Petrous Temporal Bone Cholesteatoma: A New Classification and Long-Term Surgical Outcomes

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ABSTRACT

The goals of this retrospective case review were to analyze the long-term results of surgery for petrous temporal bone cholesteatomas and to propose a new classification system for these lesions. Patients with a surgically confirmed petrous temporal bone cholesteatoma were treated at Addenbrooke's Hospital, a tertiary referral center. Postoperative facial function, hearing, residual/recurrent cholesteatoma, and other complications were assessed in relation to preoperative signs, intraoperative findings, and surgical approach. Between 1983 and 2004, 43 patients were treated. There were no perioperative deaths. There was no long-term recurrence in 95.4% of the patients, possibly because of meticulous surgical technique, bipolar diathermy, and use of the laser to denature the cholesteatoma matrix that was adherent to the dura. At presentation, 95% of the patients had no socially useful hearing in the affected ear. Facial nerve function, however, was usually preserved. Both direct anastomosis and nerve grafting can improve facial nerve function from House-Brackmann grade VI to grade III if the palsy is not longstanding. Four patients had cerebrospinal fluid leakage; other complications were rare. The proposed classification facilitates surgical planning and predicts the postoperative outcome with regards to hearing.

KEYWORDS: Petrosal cholesteatoma classification, hearing

Petrous temporal bone cholesteatomas are defined as cholesteatomas medial to the otic capsule and account for 4 to 9% of all petrous pyramid

lesions.^{1,2} Unfortunately, these lesions often invade the labyrinth and fallopian canal before they are diagnosed. They can be congenital or, more

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frequently, acquired. Histologically the two are indistinguishable. Congenital cholesteatomas are thought to arise from embryonic rests deposited in the temporal bone after the abnormal development of the first branchial groove. In contrast, acquired cholesteatomas originate in the tympanomastoid region and erode medially around or through the labyrinth. Cholesteatomas are locally destructive and erode bone. A number of mechanisms have been proposed to account for this behavior, including secretion of osteolytic enzymes, pressure necrosis, and osteitis and surrounding chronic granulation tissue.^{3,4}

Preoperatively, the size, morphology, and anatomical site of the cholesteatomas are assessed by various imaging techniques, including high-resolution axial and coronal computed tomography (CT) and T2-weighted, T1-weighted, and B1000 diffusion-weighted magnetic resonance imaging (MRI). Obliteration of the cavity and blind sac closure techniques has increased the importance of postoperative imaging. The more traditional open cavity technique has become less popular because of the potential for an otic discharge. Open techniques are also associated with a higher incidence of cerebrospinal fluid (CSF) leakage and intracranial infections. Furthermore, the medial aspect of the open cavity seals once the disease is medial to the labyrinth.

Classification of petrosal cholesteatomas has evolved from the two classes (supralabyrinthine and infralabyrinthine-apical) proposed by Fisch,⁵ to the additional translabyrinthine category proposed by Bartels,⁶ to the five classes proposed by Sanna et al⁷ (supralabyrinthine, infralabyrinthine, massive labyrinthine, infralabyrinthine-apical, and apical). Perhaps because our series is the largest yet, it became apparent that none of the existing classifications were adequate. We therefore propose a new Moffat-Smith classification. This classification system is more comprehensive than existing systems. Lesions involving the petrous apex and intracranial extension can be classified if necessary.

MATERIALS AND METHODS

Between 1983 and 2004, 43 patients (25 males, 18 females; median age, 34 years; age range, 7 to 75 years) with petrous temporal bone cholesteatomas referred to the Department of Otolaryngology and Skull Base Surgery at Addenbrooke's Hospital, Cambridge, were reviewed retrospectively. All the patients had surgical documentation of a cholesteatoma within the petrous bone medial to the otic capsule. Ten patients had undergone previous mastoid surgery elsewhere. The presenting signs and symptoms of all patients were documented, and CT scans were available to show the extent of disease.

Topographically, the petrous temporal bone cholesteatomas were grouped according to the proposed Moffat-Smith classification (Fig. 1).

