



Unusual Benign Middle Ear Tumors

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ABSTRACT

Introduction

Middle ear tumors represents a rare group of neoplasm that vary widely according to their pathology, anatomical involvements and clinical features. They mimic middle ear inflammatory pathologies intricately and hence early definitive diagnosis is a challenge. As these tumors are histologically benign, they are only locally destructive causing vestibular/ facial dysfunction.

Materials and Methods

In this case series, we have included 5 cases of rare middle ear tumors, who presented to our hospital in period of 2020-2023. All patients had a pre-operative examination with otoscope and microscope/otoendoscope. Pre-operative and post-operative hearing assessment done with tuning fork test and pure tone audiometry. Pre-operative radiological investigation done in all cases. Biopsy was usually taken pre-operatively, however in tumors suspected to be highly vascular an intra-operative sample was sent.

Results

Mastoidectomy (canal wall up/canal wall down/radical) was performed as definite surgical treatment to achieve maximum clearance.

Conclusion

Surgery is the treatment of the choice for benign middle ear tumors. High resolution computed tomography and magnetic resonance imaging are important to determine the extent and to help surgical planning in doubtful vascular tumour. Biopsy is helpful for the differential diagnosis.

Keywords

Benign; Ear Polyp; Mastoidectomy; Histopathology

Benign tumors arising from the middle ear contribute a very small fraction of all ear pathologies. Being a heterogeneous group, their presentation varies; though most mimic Chronic Suppurative Otitis Media intricately. Swift establishment of clinicopathological diagnosis is a challenge. As these lesions are benign, they are locally destructive; hence leading to audiological and facial/vestibular dysfunction. Early diagnosis is necessary to prevent these associated complications.

They are classified into two groups:

1. Primary tumour of the middle ear

2. Tumors of adjacent structures that involve the middle ear spaces.

Most common primary tumors are paraganglioma (glomus tumour), followed by adenoma in adults and by haemangiomas in children. Primary tumors may extended from the middle ear space intracranially or into the other portion of the temporal bone and skull base.¹ High resolution Computed tomography (HRCT) and magnetic resonance imaging (MRI) are useful in defining the extent and nature of the lesion.² While microscopic or otoendoscopic examination is always done preoperatively in all polyp/mass in the ear cases, biopsy is must for definitive diagnosis. It may sometimes be difficult to distinguish between polyps arising from the middle ear and those arising from the external ear since both may fill the meatus and obscure the exact site of origin. So far, the surgical intervention has been the mainstay of

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treatment. Surgical excision with radical extent is definitive to achieve maximum clearance.

The aim of this study is to present five cases of these relatively rare disease in the middle ear, discuss the clinical characteristics of the disease, and to highlight the management. In this case series we have included five rare and anatomically complex tumors. We have also discussed their optimal surgical management to achieve optimal clearance and to prevent recurrence.

Materials and Methods

Cases of middle ear tumors reported on histopathology pre-operatively and/or intra-operatively were enrolled. Pre-operatively otoscopic and otoendoscopic/microscopic examinations were done in all cases. Origin and anatomical location of the tumour was detailed. Biopsy was taken under otomicroscopy with adequate preparation for haemostasis. We have analysed the clinical characteristics of these five cases after ruling out chronic otitis media; such as gender, age, clinical manifestations, clinical images, radiological correlation, intraoperative extension, intraoperative clearance of the disease, pathological data, surgical approach. Audiological investigation done preoperatively and postoperatively (after 3 months) with pure tone audiogram and tuning fork test. All data were documented.

