Surgical Management of Petrous Apex Cholesteatoma: A Therapeutic Scheme

Abstract—From 1969 to 1990, 16 patients were treated for petrous apex cholesteatoma invading or extending beyond the internal auditory canal. This type of cholesteatoma poses various specific surgical problems and various surgical procedures are necessary for treatment. To attain maximal therapeutic effect and ensure postoperatively a satisfactory quality of life, the preoperative therapeutic scheme for each individual patient is very important, as is demonstrated by the case study presented here. The therapeutic scheme includes the following: (1) surgical approach; (2) management of a cavity following removal of the cholesteatoma; (3) handling of problems such as involvement of the dura and internal carotid artery; and (4) management of the facial nerve. (*Skull Base Surgery, Volume 2, Number 1, 1992, p. 22*)

Aural cholesteatoma may expand beyond the middle ear. It is described as temporal bone cholesteatoma, cholesteatoma extending deep into the cell tracts superior, inferior, and posterior to the bony labyrinth.^{1,2} Through expansive growth and pressure atrophy, cholesteatoma gradually invades the bony labyrinth and erodes the bone of the petrous apex of the skull base around the internal auditory canal. It may further invade the internal auditory canal and grow into the cerebellopontine angle.³ According to extension and location, Sanna et al² classified temporal bone cholesteatomas as follows: supralabyrinthine cholesteatoma, infralabyrinthine cholesteatoma, retrolabyrinthine cholesteatoma, diffuse massive cholesteatoma, and apical cholesteatoma. Fisch et al^{4,5} use the term “supra- and infralabyrinthine-apical cholesteatoma.”

From 1969 to 1990, we treated 16 patients with apical cholesteatoma invading or extending beyond the internal auditory canal. For treatment, various surgical methods were used. Our experience indicates that this type of cholesteatoma poses a number of specific surgical problems. To achieve the most beneficial therapeutic effect and give the patient satisfaction, a therapeutic scheme for each individual patient must be carefully planned, anticipating

surgical problems involved by preoperative examination. No detailed discussion on the therapeutic scheme appears in previous literature. After briefly reviewing the clinical manifestations of 16 patients, appropriate surgical procedures employed were described and one case was chosen as the most suitable to represent the importance of the therapeutic scheme. Problems involved in the scheme for surgical management of this laborious disease were then considered.

PATIENTS

The profiles of 16 patients, nine males and seven females, are presented in Table 1. Their ages ranged from 20 to 64 years, with a mean of 37.2. The lesion was unilateral in all cases. Fourteen were left-sided and the other two, right-sided. As shown in Table 2, two were assumed to be congenital and nine were acquired. In the other five, etiology could not be determined, since the cholesteatoma situated in the middle ear had been removed with surrounding structures by previous surgery.

Table 1. Clinical Symptoms of 16 Patients with Petrous Apex Cholesteatoma (1967–1990)*

Case	Age (yr)	Sex	Side	Facial Palsy	Otorrhea	Deafness	Tinnitus	Vertigo	Caloric Test
1	20	M	L	+†	+	Mixed (60 dBHL)§	+	+	Dead labyrinth
2	21	F	R	+	–	Conductive (50 dBHL)	–	–	Dead labyrinth
3	38	M	R	+†	–	Conductive (50 dBHL)	–	–	Dead labyrinth
4	49	M	L	+	–	Total	+	+	Canal paresis
5	39	F	L	+	+	Total	–	–	Canal paresis
6	25	M	L	+	–	Conductive (40 dBHL)	–	–	Canal paresis
7	38	M	L	+	+	Total	–	–	Dead labyrinth
8	25	M	L	+	–	Conductive (30 dBHL)	–	–	Normal
9	23	F	L	+†	+	Total	+	–	Not done
10	29	F	L	+	+	Total	–	–	Not done
11	40	M	L	+	–	Conductive (40 dBHL)	–	–	Not done
12	51	F	L	+	–	Mixed (75 dBHL)	+	+	Dead labyrinth
13	35	F	L	+	–	Mixed (40 dBHL)	+	–	Canal paresis
14	63	F	L	+	–	Total	+	–	Dead labyrinth
15	35	M	L	+†	–	Sensorineural (50 dBHL)	+	–	Dead labyrinth
16	64	M	L	+	–	Mixed (70 dBHL)	+	+	Not done

*M: male; F: female; R: right; L: left.