Postoperatively, patients were regularly followed both clinically and radiologically with high-definition CT, MRI, and B1000 diffusion-weighted scans. Scanning was indicated if clinical signs or symptoms suggested residual or recurrent disease. If the petrous cavity had been obliterated, patients underwent imaging annually. The median follow-up was 10 years (range, 9 months to 23 years); however, patients must be followed for life.

Pre- and postoperative morbidity, including hearing loss (air conduction measured by pure tone audiometry using the modified American Academy of Ophthalmology and Otolaryngology [AAOO] criteria of 4 rather than 3 KHz) and other cranial nerve deficits (as assessed at last consultation) were evaluated. Facial nerve function was graded according to the House-Brackmann (H-B) grading system.⁸ The site of cholesteatoma on the facial nerve was recorded as being proximal at or distal to the geniculate ganglion. The presence of CSF leakage and residual or recurrent cholesteatoma during follow-up were recorded.

The most common symptom was hearing loss (26 patients) (Table 1). However, significant hearing loss measured by pure tone audiometry was found in 95% of the patients at presentation. The second most common symptom was facial nerve palsy with vertigo/imbalance and otorrhea occurring less

Figure 1 Moffat-Smith classification. Supralabyrinthine, above the labyrinth; supralabyrinthine-apical, above the labyrinth extending to the petrous apex; infralabyrinthine, below the labyrinth; infralabyrinthine-apical, below labyrinth extending to petrous apex; massive labyrinthine, extensive destruction of inner ear; massive labyrinthine-apical, extensive destruction of inner ear extending to petrous apex; apical, confined to petrous apex.

frequently. Recurrent bacterial meningitis, sixth nerve palsy, and trigeminal neuralgia each occurred in one patient. The mean duration from the onset of symptoms to surgery was 18 months, but this period

varied from 3 days to 20 years. Once referred, the mean latency to surgery was 6 months (range, 0 to 22 months). Twenty-four operations were performed on the left side and 19 were performed on the right

Table 1 Presenting Symptoms in 43 Patients with Petrosal Cholesteatomas

Symptom	No. of Patients (%)
Hearing loss	26 (60)
Facial palsy	22 (51)
Vertigo/imbalance	14 (33)
Discharge	14 (33)
Tinnitus	5 (12)
Otalgia	3 (4)
Headaches	2 (5)

side. Ten patients had had surgery elsewhere, and almost a third (13 patients) had a history of chronic otitis media. One patient had a history of trauma. Most patients had a massive labyrinthine lesion with or without apical extension (Table 2). Only 14% were suitable for surgery to preserve the otic capsule.

RESULTS

There were no perioperative deaths, and 95.4% of the patients had no long-term recurrence. Two patients underwent re-exploration for a recurrence. One year after surgery in one patient the tegmen was eroded with a large pocket into the middle fossa. The other recurrence was found 6 years after surgery. Two years after surgery, two other patients underwent surgical re-exploration; one for a delayed

Table 2 Surgical Approaches Used to Treat 43 Petrous Temporal Bone Cholesteatomas Grouped According to Moffat-Smith Classification

Classification	No. of Patients	Surgical Approach/ No. of Patients
Supralabyrinthine	6	TT-SL (+ 1 with MF)/4
Supralabyrinthine-apical	3	TT-SL/3
Infralabyrinthine	1	ST P/1
Infralabyrinthine-apical	3	ST P/3
Massive labyrinthine	10	ST P/2 Transotic/4 Transcochlear/1 IT type A/3
Massive labyrinthine-apical	17	ST P (=1 with MF)/16
Apical	3	ST P/3

TT-SL, transtemporal-supralabyrinthine; MF, middle fossa; ST P, subtotal petrosectomy; IT type A, infratemporal type A approach.

CSF leakage and the other for a retraction pocket where no cholesteatoma was found.

Of the 21 patients with normal preoperative facial nerve function, only two deteriorated (Fig. 2): one patient to grade II and the other to grade III. Preoperatively, two patients had H-B grade II; one patient this improved to grade I, and the other maintained at grade II. Most patients who were grade III before surgery remained at this level after surgery. However, one patient improved to grade I and one patient deteriorated to grade VI. The one patient who was H-B grade IV before

Figure 2 Comparison of the pre- and postoperative facial nerve function according to House-Brackmann grades.