Case series:

Case 1

Glomus tympanic Paraganglioma : A 32-years old female presented with complaint of blood stain right ear discharge with earache for 6 months. On otoscopic examination mass was seen arising from tympanic membrane. Clinically we were suspecting vascular mass, so biopsy was not taken pre-operatively. Pure tone audiogram suggested severe conductive hearing loss. MRI was suggestive of presence of poorly defined abnormal signal intensity mass lesion involving middle ear extending up to external auditory canal, eustachian tube and mastoid air cell, reaching up to tegmen tympani. Right radical mastoidectomy done. Intra operatively tumour

extending up to antrum, aditus, external auditory canal (EAC), sinus plate, dural plate, part of eustachian tube and carotid area, lenticular process of incus necrosed, stapes partially eroded and preserved. No major complications were seen. Post-operative histopathology report suggested of glomus tympanic tumour. She is disease free till this time.

Case 2

Capillary haemangioma : A 43-year-old female presented with left-sided hearing loss with ear discharge for 1 year. On otoscopic examination a bright red polypoidal mass occupied the whole of external auditory canal, occluding the view of tympanic membrane. It seemed like vascular tumour during examination, so biopsy was not taken pre operatively. Pure tone audiogram revealed moderate conductive hearing loss. HRCT was suggestive of ill defined, non-enhancing hyperintensity involving left external auditory canal, left middle ear cavity and mastoid air cell. Intraoperatively, the tumour was found to occupy the entire epitympanum and mesotympanum. The mass had encased the entire ossicular chain without any erosion. The incus and malleus were removed to facilitate tumour excision. Canal wall down mastoidectomy with type 3 tympanoplasty done. Total clearance could be achieved. Biopsy confirmed the clinical suspicion of capillary haemangioma. No major post-operative complication seen and the cavity healed uneventfully in 4 weeks.



Fig. 1. Oto-endoscopic examination showed tumour arising from posterosuperior region of left ear

Case 3

Myxoma: A 45-year-old female presented with Left otalgia and ear discharge for six months. Oto-endoscopic examination revealed a polypoidal tissue in postero superior quadrant (PSQ) region coming from middle ear. A small bit was sampled and biopsy suggested myxoma. Audiogram suggested of moderate conductive hearing loss. HRCT of the temporal bone revealed, left mastoid air cell markedly filled with tumour. Mastoid antrum, air cell and tympanic cavity were filled with hypodense tissues. Post-operative histopathology showed marked proliferation of spindle to stellate shaped cells deposited in extensive myxoid stroma, suggestive of myxoma. Canal wall up mastoidectomy with type 1 tympanoplasty was done as no ossicular erosion was noticed. No major post-operative complication seen.

Case 4

Facial Schwannoma: A 62-year-old female came with complains of severe left ear pain and ear discharge. On Microscopic examination, polypoidal tissue was present in EAC coming from PSQ region. Pure tone audiogram

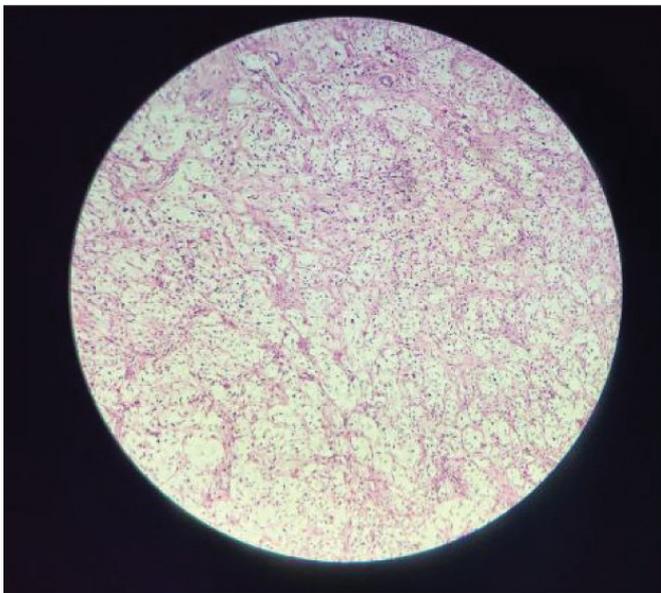


Fig. 2. Post operative Histopathological examination (hematoxylin and eosin stain under 40x magnification) suggested of schwannoma

