

Petrous Bone Cholesteatoma: Classification, Management and Review of the Literature

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Abstract

Introduction: Petrous bone cholesteatoma (PBC) is defined as cholesteatoma extending medial to otic capsule. Because of its relations with vital structures, surgical management is generally difficult. Objective: In this study, we aim to share our clinical experiences about surgical approaches, managements and follow up results in petrous bone cholesteatoma.

Methods: A retrospective clinical study was conducted on patients who had been diagnosed as PBC between 2003 and 2012 years. Sanna classification was used for the pathology extending. A total of 13 patients with 14 ears were evaluated about symptoms, clinical and radiological findings, surgical techniques, complications and follow up results.

Results: The mean age of patients was 33,3 years ranging from 18 to 52 years. Hearing loss was the most common symptom (92 %). Five patients had facial paralysis preoperatively. Massive type was the most common type. Different surgical techniques were carried out for each pathological type. Mean follow up time was 34,3 months. Recurrence was identified in one patient twice within two years and revision surgical procedures were performed.

Conclusion: Extent of the PBC should be well defined in preoperative imaging and type of surgical approach should be planned before with an appropriate classification. Regular follow up is mandatory for patients with MRI every year.

Keywords: Petrous bone; Cholesteatoma; Hearing loss

Introduction

Petrous bone cholesteatoma (PBC) is defined as cholesteatoma located medial to the otic capsule. It is an uncommon pathology and because of functional damages in the treatment it diminishes the patients' quality of life [1]. Diagnosis and surgical approach may be challenging. Advanced imaging techniques are necessary for assessment. This lesion generally has a slow-growing behavior, so it may remain asymptomatic for years. It can be congenital or acquired. Our aim with this study is to report our management and surgical experiences of fourteen cases diagnosed with PBC.

Material and Methods

This is a retrospective clinical trial. Inclusion criteria were the patients had been operated with petrous bone cholesteatoma and had long-term follow-up results. Patients with limited cholesteatoma, and recently operated were excluded. Patients diagnosed with and operated from PBC between 2003 and 2012 were analyzed retrospectively. The Ethics Committee of our institution approved the study (2015/87) and written informed

consent was taken. A total of 14 cases (13 patients) were included in this study. All patients underwent through clinical otoneurological and audiologic examinations. All patients evaluated by preoperative computed tomography (CT), temporal bone magnetic resonance imaging (MRI) and/or if necessary magnetic resonance venography (MRV) imaging. House - Brackmann (H-B) classification system was used to evaluate the facial nerve functions. Our surgical approaches, outcomes, recurrences, complications and follow up results were evaluated with the review of the literature.

Results

Patients consisted of 12 men and one woman. One patient had bilateral PBC. Mean age was 33.3 years ranging from 18 to 52. All patients were adults and patients diagnosed as acquired cholesteatoma had chronic ear disease like otorrhea, tympanic membrane retraction or perforation. Only one patient had intact tympanic membrane and no history of ear disease. Five of them had surgery for cholesteatoma before. Postoperative

follow-up period was on average 34.3 months ranging from 12 months to 8 years. The most common symptom was hearing loss (92%) followed by otorrhea (78%). Other symptoms were facial paralysis (35%), dizziness (35%), tinnitus and headache in descending order of frequency. Ear side numbers were equal (7 left sided, 7 right sided) (Table 1). Two patients had cerebellum abscess and sigmoid sinus thrombosis preoperatively. Sanna classification [2] was used to evaluate the petrous bone cholesteatomas (Table 2). Massive type was the most common type in our study and supralabyrinthine, infralabyrinthine and apical type (Figures 1-4) were observed respectively (Table 3). Surgical approaches performed for the patients are listed in Tables 4 & 5. Recurrence was seen in one patient twice within two years and revision surgical procedures were performed. Facial paralysis was observed in five patients preoperatively. According to H-B classification system, grade II facial palsy was detected in one patient, grade III in one patient and grade VI facial palsy in three patients. Facial nerve could not be protected in patient who had one and a half months history of grade III facial palsy, so facial nerve grafting was performed for the patient. Facial nerve was decompressed in patient with grade II. The remaining three patients who had fibrotic nerve and long duration of palsy received static reconstruction of the face.

