

## Audiological findings in glomus tumours

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### Abstract

Glomus tumours involving the middle ear and the cerebellopontine angle are reported with emphasis on audiological findings. Magnetic resonance imaging (MRI), angiographic and pathological results are presented. Audiological tests, including impedance audiometry, evoked otoacoustic emissions and auditory brainstem responses, are valuable in evaluation of the effect of glomus tumours on the auditory system as well as their pathological extent.

**Key words:** Glomus tumour; Ear, middle; Hearing loss, sensorineural; Hearing loss, conductive

### Introduction

Glomus tumours which may include glomus tympanicum and glomus jugulare are common vascular neoplasms of the middle ear (Glasscock and Jackson, 1993). Glomus tympanicum develops from the tympanic nerve plexus in the mucosa over the cochlear promontory and can be classified as extraadrenal paragangliomas (Stewart, 1993). The tumours originating in the adventitia of the jugular bulb area or other areas of the temporal bone may invade the bony floor of the middle ear, spread into the middle ear space and even extend into the middle cranial fossa (Carlson *et al.*, 1987; Anand *et al.*, 1993; Moffat *et al.*, 1993). Because the glomus tumours often involve the auditory pathways, audiological and radiological examinations are the primary means of the diagnosis of these tumours.

The patients with glomus tympanicum or glomus jugulare in association with significant hearing impairments are investigated here with emphasis on the audiological manifestations.

### Methods and results

Pure-tone thresholds were obtained at frequencies from 250 to 8000 Hz. Speech recognition threshold (SRT) was measured using spondee words. Word discrimination performance was tested at 40 dB SL. Tympanometry and the acoustic reflex tests were conducted using a GSI-33 middle ear analyser (Gradson-Stadler Inc., Littleton, USA). Transiently-evoked otoacoustic emissions (TEOAEs) were tested by the ILO88 otoacoustic emissions analyser (Otodynamics Inc., Hatfield, England).

Audiological findings in five patients with glomus tumours are summarized in Table I. The diagnoses of glomus tumours were all confirmed by angiography, computed tomography (CT) or MRI, surgery, and pathology. The diagnosis of glomus tympanicum for the patient TC (see Table I) was finally determined by electron microscopy (EM) following two previous negative results by the light microscopic examinations. EM of the tissue from the right middle ear in this patient shows that the tumour cells are of moderate size with abundant cytoplasm. The nuclei are mostly ovoid and occasionally have a small nucleolus. The chromatin is primarily dispersed euchromatin, but shows scattered irregular foci of clumped chromatin. The cytoplasm containing scattered, mildly swollen mitochondria is seen with normal appearing endoplasmic reticulum and Golgi complex. Moderate numbers of osmophilic, well-rounded, membrane bound secretory granules are seen which range from about 100 to 325 nm in size. Some cells contain large numbers of microfilaments. The cytoplasmic membranes are fairly regular and lack cytoplasmic processes. Small intercellular junctions are seen (Figure 1). These pathological findings are consistent with glomus tympanicum or paraganglioma. Two typical cases are presented as follows.

### Case reports

#### Case 1

A 43-year-old woman presented with a six-year history of a progressive hearing loss associated with pulsatile tinnitus in the right ear. The patient reported pressure and fullness in the affected ear with a negative history of facial paralysis, bloody

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TABLE I  
SUMMARY OF AUDIOLOGICAL AND VIDEO-OTOSCOPY FINDINGS IN PATIENTS WITH GLOMUS TUMOURS