suggested of profound mixed hearing loss. HRCT temporal bone revealed ill-defined tissue density lesion is seen in the external auditory canal, mesotympanum and hypotympanum. Intraoperatively the tumour found to be arising from second genu of facial nerve and was in close proximity to chorda-tympani nerve. Tumour could be peeled off facial involving the epineurium, however the segment of chorda in contact with lesion was sacrificed. It involved mesotympanum, hypotympanum, sinus tympani. Tumour was removed in-toto and canal wall down mastoidectomy with type 3 tympanoplasty reconstruction was done. Post-operative histopathology report suggested of schwannoma. As even post-operative no facial palsy was seen, we inferred it to be Chordal Schwannoma.

Case 5

Myxoma in childhood: A 7 years male child with complaint of swelling over left side pre and post aural swelling for 3 years. Swelling was small in size, gradually progressive. It was associated with decreased hearing; although no vertigo, nystagmus or facial palsy was present. On otoscopic examination, a polypoidal mass in the external ear canal that was covering whole of the canal and further not negotiated to see the tympanic membrane. History of twice excision biopsy from swelling done 1.5 year and 1 year back at some peripheral hospital. Histopathology report was suggestive of low grade myxoid tumour. Patient visited to us with swelling over pre and post auricular region with discharge coming from it. Audiometry suggested mild conductive hearing loss. HRCT temporal bone showed abnormal fluid density noted in left external auditory canal, middle ear cavity and mastoid air cells with abutting left parotid gland with effaced fat plane, thinning of tegmen tympani was noted. MRI showed multi-lobulated abnormal signal intensity lesion, in left mastoid air cells and middle ear extending into left external auditory canal and in left infra mastoid region; lesion was causing superior bulging and scalloping of tegmen tympani without invasion of adjacent brain. It

measures approximately 7.0 (CC) x 5.0 (W) x 4.3 (AP) cm in size. It is displacing parotid gland infero-medially without invasion. As this tumour involved preauricular area, we kept extended modified Blair's incision. Intra operatively tumour was going into zygomatic area anteriorly, parotid gland was displaced inferomedial and middle ear superiorly. Tumour separated from surrounding tissues, removed. Mastoid bone was exposed through the same incision. On drilling the mastoid bone tumour was reaching and completely filling air cell system, without significant bone destruction. Epitympanum, Mesotympanum and Hypotympanum was also completely engulfed with tumour tissue. Eustachian tube was identified and blocked with muscles tissue. Closure over preauricular region was done with rotation of temporalis muscle. Facial functions were all preserved after surgery. Post-operative histopathological report was suggestive of hyperplastic squamous epithelium, myxoid stroma with scattered spindle and stellate shaped cells suggestive of low grade myxoid tumour.



Fig. 3. Pre operative, intraoperative tumour location with closure

Table I : Summary of five cases

SR. NO.	AGE/SEX	CASES	PRE OP HPR	POST OP HPR	SURGERY REQUIRED
1.	32/F	Glomus tympanic paraganglioma	Not done	Glomus tympanic	Radical mastoidectomy
2.	45/F	Capillary haemangioma	Not done	Capillary haemangioma	CWD mastoidectomy type3 tympanoplasty
3.	42/F	Myxoma (female)	Myxoma	Myxoma	CWU mastoidectomy type 1 tympanoplasty
4.	65/F	Schwannoma	Polyp	Schwannoma (chordal)	CWD mastoidectomy type 3 tympanoplasty
5.	07/M	Myxoma (childhood)	myxoma	myxoma	Radical mastoidectomy

(op = operative, HPR = histopathology report, CWD = canal wall down, CWU = canal wall up)

Discussion

Middle ear tumors are rare and are best evaluated with imaging studies. As they imitate otitis media, High resolution Computed Tomography imaging is the key their assessment. When combined with patient's clinical findings, Computed Tomography findings can assist in disease classification which is helpful for definitive surgery. Surgical excision with histopathologic analysis is usually required for definitive diagnosis and treatment. They are classified into two groups: 1. Primary tumour of the middle ear and 2. Tumors of adjacent structures that involve the middle ear spaces. Most common primary tumors are paraganglioma (glomus tumour), followed by adenoma in adults and by haemangiomas in children.

