The Journal of Laryngology and Otology
December 1990, Vol. **104**, pp. 945–948

Inner ear cholesteatoma and the preservation of cochlear function

Roberto Gamoletti, M.D., Mario Sanna, M.D., Carlo Zini, M.D., Abdel-Kader Taibah, M.D., Enrico Pasanisi, M.D., Luca Vassalli, M.D. (Parma, Italy)

Abstract

Labyrinthine destruction by direct cholesteatoma invasion has always been considered a serious threat to the inner ear function.

A number of reports in the literature have cited both patients who had preservation of hearing despite widespread erosion of the labyrinth by cholesteatoma and patients who had retained auditory function despite surgical removal of the matrix from the labyrinth. In most cases the vestibular portion of the inner ear was invaded but cases of cochlear involvement have been described as well.

Twelve cases with pre-operative auditory function preservation despite extensive labyrinthine destruction treated at our Institution are reported. Seven cases retained cochlear function post-operatively.

Possible explanations of this occurrence and implications of related with hearing preservation in the presence of widespread inner ear destruction by chronic inflammatory disease are discussed.

Introduction

It has always been taught that destruction of any part of the membranous labyrinth by pathology or direct injury is a serious threat to the inner ear function. Even if it is true that minimal insults to the membranous labyrinth generally lead to the total loss of inner ear function, a dead ear is not always the inevitable result of such injuries. In fact a number of reports have appeared in the literature citing patients who have had preservation of inner ear function after surgical trauma or after labyrinthine invasion by cholesteatoma.

In 1911, Jenkins reported a case of hearing preservation following the surgical opening of the perilymphatic space of the lateral semicircular canal for treatment of vertigo. Preservation of hearing following accidental damage of the semicircular membranous canals during fenestration procedures was described by Jongkees (1950), Thomas (1951), Bateman (1952), Vetter (1952), Sadé-Sadowsky (1955). Cases of preservation of cochlear function after semicircular canal injury occurring during surgery for chronic ear disease were reported by Palva *et al.* (1976), Jahrsdoerfer *et al.* (1978), Cullen and Kerr (1986).

A number of reports have cited patients who had preservation of hearing before surgical treatment for extensive cholesteatoma that destroyed the labyrinth: some of them had preservation of cochlear function post-operatively despite a wide labyrinthectomy for removal of the cholesteatoma (Phelps, 1969; Bumsted *et al.*, 1977; Thomsen *et al.*, 1980; Bagger-Sjoback, 1984; Bagger-Sjoback and Phelps 1985).

It follows that, even if in most cases the destructive pathological process spreads throughout the membranous labyrinth causing a massive labyrinthitis, with loss of the inner ear function, there are however exceptions to this rule.

The scarcity of documented cases, the interest raised by the possible explanations of this uncommon evenience and the implications related to preservation of part of the inner ear function despite its involvement from chronic inflammatory pathology, have prompted us to review similar cases that occurred at the ENT Department of the University of Padua.

Materials and methods

Twelve patients with pre-operative auditory function preservation despite extensive labyrinthine destruction by cholesteatoma have been considered for the present study (Table I).

Five patients presented with a long standing chronic cholesteatomatous middle ear disease and seven with a temporal bone cholesteatoma. The cases came from a series of about 2,000 middle ear cholesteatomas treated at our Institution between 1975 and 1988 and from a series of 48 temporal bone cholesteatomas treated between 1978 and 1988. Labyrinthine invasion by the cholesteatoma matrix was revealed in some cases by high resolution CT scan but it was comfirmed in all cases during surgery. In all cases only the intra-operative findings will be considered. The pre- and post-operative masked bone conduction threshold is the average of the hearing thresholds for the frequencies 250, 500, 1000, 2000 Hz.

Results

Labyrinthine destruction involved at least two semi-

Accepted for publication: 7 September 1990.

https://doi.org/10.1017/S0022215100114446 Published online by Cambridge University Press

