionally blurred as compared to the preoperative condition (Fig. 2). No signs of middle ear ossicles.

of the labyrinth and preservation of hearing. Pedersen (1969) reported a case of Menière's disease with partial labyrinthectomy and removal of the lateral semicircular canal. The hearing and vestibular function were preserved 12 years after the operation. Shambaugh (1967) reported one case of chronic otitis media, in which removal of parts of the bony semicircular canals in both ears resulted in total loss of vestibular function, but in which hearing was retained in one ear. In addition, Phelps (1969) has reported three cases with preserved hearing with labyrinths invaded by cholesteatomas. To these cases Bumsted *et al.* (1977) report 3 additional cases (4 ears) with far more extensive destruction of the labyrinth than in previously described cases, but with preservation of hearing. One of these patients suffered from a congenital cholesteatoma, with hearing preserved 18 months after extensive labyrinthine destruction.

Congenital cholesteatomas involving the temporal bone arise from congenital epithelial inclusion remnants. When they involve the cerebellopontine angle one might suspect such a diagnosis because of the distinctive scalloped contour of the surface in Pantopaque cisternography (Thomsen, 1976). However, when the cholesteatoma is confined to the temporal bone, the diagnosis may not be so easy. According to Peron and Schuknecht (1975), who reviewed the literature, five sites of occurrence within the temporal bone are mentioned: 1. The petrous apex, 2. The mastoid, 3. The middle ear, 4. The middle ear and mastoid, 5. The external auditory canal. The symptoms vary according to the location. Eighty-three per cent of patients with a lesion in the petrous apex were found to have facial nerve involvement at the time the diagnosis was made, and 94 per cent of these patients had a unilateral, sensori-neural hearing loss at the

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time. Our patient, whose cholesteatoma probably originated in the petrous apex, presented herself with facial paralysis, which is in accordance with the above-mentioned findings from the literature. However, she differed in so far as her hearing was completely normal at the time of diagnosis, in spite of the fact that tomography demonstrated extensive destruction of the basal coil of the cochlea. In this way our patient resembles one of the patients presented by Phelps (1969), who had a similar cholesteatomatous erosion of the cochlea preoperatively, yet with preserved hearing. However, Phelps' patient lost her hearing at surgery. It was his impression that,when destruction of a labyrinth is sufficiently slow, the inflammatory response so induced may allow a layer of granulation tissue to be laid down, thus walling off the adjacent parts of the labyrinth, with retention of function. This mechanism may also explain the favorable outcome in three of the ears presented by Bumsted *et al.* (1977), but in the fourth ear in their series, as well as in our own case, this mechanism cannot adequately explain the preservation of function. In Phelps' patient the surgical procedure produced a dead ear; in the case of Bumsted *et al.* and in our own, only a loss of caloric response was produced.

According to Bumsted *et al.* (1977), there are 4 places where the inferior part of the labyrinth (cochlear duct and saccule) can be sealed off from the superior part (semicircular canals and utricle): 1. ductus reuniens, 2. the saccular duct, 3. the utricular duct and 4. the utriculoendolymphatic valve, which is located in the antero-inferior wall of the utricle at the orifice of the utricular duct. This valve was first described anatomically by Bast (1928), and its likely functional significance was described by Schuknecht and Belal (1975). They concluded that the utriculoendolymphatic valve is ideally suited to preserve the humoral and anatomical features of the superior part of the labyrinth from the developmental, pathological and traumatic susceptibilities of the inferior part. Bumsted *et al.* (1977) concluded, and we agree with them, that it may also function in a reverse manner; that is, it may protect the cochlear duct and sacculè from a destructive process occurring in the superior part, especially if the pathological process is a gradual one. In our case it may therefore be surmized that, owing to the slow progression of the congenital cholesteatoma, the utriculo-endolymphatic valve was sealed off, and therefore able, at least for a time, to withstand the consequences of removal of the entire superior part of the labyrinth.

Abstract

A patient with a large congenital cholesteatoma of the left temporal bone is presented. The case is unique in that, although the cholesteatoma invaded all three semicircular canals as well as the basal coil of the cochlea, the patient retained normal hearing. She had an almost complete left-sided facial paralysis, yet she retained sensori-neural hearing postoperatively, in spite of removal of the superior part of the labyrinth (semicircular canals

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and utricle). The possible mechanisms behind such an unusual event are discussed together with the possible function of the utriculo-endolymphatic valve.

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