The paraganglioma are also known as glomus tumour or chemodectomas. Incidence of paraganglioma tumour said to be about one per 1.4 million people per year. These are frequently seen in women (66-90%) in 4th and 7th decades of life. It can cause hearing loss, tinnitus, dizziness, facial palsy. Paragangliomas can be divide into three categories: 1) Jugular paragangliomas 2) Tympanicum paragangliomas 3) Jugulo-tympanicum paraganglioma. MRI helps in recognizing the characteristic vascularity of the tumor.³ Histopathologically, epitheloid or spindle-shaped cells are seen to form nests or layers, and stroma has a rich vascular network. The tumour resection is usually done via trans mastoid approach and almost universally converted into canal wall down mastoidectomy in the view of the size of the tumour.

Capillary haemangioma is classified as a vascular tumour, while cavernous lesions are vascular malformations.⁷ Vascular anomalies are common in head and neck but rarely reported in the ear. Only 18 cases have been reported in English literature and out of which, only one with hemangioma of middle ear was extended to the mastoid cavity. Haemangiomas are tumors growing with age. They can cause significant morbidity in the enclosed spaces of the ear. Clinically usually reddish polypoidal mass is seen in EAC occluding further visualization. Imaging is indicated to classify and assess the extent of the lesion. Bony structures are usually intact, however erosion of EAC, ossicles and facial canal may

be seen in some cases due to pressure effect. Their management depends on the diagnosis and its presentation. Radical Surgery with complete excision is the treatment of choice.⁸

Schwannomas are generally seen over the age of 40 years. But Age of presentation varies from 5 to 84 years. No gender or side predilection is observed.⁴ It is mostly seen unilateral. It is seen bilaterally in the syndrome of neurofibromatosis type 2. One study of 600 cadaver temporal bone studies reported facial Nerve Schwannoma incidence of 0.8%. The incidence of infratemporal facial neuromas was 0.8% in a cadaveric study, although this figure is higher than the rate of clinical presentation.⁵ They are generally encapsulated well-circumscribed lesions. As in schwannomas seen elsewhere, it consists of Antoni A fields and Verocay bodies. Hyalinized vessel walls are encountered. Degenerative changes such as haemorrhage, necrosis, and myxoid changes can be observed. Diagnostic workup includes audiometry (audiological test), auditory brain stem evoked response audiometry, Computed Tomography, and contrast-enhanced MRI. Treatment includes surgical removal. Approach depends on the site of tumour, size of tumour, and hearing loss. Timing for surgery is controversial because facial nerve neuromas almost always grow slowly.⁶

Myxoma is the benign tumour originating from the mesenchymal tissue. It is rare tumour and its molecular mechanism and prognosis remain unclear till date. Myxomas in the head and neck areas are most commonly seen in maxilla and mandible. Myxoma is very rare benign tumour of the middle ear. However, it may also develop in the auricular, ear canal, middle ear and temporal bone. Only 5 cases of isolated myxoma of the external auditory canal have been reported in the literature.⁹ The typical myxoma develops from the atrium of the heart. It is mostly seen as carney complex. Carney complex is a hereditary condition associated with spotty skin pigmentation with connective tissue tumors; and a number of other tumours of the endocrine (hormone-producing) glands. Carney complex is inherited autosomal dominantly and PRKARIA gene mutations or deletions are seen in

patients. Management is mainly complete surgical resection.