†Relapsing.

*Noted from her past history.

§HL: hearing level.

CLINICAL MANIFESTATIONS

In Table 1, the clinical manifestations of each patient are summarized. Deafness and facial palsy were common symptoms. All the patients complained of deafness to varying degrees and had facial palsy of varying duration. In three patients, facial palsy relapsed. In case 9, the palsy remitted following referral to us and in case 14, it was at the onset. In the other 14 patients, palsy was long-standing with pronounced denervation. Otorrhea was noted in four

patients. Eight patients complained of tinnitus and four of vertigo. Some complained of persistent headache.

Otoscopic findings indicated possible aural cholesteatoma, such as a white mass behind the eardrum and a retraction pocket of the eardrum. A punched out radio-lucent lesion in the petrous apex was shown by the conventional Stenvers view and polytomography of the temporal bone provided confirmation. High resolution computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone gave more accurate and decisive information on the extent of the cholesteatoma.

Table 2. Surgical Procedures of 16 Patients with Petrous Apex Cholesteatoma

Case	Etiology	Surgical Approach*	Cavity Obliteration*	Management of Facial Palsy*
1	Acquired	TL + RM	+ (Secondary)	MS
2	Acquired	TL + RM	+ (Primary)	XII–VII anastomosis
3	Acquired	MCF + L + RM	–	Nerve grafting
4	Congenital	TL + ICW	+ (Primary)	MS
5	Unknown	TL + RM	+ (Secondary)	MS
6	Acquired	TL + RM	+ (Primary)	MS + MT
7	Unknown	TL + RM	+ (Primary)	XII–VII anastomosis
8	Unknown	TL + RM	+ (Primary)	MS + MT
9	Unknown	TL + RM	–	
10	Unknown	TL + RM	–	
11	Acquired	TL + RM	+ (Secondary)	MS
12	Congenital	TL + ICW	–	
13	Acquired	TL + RM	+ (Primary)	MS + MT + TT + nerve grafting
14	Acquired	TL + ICW	+ (Primary)	
15	Acquired	MCF + TL + ICW	+ (Primary)	MS + TT + Rerouting
16	Acquired	MCF + TL + RM	+ (Primary)	MS + TT + XII–VII anastomosis

*RL: translabyrinthine approach; MCF: middle cranial fossa approach; L: labyrinthectomy; ICW: intact canal wall mastoidectomy; RM: radical mastoidectomy; MT: masseter muscle transfer; TT: temporal muscle transfer; MS: multiple suspensions.

SURGICAL PROCEDURES

Table 2 summarizes the surgical procedures for each patient. The translabyrinthine approach combined with radical mastoidectomy was conducted in ten patients. In three patients, the middle cranial fossa approach was used. In conjunction with this approach, labyrinthectomy combined with radical mastoidectomy was used in case 3, labyrinthectomy with intact canal wall mastoidectomy in case 15 and the translabyrinthine approach with radical mastoidectomy in case 16. In three patients, intact canal wall mastoidectomy was conducted with the translabyrinthine approach.

In cases 3, 9, and 10, in which the cholesteatoma was removed by the translabyrinthine approach with radical mastoidectomy, the cavity had to be left open to the orifice of the external ear canal for one or both of the following reasons: (1) The cholesteatoma matrix tightly adhering to the internal carotid artery could not be peeled off for fear of rupture of the artery; and (2) besides chronic infection, the matrix adhered extensively and tightly to the dura of the middle and posterior fossa and wall of the sigmoid sinus. In case 12, the cavity was left because noninfected cholesteatoma of congenital origin was removed without sacrificing the bony external ear canal or the ear drum. Only the dural defect in the internal auditory canal was subsequently closed by a free temporalis muscle flap.