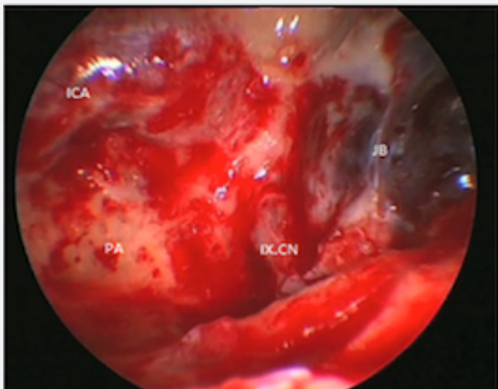


Figure 1: Apical type PBC. Cholesteatoma located petrous apex is marked with white arrow, also seen clivus extension at medial.

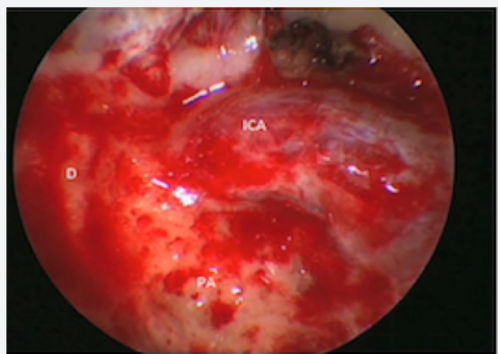


Figure 2: CT image of the patient that had bilateral petrous bone cholesteatoma. Especially on the right, it is seen that the massive type of cholesteatoma eroded the dura.

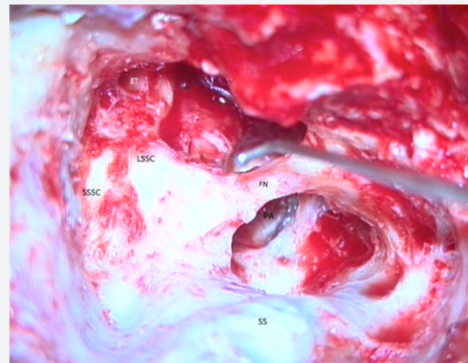


Figure 3: MR image of the patient that had bilateral petrous bone cholesteatoma. Indicated by the white arrow on the left cholesteatoma located superior to the labyrinth. Supralabyrinthine type petrous bone cholesteatoma.

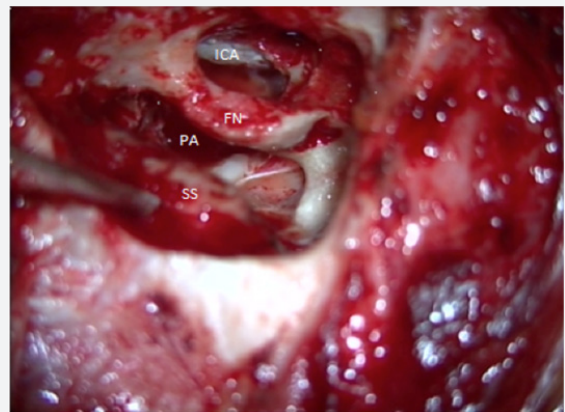


Figure 4: Coronal section of CT, infralabyrinthine type.

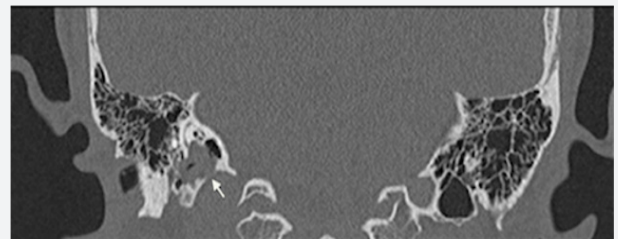


Figure 5: Massive type cholesteatoma excision by transotic approach. FN: Facial nerve, PA: Petrous apex, SS: Sigmoid sinus, ICA: Internal Carotid Artery.