Figure 3 Comparison of the pre- and postoperative hearing levels as measured by mean air conduction thresholds.

surgery deteriorated to grade VI after surgery. Both patients who were H-B grade V before surgery improved: one to grade II and the other to grade IV. More than half of the patients who were H-B grade VI before surgery improved to grade III after surgery. One grade VI patient improved to grade II after primary anastomosis and nerve grafting. Other techniques used to treat patients with impaired facial nerve function included static slings, gold weights, and tarsorrhaphy (Table 3). In summary, if the preoperative facial nerve function was good; the likelihood of preserving facial nerve function after surgery was high.

The otic capsule was preserved in only 14% of the patients; therefore, only six patients had residual hearing (Fig. 3).

Postoperatively, four patients developed CSF leaks. The CSF leaks were managed by surgical exploration in one patient. Lumbar drains were

inserted for 5 days in two patients, and one patient required repeated lumbar punctures. One patient developed deep vein thrombosis (DVT). One patient experienced a septal cardiac infarction 6 days after surgery. One patient developed seizures and another a wound infection. Secondary procedures included resuturing a wound and the prophylactic insertion of lumbar drains at surgery in four patients.

DISCUSSION

This is the largest series of petrous bone cholesteatomas reported in the world literature to date. The most common presenting symptom, hearing loss in the affected ear, occurred in 60% of our patients. However, as measured by pure tone audiometry at presentation, 95% had a significant hearing loss. These findings are consistent with reported rates of hearing loss of (64 to 100%) in other series.^{3,9-11} In this series, 51% had facial nerve weakness. Rates for facial nerve weakness preoperatively varies in series from 20 to 64%^{3,7,9-12} Vertigo/imbalance and discharge from the ear affected almost a third of our patients at presentation before surgery, but Burggraaff and coworkers reported that 61% of their patients suffered dizziness before surgery.¹¹ Tinnitus, otalgia, and headaches were less common manifestations.

A diagnosis of petrosal cholesteatoma must be considered in any patient presenting with recurrent bacterial meningitis, a sixth cranial nerve palsy,

Table 3 Facial Nerve Management

Procedure	No. of Patients
Dynamic	
Primary anastomosis	5
Great auricular nerve (initial surgery)	1
Sural nerve graft (13 months)	1
Static	
Fascia lata sling (18 months)	1
Temporalis hitch (42 months)	1
Gold weights	4*
Tarsorrhaphy	10†

*Two were revised once, one was revised twice, and two were removed.

†Five were later divided.

or trigeminal neuralgia.³ It is apparent from our series and others that patients can have symptoms for as long as 20 years.³ The diagnosis was delayed in a quarter of patients who had undergone surgery for assumed tympanomastoid disease. Others had their otorrhea treated medically. The mean wait for surgery in our department was 6 months, but immediate surgery was performed when clinically indicated.

Advances in CT and MRI have improved the detection and definition of the extent of the cholesteatomas, thereby improving the accuracy of preoperative assessment. High-resolution CT provides accurate details of the location of the disease, which appears as a "punched-out" or scalloped area of bone destruction. It has also been associated with elevation of the petrous ridge.¹³ MRI differentiates cholesteatomas from other lesions such as cholesterol granulomas. Cholesteatomas are only hyperintense on T2-weighted MRIs,¹³ whereas cholesterol granulomas are hyperintense on both T1- and T2-weighted MRIs. The use of imaging before any mastoid surgery is performed would allow a petrosal cholesteatoma to be identified and hence result in earlier referral to a tertiary center. Earlier detection of cholesteatomas may reduce both preoperative and postoperative morbidity.

The routine use of CT scanning before any cholesteatoma surgery is important for determining the extent of bone erosion and the prognosis, which are critical for surgical planning and for obtaining informed consent for treatment. Preoperative CT is recommended for all patients with a history of facial nerve palsy, headaches, dizziness, or sensorineural hearing loss.¹¹ B1000 diffusion-weighted MRI is a promising technique for detecting recurrences or residual disease without resorting to further surgery. The advent of non-echo-planar imaging (NON EPI) is exciting. This modality can differentiate cholesteatomas from cholesterol cysts and mucosal thickening (Moffat D, unpublished observation).

