

# Glomus Tympanicum: A Report of Two Cases

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## Summary

Two cases of glomus tympanicum tumor of the middle ear stage 'type A' according to Fisch classification, are presented due to their rarity in this part of the world. These tumors were excised by end aural tympanotomy approach and there has been no recurrence in both patients to date.

**Key Words:** Glomus tympanicum, Paraganglioma

## Introduction

Glomus tumors are the most commonly diagnosed neurotologic neoplasm after acoustic neuroma<sup>1</sup>. When they are confined to the middle ear space they are termed glomus tympanicum. The term glomus is a misnomer because these tumors originate from special neural crest elements, the paraganglion cells, which with autonomic ganglion cells, form the paraganglia<sup>2,3</sup>. Temporal bone glomus bodies are vascularized by the inferior tympanic branch of the ascending pharyngeal artery and are in association with Jacobson's and Arnold nerves. Glomus tympanicum have been reported in all age groups but most frequently occur in the fifth and sixth decades of life. The ideal management of glomus tympanicum is complete surgical excision<sup>4</sup>.

## Case Report 1

A 49 year old Indian lady presented with three months history of headache with intermittent episodes of nausea and vomiting. Otoscopy revealed a reddish mass behind the left tympanic membrane. Pure tone audiometry revealed a mild conductive deafness of the left ear. CT scan of the temporal bone as well as MRI demonstrates mass in the left middle ear, based on the

cochlear promontory and in contact with all the ossicles but not eroding the ossicles. This tumor mass extended from the inferior most portion of the middle ear up to the level of horizontal portion of the facial nerve, which was intact i.e. 'type A' according to Fisch classification. Complete excision of tumor was performed by an end aural approach. Intra-operatively the tumor was found to be adherent to the tympanic membrane. There was a central perforation of the tympanic membrane post surgery. Further follow up on the patient for a year has revealed no recurrence to date.

## Case Report 2

A 52 year old lady was referred to the ENT department complaining of right pulsatile tinnitus and progressive hearing loss for the past two years. Otoscopy showed a reddish mass behind the tympanic membrane and pure tone audiometry revealed moderate conductive hearing loss on the affected side. CT scan (Fig 1) revealed a mass behind the right tympanic membrane which was consistent with glomus tympanicum 'type A' according to Fisch classification. Excision of tumor was performed by an end aural approach. Intra-operatively, a soft tissue mass was noted to be behind the tympanic membrane extending to the attic

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superiorly and excised completely. Patient was discharged well. Further follow up on the patient for a year has revealed no recurrence to date.

## Discussion

Glomus tympanicum is usually considered to be a nonchromaffin paraganglioma with no endocrine function<sup>2,3,4</sup>. Histologically glomus tumors resemble glomus bodies with epitheloid cells interspersed in a highly vascular stroma of capillary and pre capillary vessels. Cytologically they are not very active with only rare mitotic figures. Clinically, these can be locally invasive and destructive of bone and facial nerve. Glomus tympanicum shows predominance in females. Clinically it is important to look for evidence of endocrine activity by urinary assay of metabolites dopamine and VMA<sup>5</sup>. All patients with glomus tumors should be evaluated for multiple lesions. These tumors are generally considered to be a low grade malignancy mainly causing problems because of its site in the complex anatomy of skull base. Progression can be so slow that ongoing neurologic deficits may undergo simultaneous compensation and go unnoticed by the patient. There have been many attempts at tumor classification. The most widely accepted classification was proposed by Fisch based on tumor site and size as follows -

- Type A - Tumors localized to middle ear cleft
- Type B - tympanomastoid tumors with no destruction of bone in the infralabyrinthine compartment of the temporal bone
- Type C - tumors invading the bone of the infralabyrinthine compartment of the temporal bone
- Type D - tumors with intra cranial extension.

The most common physical sign in glomus tympanicum is a vascular middle ear mass. Two other conditions that mimic this appearance are a high jugular bulb and an aberrant internal carotid artery. The red drum of otitis media should be obvious from the history, a more extensive spread involving the external auditory meatus may appear to be a squamous cell carcinoma which can bleed profusely. The most common presenting symptom is pulsatile tinnitus (80%) followed by hearing loss<sup>2,3,4,5</sup>. Tympanic membrane erosion and bleeding are late symptoms. Facial nerve weakness signals advanced disease, and an indication of poor facial nerve prognosis. Otoscopy can be misleading<sup>1</sup>.

The first step in investigation entails a careful clinical examination, observation of the drum under the microscope will frequently show a pulsation of mass which can be soft and often blanches on palpation. Hearing loss is conductive and neurological assessment of the cranial nerves give a considerable information regarding the extent of tumor.

Myringotomy and biopsy are to be avoided. If tissue is required for diagnosis, a post auricular, transmastoid approach is recommended as vital anatomy is identified and bleeding can be safely controlled.

The main investigation is radiological where a very detailed assessment of the tumor can be made. CT scan remains the most valuable method of imaging giving information about bone erosion and extent of tumor to bony anatomy of the ear which is vital in operative planning. MRI with gadolinium enhancement gives a better indication of soft tissue involvement but is more difficult to correlate with anatomy. Angiography indicates blood supply of tumor and with preoperative embolization allows a significant reduction in operative bleeding. More recently MRA has also demonstrated its value in diagnosis.

The treatment plan is based on tumor size, tumor location, patient's age and health. Current treatment options are surgical excision or primary radiotherapy or surgical excision with planned adjunctive radiotherapy or no active treatment and continuous observation<sup>1,2,3</sup>.

At present the preferred method of management is surgical, but management strategies must be tailored to the individual patient. Objectives of the surgery are total resection of tumor where possible and without increasing the patient's neurological deficit<sup>1,4</sup>. Type A tumors can usually be approached via the external auditory meatus. If the margins of the tumor are indistinct, these tumors can often be encompassed by combined approach (intact canal wall) procedure.

Glomus tympanicum are extremely slow growing and may have a long natural history. Some patients do not present until the latter part of the sixth or seventh decades and if the repeat scans do not show very extensive spread or rapid growth with minimum symptoms, no treatment is indicated apart from reassurance. An elderly or infirm patient with a symptomatic, growing tumor should be treated solely with radiotherapy. In those extensive tumors for which

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surgery may well increase the neurological deficit, a subtotal resection with post-operative radiotherapy is probably the method of choice.

Data regarding best treatment outcome by radiation therapy or surgery do not exist. Radiation therapy should not represent primary treatment for glomus tympanicum tumor, because the risks of surgery are low<sup>4</sup>.

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