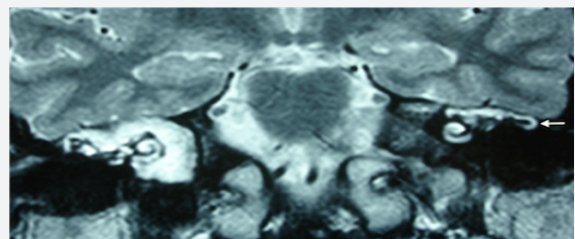


Figure 6: Subtotal Petrosectomy. LSSC: Lateral Semisircular canal, SSSC: Superior Semisircular Canal, ET: Eustachian Tube, FN: Fasiyal Nerve, ICA: İnternal Carotid Artery, PA: Petrous Apex, SS: Sigmoid Sinus.

In terms of the surgical approach to remove the cholesteatoma matrix; modified trans cochlear approach type A (MTCA) was used for three patients, enlarged trans labyrinthine approach (ETLA) for four patients, transotic approach (TO) for three patients (Figure 5), modified trans cochlear approach type B (MTCB) for one patient, combined middle fossa transmastoid approach for one, transmastoid retrolabyrinthine approach for one patient and subtotal petrosectomy (Figure 6) operation for

one patient were performed. We performed the right subtotal petrosectomy in a patient with infralabyrinthine type. This surgical procedure included after canal wall down mastoidectomy and complete drilling of the air cells of the temporal bone was carried out, the cholesteatoma sac was followed and retrofacial, retrolabyrinthine, infralabyrinthine, peritubal, and pericarotid air cells were removed and only a few cells in the petrous apex left behind.

Table 1: Clinical findings of 13 patients with petrous bone cholesteatoma.

Case no	Age (years)	Sex	Side	Symptoms	Etiology	H-B Grade
1	34	M	L	HL1, otorrhea	Acquired	
2	30	M	L	HL, otorrhea	Congenital	
3	26	M	L	HL, FP2, otorrhea	Recurrent/Iatrogenic	Grade III
4	50	M	L	HL, FP, otorrhea, dizziness	Acquired	Grade VI
5	18	M	L	HL, otorrhea	Recurrent/Iatrogenic	
6	23	M	R, L	HL, FP, otorrhea	Recurrent/Iatrogenic	Grade VI
7	52	M	R	HL, FP, otorrhea, dizziness	Acquired	Grade II
8	50	M	R	HL, otorrhea	Acquired	
9	41	M	R	HL, FP	Acquired	Grade VI
10	23	F	R	HL,otorrhea,dizziness, tinnitus	Acquired	
11	36	M	R	HL, otorrhea	Acquired	
12	25	M	L	HL, dizziness	Acquired	
13	26	M	R	Dizziness	Acquired	

- 1.Hearing Loss
- 2.Facial Paralysis

Table 2.

Class	Spread
Class I: Supralabyrinthine	Anterior: horizontal part of Intrenal carotid artery (ICA)
	Posterior: posterior bony labyrinth
	Medial: Internal acoustic canal (IAC), petrous apex
	Inferior: basal turn of the cochlea
Class II: Infralabyrinthine	Anterior: ICA vertical part, petrous apex, clivus
	Posterior: dura of the posterior cranial fossa and sigmoid sinus
	Medial: IAC, lower clivus, occipital condyle.
	Inferior: jugular bulb, lower cranial nerves
Class III: Infralabyrinthine – Apical	Anterior: ICA vertical ± horizontal parts
	Posterior: posterior fossa through the retrofacial air cells
	Medial: petrous apex, clivus, sphenoid sinus, rhinopharynx
	Inferior: jugular bulb, lower cranial nerves
Class IV: Masivve	Anterior: ICA vertical ± horizontal parts
	Posterior: posterior fossa dura and IAC
	Medial: petrous apex, superior and midclivus, sphenoid sinus
	Inferior: infralabyrinthine compartment
Class V: Apical	Anterior: Meckel’s cave area and may involve the V. Nerve
	Posterior: IAC and posterior cranial fossa
	Medial: superior or mid clivus, sphenoid sinus
	Inferior: infralabyrinthine compartment

Table 3: Sanna Classification of Petrous Bone Cholesteatoma.

Subclass	Features
Clivus (C)	superior and mid clival extensions are seen from massive, infralabyrinthine - apical and apical PBC whereas the lower clival involvement is a feature of infralabyrinthine-apical PBC
Sphenoid sinus (S)	sphenoid sinus involvement is seen from anteromedial extensions of massive, infralabyrinthine apical and apical PBC; it is a rare extension
Rhino pharynx (R)	it is the rarest extension of the PBC; it is an extension of infralabyrinthine- apical or massive PBC, which may extend through the clivus beneath the sphenoid sinus into the rhino pharynx.