No.	Patient	Sex	Age	Ear	Diagnosis*	Pure-tone audiometry	Speech audiometry	Impedance audiometry	EOAEs	ABR	Video-otoscopy findings
1	MW	F	43	Right	Glomus tympanicum	Moderate conductive hearing loss	50 dB HL of SRT; 92% of WRS	Flat tympanogram; Absent acoustic reflex; Pulsating baseline	Absent	Did not test	Reddish mass bulging TM toward the canal; pulsating movement of mass
2	KA	M	27	Left	Glomus jugulare with CPA extension	Mild SNHL	15 dB HL of SRT; 96% of WRS	Normal tympanogram; Elevated ART; Pulsating baseline	Absent	Delayed wave V	Reddish tissue on the floor of the canal and inferior TM
3	TC	F	21	Right	Glomus tympanicum	Moderate conductive hearing loss	40 dB HL of SRT; 100% of WRS	Flat tympanogram; Absent acoustic reflex	Absent	Did not test	Polyp-like reddish mass in the middle ear with pulsating movement
4	OF	F	57	Right	Glomus jugulare	Conductive hearing loss	45 dB HL of SRT; 96% of WRS	Flat tympanograms; Absent acoustic reflex; Pulsating baseline	Absent	Did not test	Reddish mass behind the tympanic membrane with Brown's sign
5	LG	M	40	Left	Glomus jugulare	Mixed mild-to-moderate hearing loss	20 dB HL of SRT; 92% of WRS	Flat tympanogram	Did not test	Did not test	Reddish mass in the middle ear with extension to the external canal

Note – SNHL: sensorineural hearing loss; CPA: cerebellopontine angle; SRT: speech reception threshold; TM: tympanic membrane; WRS: word recognition score.

\*The diagnosis was confirmed by CT or MRI, angiography and pathology.

drainage, otalgia and vertigo. She was misdiagnosed as having otitis media and had undergone myringotomy with a pressure-equalization tube in the right ear at another hospital a month earlier. She reported no hearing improvement following the myringotomy. Video-otoscopy in the right ear demonstrated a reddish, pulsating mass which bulged the tympanic membrane (TM) toward the external auditory canal. The wall of the external canal was normal. CT scan of the temporal bone showed opacification in the right mastoid air cells with soft tissue opacity in the

right middle ear which appeared to erode the scutum. An MRI scan revealed abnormally increased signal contrast in the right middle ear cavity and the mastoid air cells. On contrast MRI, this region was enhanced uniformly, suggesting a vascular tumour in the middle ear (Figure 2). Angiography demonstrated a well-circumscribed  $1.7 \times 1 \text{ cm}^2$  vascular tumour in the area of the middle-ear cavity with its blood supply from the branches of the ascending pharyngeal artery (Figure 3).

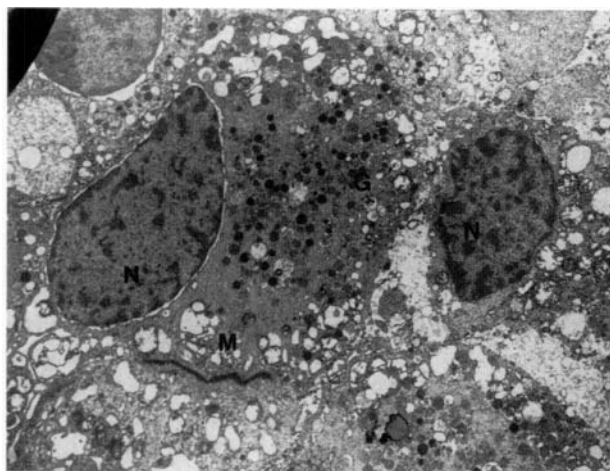


FIG. 1

Electron microscopy of the tissue from the middle ear confirms diagnosis of glomus tympanicum in a 21-year-old woman. G: Osmophilic secretory granules; M: Mitochondria; N: Nucleus.