TABLE I

No	Pathology	Pre-op. hearing	Labyrinth invasion	Type of surgery	Labyrinth management	Post-op. hearing
1	Middle ear cholesteatoma	16 dB	L.S.C. S.S.C.	Radical mastoidect.	Bone paté and autolog. fascia	15 dB
2	Middle ear cholesteatoma	22 dB	L.S.C. S.S.C. vestibule	Radical mastoidect.	Bone paté	no hearing
3	Temporal bone cholesteatoma	22 dB	L.S.C. S.S.C.	Trans-lab. removal	Abdominal fat	no hearing
4	Temporal bone cholesteatoma	5 dB	L.S.C. S.S.C. vestibule	Trans-lab. removal	Abdominal fat	no hearing
5	Middle ear cholesteatoma	12 dB	L.S.C. S.S.C. vestibule	Radical mastoidect.	Bone paté Cartilage Palva flap	40 dB
6	Temporal bone cholesteatoma	20 dB	L.S.C. S.S.C. P.S.C.	Hemi-petrosect.	Cartilage	30 dB
7	Temporal bone cholesteatoma	13 dB	L.S.C. S.S.C. P.S.C. vestibule	Open tympanopl.	Bone paté Palva flap	40 dB
8	Temporal bone cholesteatoma	10 dB	L.S.C. S.S.C. P.S.C. vestib. Sac and duct	Closed tympanopl.	Bone paté	16 dB
9	Middle ear cholesteatoma	15 dB	L.S.C. P.S.C.	Radical	Bone paté	30 dB
10	Middle ear cholesteatoma	17 dB	L.S.C. S.S.C. P.S.C. vestibule	Open tympanopl.	Bone paté Palva flap	15 dB
11	Temporal bone cholesteatoma	20 dB	L.S.C. S.S.C. cochlea	Combined approach	Bone paté autolog. fascia	no hearing
12	Temporal bone cholesteatoma	15 dB	P.S.C. S.S.C. cochlea	Trans-lab. removal	Abdominal fat	no hearing

circular canals in all 12 cases presented herein; all semicircular canals were involved by pathology in five cases; in six cases the vestibule was involved. In one case cholesteatoma was well into the three semicircular canals, the vestibule, the sac and the endolymphatic duct. In two cases the cholesteatoma invaded the basal turn of the cochlear together with the superior semicircular canal and the lateral or posterior one.

Pre-operative bone conduction threshold in the twelve patients ranged from 5 dB to 22 dB (mean 16 dB).

The surgical procedures performed in each case for cholesteatoma removal are reported in Table I.

The residual labyrinthine defect was treated in different ways: obliteration with bone paté, cartilage, abdominal fat, sometimes associated with a Palva flap.

Cochlear function was lost post-operatively in five cases and was maintained in the remaining seven cases.

Examples of cases in which hearing was maintained post-operatively will be described in detail.

Case 1

A 40-year-old patient was seen at our Institution with a history of intermittent purulent otorrhea and progressive left hearing loss without vertigo. Examination showed a small epitympanic perforation with squamous debris and erosion of the scutum. The fistula test was uncertain. The right tympanic membrane was thickened and opaque.

Pre-operative audiogram showed a left conductive hearing loss with an average bone conduction threshold of 16 dB. Hearing was normal on right.

At surgery a wide fistula of the lateral semicircular canal was found. A large portion of the labyrinthine bone was sequestrated and surrounded by cholesteatoma matrix which at removal showed a large opening of the superior semicircular canal. The labyrinthine defect was sealed with bone-paté and a piece of autologous fascia. A radical mastoidectomy with a wide meatoplasty was performed.

Post-operatively the patient experienced moderate vertigo. Audiometric testing performed a few days after surgery showed an average bone conduction threshold of 40 dB. Ten years from operation the patient had an average bone conduction threshold of 15 dB and a dry well-healed radical cavity.

Case 8

A 33-year-old man, with a history of recurrent epi-

https://doi.org/10.1017/S0022215100114446 Published online by Cambridge University Press

sodes of otitis in the right ear since childhood, presented with recurrent ear discharge of one year duration, progressive hearing loss but no vertigo or tinnitus.

Examination showed a dry epitympanic erosion with squamous debris coming out of it. The left ear was normal.

Pre-operative audiogram showed a right conductive hearing loss with an average bone conduction threshold of 10 dB.

At surgery, cholesteatoma was invading all the semicircular canals, the vestibule, the sac and the endolymphatic duct. The matrix was completely removed and the labyrinth was obliterated with bone paté. The middle ear and adnexed cavities together with the Eustachian tube orifice were obliterated with cartilage.

Post-operatively the patient experienced vertigo and vomiting which subsided one week after surgery. Post-operative audiogram performed 15 days after surgery showed residual bone conduction threshold for the frequencies 500 and 1000 Hz at 50 and 30 dB respectively. One year after surgery the average bone conduction threshold for the frequencies 250, 500, 1000, 2000 Hz was 16 dB.