In the other 12 patients, the temporal bone defect was obliterated as a primary or secondary procedure using an anteriorly pedicle temporalis muscle flap supplemented by fatty tissue from the lateral aspect of thigh. The orifice of the external auditory canal was closed by suturing. In eight patients, the cavity was obliterated primarily because a large cystlike cholesteatoma had been totally removed without leaving the matrix. In three patients, the cavity was obliterated secondarily, because of concomitant infection or of high suspicion of residual cholesteatoma. The cavity was left open to retroauricular wound postoperatively and cleaned for a month or two until asepsis of the cavity was confirmed by repeated bacteriologic examination. The cavity was then reexplored. Removing the granulation tissue in the cavity, the bone and dura were reexposed to check for residue of the matrix of the cholesteatoma. The cavity was obliterated only when we were fully confident of the absence of residual cholesteatoma and infection. In case 16, multiple dural tears were produced by elevating the matrix very tightly adhering to the dura of the middle fossa. Although complete removal of the cholesteatoma was doubtful, the cavity was intentionally obliterated to prevent liquorrhea. Nine months later, it was reexamined. An epidermoid cyst close to the middle fossa dura was found and removed without difficulty.

The facial nerve was preserved in 11 patients. In the other five patients, it was resected for one of the following reasons: (1) The matrix adhered to the nerve and could not be removed without being resected; (2) due to compression, the nerve underwent profound atrophy; and (3) due

to long-standing facial palsy, preservation of the damaged portion of the nerve was insignificant. In these five, facial nerve grafting was performed in two, facial hypoglossal anastomosis in two, and end-to-end suturing by rerouting in one patient. Facial reanimation was conducted in eight patients in whom return of the facial nerve function could not be assured 1 year or more following removal of the cholesteatoma.

FOLLOW-UP STUDY

Six early cases (1 through 6) of the present series were treated by one of us (NY) at Kyoto University Hospital. Until he moved to Ehime University in 1976, these six patients had been followed up with no recurrence of cholesteatoma. The other ten cases (7 through 16) were treated at Ehime University with checks once or twice a year. The average follow-up period was 3 years. Until now, no recurrence of cholesteatoma or postoperative complications have been noted.

REPORT OF CASE 10

The patient was a 29-year-old woman with sudden hearing loss and tinnitus of the left ear at 12 years of age. At 15 years of age, she had acute otitis media of the left ear, which was once controlled by medication. She had been bothered by persistent otorrhea since the recurrence of otitis media at 17 years of age. One year later, she consulted a neurosurgeon due to severe headaches and disequilibrium. A temporal bone cholesteatoma extending to the cerebellopontine angle was found and tumor resection was conducted by suboccipital craniectomy but without notable improvement. Radical mastoidectomy was then carried out by an otologist, again without beneficial result. After the operation, even facial palsy developed. The facial palsy for a time improved naturally, but recurred within a few months. The following year, another otologist attempted to remove the cholesteatoma but without success. Facial palsy has remained total and the patient gave up therapy.

On September 30, 1985, she was referred to us with complaints of headache, otorrhea, and facial disfigurement. Otoscopy indicated a large cavity of the temporal bone with draining otorrhea of foul odor. All middle ear structures were missing. Medial to the deformed bony block, a large cholesteatoma mass was seen. As shown in Figure 1, an axial view by CT disclosed a large cholesteatoma mass occupying the entire apical region of the petrous pyramid. The posterior bony wall of the petrous pyramid was eroded and the internal auditory canal was filled with the mass. An audiogram demonstrated total deafness of the left ear. Facial palsy was total. Electrodiagnostic testing indicated profound denervation. Bacteriologic examination of the discharge showed infection of *Pseudomonas aeruginosa*.