As found by other authors, supralabyrinthine involvement was common and direct labyrinthine involvement was even more so.^{14,15} In 1969, Glasscock and coworkers described the middle fossa approach, which can be used successfully to remove

supralabyrinthine cholesteatomas that extend toward the petrous apex.¹⁴ We used the transtemporal supralabyrinthine approach in patients with supralabyrinthine lesions with or without an apical extension. The transtemporal approach has two main advantages. It is the most direct route to the pathology, and bone removal avoids dural retraction. A variety of approaches can be used in patients with massive labyrinthine cholesteatomas. A subtotal petrosectomy is appropriate for infralabyrinthine lesions with or without an apical extension. The transotic (translabyrinthine-transcochlear) and transcochlear approaches enable access to the petrous apex and clivus.^{16,17} Fisch described several infratemporal fossa approaches to access more anterior structures, including the internal carotid artery.¹⁸ We used this approach to resect massive labyrinthine lesions. Massive labyrinthine lesions with apical extensions and apical cholesteatomas were removed using a subtotal petrosectomy approach.

Our proposed classification allows logical surgical planning based on extent of disease and the involved anatomical areas. The goals of surgery for treatment of supralabyrinthine disease are preservation of preoperative hearing and removal of part of the middle fossa plate and squamosa. Superior retraction of the middle fossa dura using a Yasargil flexible arm retractor permits visualization and dissection of the cholesteatoma matrix from the otic capsule and middle fossa dura. An apical extension also can be excised through this approach. If, however, the cholesteatoma is extensive, it may be necessary to combine a supralabyrinthine approach with a classical middle fossa craniotomy. Only infralabyrinthine cholesteatomas can be excised via a subtotal temporal bone resection. The inferior portion of the temporal bone is removed, and this dissection can be extended to the petrous apex.

The approach to massive labyrinthine cholesteatomas depends on the extent of the anatomical site and on the difficulties of access and visualization. Various approaches such as the transotic, transcochlear, subtotal petrosectomy and a classical infratemporal Fisch type A approach can be

considered. Massive labyrinthine apical cholesteatomas require a subtotal petrosectomy possibly combined with a middle fossa craniotomy. Again, only apical cholesteatomas require subtotal petrosectomy, although approaches via the middle cranial fossa and a retrosigmoid approach have been described. Thus, the grade of a cholesteatoma will determine the approach to use.

In this series, there were no perioperative deaths and 95.4% had no long-term recurrence. Two patients developed recurrence. One was detected at 1 year after initial surgery; the other was found at 6 years after initial surgery. Consequently, these patients are never discharged even though a recurrence after petrous apex surgery is uncommon.¹⁴ Macroscopically, it can be very difficult to remove a cholesteatoma matrix completely from dura, facial nerve, and carotid artery. Some surgeons have advocated wide excision of dura-bearing matrix and closure of the dural defect with connective tissue grafts. However, graft necrosis can lead to CSF leakage or infection, and this technique has now been abandoned.¹⁵ At surgery, cholesteatomas adherent to dura were managed by the accepted technique of bipolar diathermy,^{2,7} and since 1995 we have used a diode laser with a defocused beam to denature the matrix.

B1000 diffusion-weighted MRI is used to aid detection of recurrence and is performed on all patients annually for 3 years, every 2 years for the next 6 years, and every 3 years thereafter unless patients develop symptoms suggestive of a recurrence. Because only two patients had a recurrence, it is difficult to predict the level of postoperative morbidity associated with the surgical approach to any given grade of lesion. However, one could anticipate a recurrence when visualization is difficult during attempts to preserve the otic capsule and when it is difficult to clear cholesteatoma matrix from the dura.

In only 14% of the cholesteatomas could the otic capsule be preserved. Only 3 of the 43 patients had normal preoperative hearing, but their supralabyrinthine cholesteatomas extended to the petrous apex and required a transtemporal

supralabyrinthine approach. Few patients had useful hearing in the affected ear at presentation. Residual hearing was sacrificed to enable sufficient access to remove the cholesteatoma. This finding is consistent with the results of other studies, although Bartels preserved hearing in 34% of patients treated surgically.⁶ When a cholesteatoma involves the only hearing ear, an open technique has been recommended. Exteriorizing the disease rather than excising it allows hearing to be preserved. Such patients must be monitored closely to avoid complications.^{7,14,18}