Table 4: Classification of PBC versus pathologic type.

Pathologic type	Congenital	Acquired	Recurrent/ Iatrogenic	Total
Supralabyrinthine	0	4	0	4
Infralabyrinthine	0	3	0	3
Infralabyrinthine-apical	0	0	0	0
Massive	0	3	3	6
Apical	1	0	0	1

Table 5: Surgical approach and class of PBC.

Surgical Approach	No. Of Patients	Classification
TO	3	2 Massive - 1 Apical
ETLA	4	2 SL-2 IL
SP	1	IL
MF+TM	1	SL
TM+RL	1	SL
MTCA	3	Massive
MTCB	1	Massive

TO: transotic; ETLA: enlarged trans labyrinthine approach; SP: subtotal petrosectomy; MF+ TM: Middle Fossa + trans mastoid approach; TM+RL: Transmastoid+Retrolabyrinthine; MTCA: modified trans cochlear approach type A; MTCB: Modified trans cochlear approach type B; SL: Supralabyrinthine; IL: Infralabyrinthine.

Discussion

The largest series of petrous bone cholesteatoma reported in the world literature to date belongs to Sanna et al. with 129 cases [2]. Incidence of PBC in their series was 2.9%. Omran et al had 93 cases of PBC in their study [3]. In our retrospective study with 14 cases we evaluated our management, results and postoperative follow-up experiences accompanied by literature knowledge. According to the Sanna classification massive type was the most common type and hearing loss was the most common symptom in our study similar to the series of Omran et al. PBC treatment is surgery and cholesteatoma needs to be removed clearly. Its extension should be evaluated before the surgery so imaging techniques have critical roles on management of PBC. Computed tomography and MRI are the best modalities for definition

of petrous apex lesions, differential diagnosis, or making a decision for surgical approach and to see the recurrences. In MRI cholesteatoma can be distinguished from the cholesterol granuloma by the presence of intermediate-to-low T1-weighted signal, from a mucocele by the presence of restricted diffusion on diffusion weighted images, and from a benign bone tumor by the absence of enhancement on contrast enhanced images [4]. In some extensive cases sigmoid sinus obliteration could be necessary by intraluminal or extraluminal, so the dominant sinus identification must be done before the surgery. Therefore, MR venography can make a contribution to treatment plan. As Pandya et al [5] described before, some points have to be taken into considerations when treating patients. These are

- i. Complete eradication of the disease
- ii. Preservation of facial nerve function
- iii. Prevention of CSF leakage and meningitis; and
- iv. Cavity obliteration. Hearing preservation is not the primary aim of treatment after radical removal of cholesteatoma. Surgical approaches for PBC can vary from subtotal petrosectomy to modified trans cochlear approaches [2,3,5] including lateral transtemporal (trans labyrinthine, transcochlear, infralabyrinthine, infracochlear) and middle fossa approaches. Furthermore, use of intraoperative navigational systems can lead combination of an endoscopic transsphenoidal approach with transtemporal or middle fossa (MF) approaches [6].

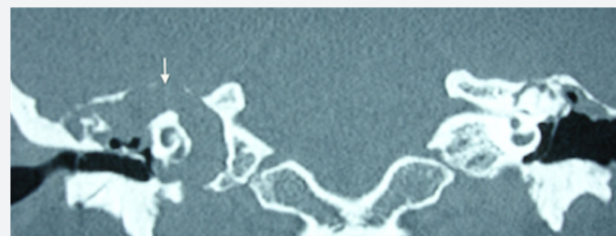


Figure 7: Modified Transcochlear Approach type A. Endoscopic examination of right ear. D: Dura, ICA: Internal carotid artery, PA: Petrous Apex.

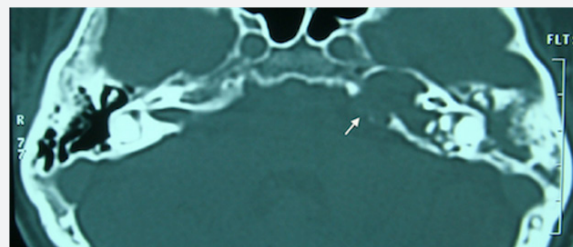


Figure 8: Right ear Modified Transcochlear Approach surgery endoscopic view. ICA: Internal carotid artery, JB: Jugular Bulb, IX. CN: IX. cranial nerve, PA: Petrous Apex.