Figure 1. Axial view of the preoperative computed tomography scanning indicates a large cholesteatoma mass occupying the petrous apex (asterisk) and medial to the eroded labyrinthine block (arrow). The cholesteatoma involved the internal auditory canal and the internal carotid artery.

The patient's condition was considered to be due to failure in conducting an appropriate therapeutic scheme. We informed her that at least three operations would be necessary to improve her condition and eliminate the serious complications. On November 12, 1985, the first operation was conducted. By a retroauricular transmastoid approach, the mass in the apical region with dirty granulation tissue due to osteitis was removed as much as possible, leaving the labyrinthine bone block. Posterior fossa and middle fossa dura, internal auditory canal and internal carotid artery were left untouched. The retroauricular wound was left open and cleaned by antibiotic solution. A month later, wound secretion was bacteriologically negative. The second surgery was done on December 24, 1985. By an extensive translabyrinthine approach, the entire cholesteatoma mass in the entire petrous apex was removed. The internal auditory canal was widely opened and the mass extending to the canal was eradicated. The facial nerve was sacrificed. The internal auditory canal was closed by layers of fascia lata using cyanoacrylate glue. The matrix adhering to the dura in the depth of petrous apex and the internal carotid artery could not be removed and thus the large cavity had to be left open to the orifice of the external ear canal so as to close the retroauricular wound. To prevent liquorrhea, a spinal drainage was left inserted for a week. Fortunately, the dural defect was closed and the cavity became clean and dry 2 months postoperatively. Facial rehabilitation operation was then carried out as a multiple facial suspension operation along with temporalis muscle transfer to the

orbicularis oculi and masseter muscle transfer to the orbicularis oris.

Postoperatively, she has ceased to have headaches and disequilibrium. In about 3 months, the large cavity shown in Figure 2 epithelized and was perfectly dry. A year later, good tone and some movement of the involved face were restored. Every half a year, debris in the cavity must be removed, but the patient is satisfied with the results. She has been followed for 6 years.

DISCUSSION

Petrous apex cholesteatoma may arise from an acquired middle ear cholesteatoma or embryonic epithelial remnant. It must also be remembered that iatrogenic failure of the middle ear surgery may lead to apical cholesteatoma.² Findings attributable to otitis media such as chronic otorrhea, retraction of the ear drum, and inhibited pneumatization of the mastoid air cells indicate possible acquired origin. The congenital cholesteatoma develops behind the intact tympanic membrane without manifestation of preceding otitis media. However, their distinction is not always easy owing to inflammation of the middle ear that may often be associated with congenital cholesteatoma.⁶

The complete removal of the cholesteatoma is of prime importance in the treatment of cholesteatoma in the temporal bone. In petrous apex cholesteatoma, no single surgical approach will adequately achieve this purpose.

Figure 2. Axial view of the postoperative computed tomography scanning of petrous bone. All the bony structures in the petrous bone were removed together with cholesteatoma. The cavity was open to the orifice of the external ear canal.

Conducting the operation in stages is frequently required to prevent complications such as liquorrhea and meningitis. Management of the large cavity following removal of the cholesteatoma and rehabilitation of facial expression must also be included in the therapeutic scheme to ensure good postoperative quality of life.

Surgical Approach

The translabyrinthine approach with or without the transcochlear approach is the basic technique for removal of petrous apex cholesteatoma. However, this approach alone may not allow sufficient working space when the cholesteatoma is situated deep in the petrous apex and its matrix tightly adheres to the middle fossa dura extensively. In such a situation, the middle cranial fossa approach should also be conducted in that the entire superior surface of the petrous pyramid can be seen from above and the matrix situated in a very deep apical area or adhering to the middle cranial fossa dura can be removed safely.