Facial paresis is the most debilitating long-term complication associated with petrosal cholesteatomas. Preoperatively, only 21 (49%) of the patients had H-B grade I function. This level was maintained in all but two patients who deteriorated to H-B grades II and III after surgery. Of the two patients who were H-B grade II preoperatively, one improved to grade I and the other remained H-B grade II. Their outcomes suggest that if the facial nerve is functioning well before surgery, surgeons can be optimistic that facial nerve function can be preserved after surgery. Outcomes of patients with poor preoperative facial nerve function (H-B grades III–VI) were less predictable, resulting in H-B grades II and IV after surgery.

Axon et al³ suggested that nerve ischemia occurs when the cholesteatoma involves the geniculate ganglion. Sanna et al⁷ noted that the facial nerve was present as a fibrous band at the geniculate ganglion. We think that involvement of the intralabyrinthine segment of the facial nerve is significant in the development of ischemia because this segment of the nerve has the poorest blood supply. Postoperatively, more than half of the patients who were H-B grade VI who underwent primary anastomosis or nerve grafting achieved H-B grade III, and one patient achieved H-B grade II.

Because the motor end plate atrophies after 1 year, the duration of preoperative facial paralysis affects the likelihood of recovery of function postoperatively despite aggressive treatment. Various techniques involving gold weights and tarsorrhaphy have been used to reduce the morbidity associated with facial paresis (Table 3). In half the patients,

these measures were temporary until facial nerve function recovered sufficiently.

Can any conclusions be drawn about which patients' facial nerve function will improve? The difficulty is that the series spans 23 years. In the early years, the decision of whether to decompress the nerve or whether to anastomose, or whether to interpose a cable graft depended on the surgeon's intraoperative assessment. If 50% or more of the cross-sectional area of the nerve was visibly damaged, excision and mobilization were pursued followed by direct anastomosis or placement of an interposition cable graft. If less than 50% of the cross-sectional area of the nerve was visibly damaged, decompression was performed. In recent years, it has become clear that patients with H-B grade IV or worse may do better with a graft. Only one of the four patients with a postoperative CSF leak required surgery. Other complications, including DVT, cardiac infarction, seizures, and wound infection, have been rare.

Traditionally, open cavities were fashioned to enable visualization of a potentially unsafe ear. However, these procedures may result in postoperative CSF leakage, otorrhea, otalgia, intracranial infections, and trauma to intrapetrous structures during aural toilet. CT and MRI, including B1000 diffusion-weighted MRI, enable detection of a cholesteatoma even if fat obliterates the mastoid cavity. Patients appear to remain asymptomatic with complete obliteration and blind sac closure, and this procedure is now preferred by several authors.^{7,9,15,17-21} Others still use an open cavity technique, especially if the matrix cannot be removed completely.^{12,14} However, an open cavity will not remain exteriorized if it is medial to the otic capsule; the pyramidal shape of the temporal bone will cause it to seal.

CONCLUSION

The best surgical approach to a cholesteatoma is based on several factors, including the extent and

location of the disease, the presence of hearing and facial nerve function, and anatomical variation. Our proposed classification indicates the optimal surgical approach and therefore preoperatively predicts the level of surgical morbidity. The classification also enables results from different series to be compared. The recurrence rate in our series was low. Facial nerve function can usually be preserved. Preoperative facial nerve weakness may be improved by decompression or by using both primary anastomosis or nerve grafting techniques. However, remaining hearing in the affected ear may need to be sacrificed to enable complete removal of a cholesteatoma.

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Commentary

Moffat et al present 43 patients surgically treated for petrous bone cholesteatomas. Given that 10 patients had prior surgery and 13 suffered from chronic otitis media, their population was complex. Nonetheless, only four patients developed cerebrospinal fluid leakage, and the House-Brackmann grade of only four patients worsened across follow-up. Based on their rates of hearing preservation, the authors favor aggressive resection to preserve residual hearing. However, fewer than 5% of their patients had serviceable preoperative hearing. The follow-up was extremely long, with a median of more than 10 years. Overall, these impressive patient outcomes and anatomically based classification scheme showcase the senior author's tremendous clinical experience with this challenging entity.

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