Discussion

The inner ear reaction to invasion by cholesteatoma is unpredictable: sometimes even minimal insults can lead to a complete loss of labyrinthine function, but there are cases which have retained auditory function both before and after surgical treatment despite widespread erosion of the labyrinth.

The reason why auditory function prevails in an otherwise affected labyrinth is not clear and we can only speculate how the cochlear function is spared in such conditions.

It is believed that the destructive action of cholesteatoma is greater against bone, but may respect the integrity of soft tissue structures (Bagger-Sjoback and Phelps, 1985). Direct invasion of the membranous labyrinth, which occurs slowly over long periods of time, may induce in particular cases only localized responses. According to Phelps (1969) in such circumstances a layer of granulomatous tissue divides cholesteatoma from the labyrinth which retains its function. Bumsted *et al.* (1977) suggested that parts of the labyrinth become gradually walled off because a sealing process occurs where narrow ducts connect the various parts of the inner ear. In accordance with Schuknecht and Belal (1975) the role of the utriculo-endolymphatic valve of Bast and Eyster should be outlined even if the valve is not essential to the functional integrity of the inner ear under normal conditions. The fact that the valve developed phylogenetically in association with the development of the saccule and the primitive cochlear duct indicates a probable role in maintaining the anatomical and humoral independence between the superior portion (utricle and canals) and the inferior portion (cochlear duct and saccule) of the labyrinth while preserving its accessibility to the endolymphatic duct and sac. Bast and Eyster (1935) showed that following experimentally induced collapse of the saccule and cochlear duct, the valve remained closed and the utricle and the canals remained distended. The anatomical configuration of the valve seems ideal for the purpose.

Review of the literature together with the cases presented herein confirm the unpredictability of cochlear reaction following labyrinthine damage from chronic inflammatory disease or surgical trauma. It is evident that no correlation exists between the extension of pathology and hearing preservation because nearly normal hearing has been described even in presence of massive cochlear invasion by cholesteatoma. To date it is not possible to correlate the grade of hearing preservation and the extension of labyrinthectomy required to remove the cholesteatoma matrix from the bony labyrinth. The literature and our experience confirm that removal of cholesteatoma from the cochlea always results in total hearing loss. Only one case in the literature (Bagger-Sjoback, 1984) reported a partial post-operative hearing preservation in a patient with marginal involvement of the basal turn of the cochlea by cholesteatoma that had otherwise destroyed the semicircular canals and the vestibule. Labyrinthectomy involving the vestibule may sometimes allow the preservation of hearing as reported in the literature; we had hearing preservation in four out of six cases in which the canals and the vestibule were opened at surgery. Labyrinthectomy involving only the semicircular canals should theoretically increase the possibility of hearing preservation if compared with the more destructive procedures required when the vestibule and the cochlea are invaded by pathology.

The management of the labyrinthine defect resulting from partial labyrinthectomy is aimed at sealing the residual portion of the intact labyrinth but the different proposed techniques have scarce influence on the final outcome. Nevertheless direct aspiration of lymph from the labyrinth during surgery should be avoided as far as possible to prevent further damage to the sensory-neural structures of the inner ear.

Cases with hearing preservation and concomitant inner ear invasion by cholesteatoma or cases of maintained hearing function after partial labyrinthectomy following matrix removal, should be thoroughly investigated because of the relevance in the surgical management planning.

The exact amount of labyrinthine invasion cannot be determined with certainty by CT scanning in all cases so that the surgeon should be prepared to modify the type of surgery '*a la demande*'. The retro-auricular approach is the preferred route because labyrinthectomy can be easily performed and may be extended to the cochlea with re-routing of the facial nerve.

Cholesteatoma invasion of the labyrinth should be managed with radical removal of the matrix and the surgeon should not hesitate to sacrifice the labyrinthine structures involved. Such attitude reduces the risk of residual cholesteatoma to a minimum and allows adequate management of the facial nerve which is often engulfed by cholesteatoma. The residual labyrinth should be walled off using autologous fascia or bone-patè to reduce the risks of infection.

In our opinion the above mentioned modalities are fundamental steps for complete removal of pathology and for possible post-operative preservation of inner ear function.

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Address for correspondence:
Roberto Gamoletti, M.D.,
Via Fatebenefratelli 20,
20075 Lodi (Mi)—Italy.

https://doi.org/10.1017/S0022215100114446 Published online by Cambridge University Press