Preservation of cochlear and vestibular functions is usually not considered when eradicating the cholesteatoma in the petrous apex. As shown in Table 1, caloric response is so severely impaired in most patients that labyrinthectomy would not lead to additional postoperative distress. Exceptionally useful hearing and normal caloric response are preserved despite considerable bony erosion of the otic capsule. Although the middle cranial fossa approach along with intact canal wall mastoidectomy or radical mastoidectomy is reasonable in such a

case, the preserved labyrinthine block limits the surgical view and causes residue of the cholesteatoma in the depth of the petrous apex. Once cholesteatoma regrows, preservation of function will eventually cease to be possible. Total labyrinthectomy even in cases with near normal cochleovestibular function should be conducted whenever complete removal of the matrix does not appear possible.

Removal of the Posterior External Ear Canal and Ear Drum: the Cavity Problem

In limited congenital cholesteatoma, the middle ear does not incur severe invasion of cholesteatoma and the cholesteatoma can be removed without sacrificing the posterior ear canal. However, in most cases, radical mastoidectomy should be conducted for wide field exposure to ensure radical removal of the cholesteatoma in the middle ear, particularly when the matrix adheres to the dura of the posterior fossa, sigmoid sinus, or jugular bulb. Should the radical removal of cholesteatoma by preserving the posterior external ear canal and ear drum not appear advisable, these structures must be sacrificed so as to facilitate eradication of the cholesteatoma.

Closure of Temporal Bone Defect to Prevent Liquorrhea and Meningitis

For the complete removal of the cholesteatoma matrix, the involved dura of the internal auditory canal must

be resected and resultant dural defect closed using a sheet of fascia. The temporal bone defect is then obliterated by bulky fat from the abdominal wall or lateral side of the thigh. If the posterior wall of the external ear canal is sacrificed for radical mastoidectomy, the orifice of the external ear canal is closed by suturing after eliminating the canal skin and ear drum. If the cholesteatoma is contaminated with infection, removal of the cholesteatoma mass around the internal auditory canal is delayed and local and systemic antibiotic treatment should be continued until the cavity is found to be aseptic. When a dural defect is closed by a sheet of fascia, a spinal drainage should be left in place for a week to prevent meningitis. Spinal drainage is quite valuable when the cavity must remain open for fear of meningitis or a hidden residue of the cholesteatoma matrix.

Involvement of Dura and Internal Carotid Artery

In the present series of patients, either posterior fossa or middle fossa dura or both were involved in the cholesteatoma. Usually, the matrix could be peeled off from the dura. If a microresidue is probable, the involved area should be carefully electrocoagulated. In three of the 16 patients, the matrix adhered tightly and extensively to the dura of both the middle fossa and posterior fossa. For fear of liquorrhea and meningitis, the matrix was left untouched and the cavity remained open. In one case, the middle fossa dura was so atrophic that multiple dural tears resulted. The cavity was obliterated. A revision operation was conducted 9 months later and the residual cholesteatoma was removed without difficulty.

Involvement of the internal carotid artery is a very serious problem. In two patients, the matrix adhered to the artery in its intracanalicular portion and was difficult to separate from the arterial wall. The cavity had to be left open in these patients.

Management of the Facial Nerve

Preservation of the facial nerve will no longer be possible when the nerve is severely damaged and has undergone pronounced denervation for a long period of

time. Even the facial nerve retaining good function must eventually be sacrificed for complete removal of the cholesteatoma. For restoration of facial expression or facial symmetry, three fundamental procedures (nerve grafting, hypoglossal-facial anastomosis, and facial reanimation procedure) are available. Selection of the procedure is based on the severity of damage, length and location of the damaged segment, and duration of facial palsy. The gap of the facial nerve is bridged by nerve grafting, using the greater auricular nerve. For long-standing palsy of a year or two, hypoglossal-facial anastomosis with adjunctive facial reanimation should provide satisfaction. In our experience, facial suspension using multiple strips of Gore-Tex, temporalis muscle transfer, and masseter muscle transfer have been found best for facial reanimation.^{7,8}

Follow-up Study

Long-term follow-up study with MRI is quite helpful to detect recurrence. Although recurrence is infrequent when the cholesteatoma is eradicated according to the procedure described, MRI once a year is advisable for at least 5 years or more postoperatively.

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