In massive type PBC, we preferred transotic approach or modified trans cochlear approaches type A and type B (Figures 7-8). Facial nerve function was normal preoperatively in transotic approach group. Sanna et al described in their study an algorithm

for management of the PBC [2]. Our approach was similar to them about massive type cholesteatoma surgery depending on facial nerve function. Although kinds of surgical techniques have come up with this entity, recurrence is still a big problem. The most important reasons for this problem are incomplete removal of cholesteatoma because of critical structures like dura, facial nerve, carotid artery etc. and inadequate consideration of secret areas. Therefore, an endoscopic examination with 30 degree and/or 45 degree endoscopes should be done after removing the cholesteatoma in terms of residual cholesteatoma. Generally, it is hard to preserve hearing level in transotic approach, trans labyrinthine or trans cochlear approaches. The aim of complete eradication of the disease necessitates sacrificing the labyrinth. We could preserve hearing in only three patients. One of them underwent middle (MF) fossa and trans mastoid approach (TM); one of trans mastoid and retro labyrinthine and the other one underwent subtotal petrosectomy. Patient with bilateral PBC had massive type in his right ear and supralabyrinthine type in left ear. Combined MF-TM approach was used for the only hearing ear and MTCA for the other one. Senn et al. [7] reported in their study that hearing preservation was feasible [7]. Kim et al. [8] reported in their study that preserving the hearing level was possible in supralabyrinthine type with appropriate surgery. It is possible to have control on cholesteatoma extending from medial side of superior semicircular canal to internal acoustic canal and geniculate ganglion by middle fossa and trans mastoid approach together, so we had this operation in one patient mentioned above. Infralabyrinthine type cholesteatoma usually extends from medial side of semicircular canals to internal acoustic canal along the posterior cranial fossa dura or extends to petrous apex from medial side of facial nerve and inferior cochlea between the jugular bulb and internal carotid artery. It is possible to remove cholesteatoma and preserve the hearing by subtotal petrosectomy or trans mastoid retro labyrinthine approach for these selected patients.

Patients may suffer for prolonged facial paralysis in extensive cases. Axon et al noted that significant preoperative paresis or paralysis was almost always associated with erosion at the geniculate ganglion, which suggested ischemic etiology. Danesi et al. [9] reported in their series labyrinthine segment was the most frequent site of injury conversely Sanna and Tutar et al. [10] Among the five patients with facial paralysis, only three had prolonged facial asymmetry history. The remaining two patients had grade II and grade III facial paralysis that had been occurred recently. One was treated by extended trans labyrinthine approach with decompressing the nerve. In the patient with grade III facial paralysis, cholesteatoma was extending to medial to labyrinth and internal acoustic canal invading the facial nerve in labyrinthine and intracanalicular segment. Facial nerve could not be preserved, and great auricular nerve graft was used for rehabilitation. One year after graft surgery patient had minimal movement in his left mouth angle. Facial nerve was considered as fibrotic in three patients with massive type PBC so static reconstruction was carried out for these patients. End-to-end

hypoglossofacial anastomosis is still effective procedure in facial reanimation surgery [11]. Generally open radical cavity is favorable and conventional procedure for cholesteatoma surgery however, cavity obliteration technique is recommended in PBC surgery [12]. A deep cavity forms by subtotal petrosectomy, TO, Infratemporal fossa approach and modified trans cochlear approaches so it creates a risk for facial nerve and vascular structures. Obliteration of cavity with abdominal fat also provides to recognize the recurrences and makes the revisions easier.

Conclusion

It is hard to manage the cholesteatoma located in petrous part of temporal bone because of vital structures. So that, advanced imaging techniques are required for describing the pathology and planning the surgical approach. In addition, a proper classification depending on extent of pathology should be done before the surgery and patient should be informed about surgical outcomes. Finally, appropriate surgical technique should be chosen, and patients have to be followed up every year with MRI because of asymptomatic recurrences.

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