Traditional trans canal approaches should be reserved for small, discrete masses in the mesotympanum. A postauricular incision is more appropriate for excising small, discrete lesions of the anterior mesotympanum or when mastoidectomy is anticipated for tumors that extend to the epitympanum, hypotympanum, or outside the middle ear space. When the tumour invades the pre-auricular/parotid space an extended parotid-neck incision can also serve as approach for mastoidectomy with maximal

retraction. In all cases hearing outcome was noted. Preoperative and postoperative audiological assessment done with Tuning fork test (table II) and Pure tone audiometry. For tumors that involve the mastoid cavity, canal wall-up mastoidectomy is preferred in uncomplicated resections. However, there should be little hesitation to convert it to a canal-wall-down procedure to achieve better exposure and haemostasis. Finally, a complete and safe resection of a middle ear tumour often requires disarticulating the ossicular chain, and surgeons should have a low threshold for doing so.¹⁰

Table II : Pre operative and post operative hearing assessment with Tuning fork test

TUNNING FORK	CASES	RINNE NEGATIVE		RINNE POSITIVE		WEBERS LATERALISED TO AFFECTED EAR		WEBERS LATERALISED TO OPPOSITE EAR		ABSOLUTE BONE CONDUCTION TEST (NOT DECREASED)		ABSOLUTE BONE CONDUCTION TEST (DECREASED)	
		Pre op	Post op	Pre op	Post op	Pre op	Post op	Pre op	Post op	Pre op	Post op	Pre op	Post op
256 Hz		2	0	1	2	2	2	1	2	2	0	1	0
516 Hz		4	3	1	2	4	2	1	2	4	3	1	0
1024 Hz		4	3	1	2	4	2	1	2	4	3	1	0

Conclusion

Benign middle ear tumors should be differentiated from chronic otitis media as management protocols differs. Otomicroscopic examination and biopsy are ideal though should be reserved for clinico-radiological non-vascular tumour. High resolution computed tomography and magnetic resonance imaging are important to determine the extent and to help surgical planning in doubtful vascular tumour. It is also important for the differential diagnosis. Surgery (modified radical mastoidectomy/radical mastoidectomy) with hearing reconstruction is the treatment of the choice for benign middle ear tumors. Even though the tumour can be successfully treated with surgical resection and long term follow up is required to assess recurrence.

References

1. Razek AA, Huang BY. Lesions of the petrous apex: classification and findings at CT and MR imaging. *Radiographics* 2012;32(1):151-73
2. Isenring D, Pezier TF, Vrugt B, Huber AM. Middle ear adenoma: Case report and discussion. *Case Rep Otolaryngol* 2014; 2014: 342125
3. Pellitteri PK, Rinaldo A, Myssiorek D, Gary Jackson C, Bradley PJ, Devaney KO, Shaha AR, Nettekville JL, Manni JJ, Ferlito A. Paragangliomas of the head and neck. *Oral Oncol* 2004;40(6):563-575
4. Mundada P, Purohit BS, Kumar TS, Tan TY. Imaging of facial nerve schwannomas: diagnostic pearls and potential pitfalls. *Diagn Interv Radiol* 2016;22(1):40-46
5. Saito H, Baxter A. Undiagnosed intratemporal facial nerve neurilem *Arch Otolaryngol* 1972;95(5):415-419
6. Angeli SI, Brackmann DE. Is surgical excision of facial nerve

- schwannomas always indicated. *Otolaryngol Head Neck Surg* 1997;117(6):S144–S147
7. Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. *Oral Dis* 2010;16(5):405-418
 8. Pavamani SP, Surendrababu NR, Ram TS, Thomas M, Viswanathan PN, Viswanathan FR. Capillary haemangioma involving the middle and external ear: radiotherapy as a treatment method. *AustralasRadiol.* 2007 Aug;51(4):394-7. doi: 10.1111/j.1440-1673.2007.01733.x
 9. Shadfar S, Scanga L, Dodd L, buchman CA. Isolated myxoma of the external auditory canal 2014;124(5):1220-1222
 10. Z.Jasonqian, MD; Amy M.Coffey, MD; Kathleen M. Management of benign middle ear tumors:series of 7 cases.*ENT journal* 2017;96(10-11):426-432b.