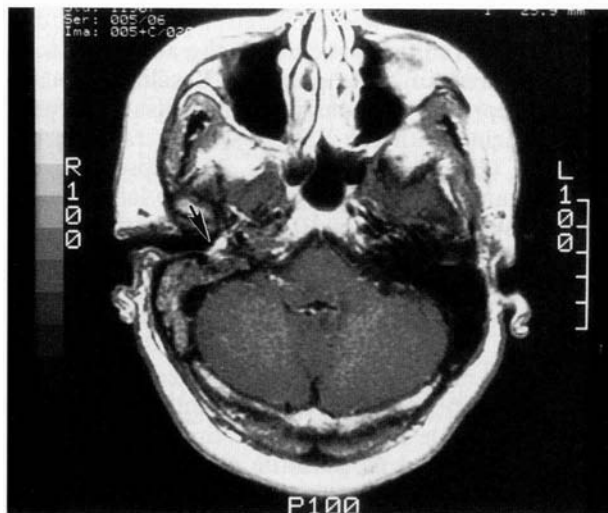


FIG. 2

Axial T1 weighted, post-contrast MRI scan of the brain in a 43-year-old woman (MW), demonstrates abnormal enhancement in the right middle-ear cavity, suggesting a vascular tumour.

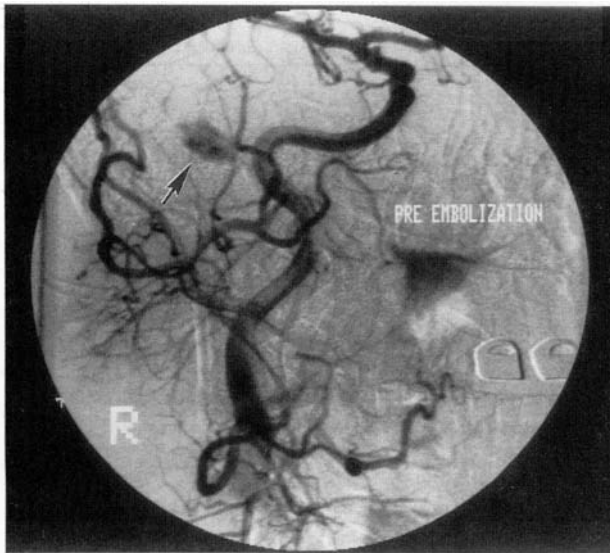


FIG. 3

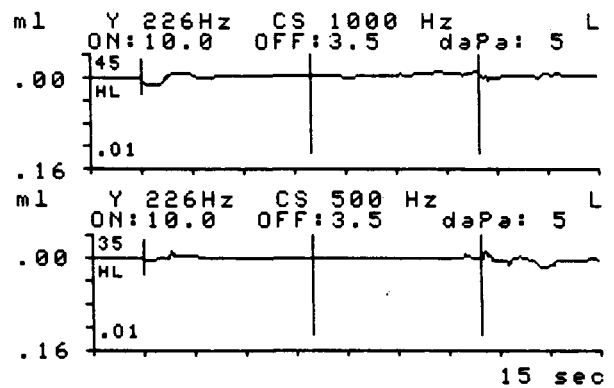
Cerebral angiogram reveals a well circumscribed  $1.7 \times 1$  cm tumour blush in the area of the right middle ear supplied by branches of the ascending pharyngeal artery in a 43-year-old woman (MW). No abnormal venous drainage was noted during the venous phase of the arteriogram.

A pure-tone audiogram showed a conductive hearing loss in the right ear with 55 dB HL of the average threshold at 500, 1000 and 2000 Hz. Normal hearing sensitivity was found in the left ear. The speech recognition threshold (SRT) was 50 dB HL for the right ear and 5 dB HL for the left ear. WRS was 100 per cent at 50 dB HL on the left ear and 96 per cent at 90 dB HL on the right ear. Tympanometry showed a flat tympanogram (type B) with unmeasurable pressure and compliance on the right ear. The left ear demonstrated a normal tympanogram (type A) with pressure of 0 dapa and 0.6 ml of compliance. Ipsilateral acoustic reflex threshold (ART) was within normal limits when a probe tone and activating tone were delivered in the left ear, while contralateral ART was elevated when probe tone was produced in the left ear and the activating tone was delivered in the right ear. Baseline acoustic reflex recording demonstrated a pulsating curve synchronizing with the patient's heart rate on the right ear (Figure 4). TEOAEs were absent in the right ear and normally presented in the left ear with a response amplitude of 10.5 dB SPL and reproducibility of 98 per cent.

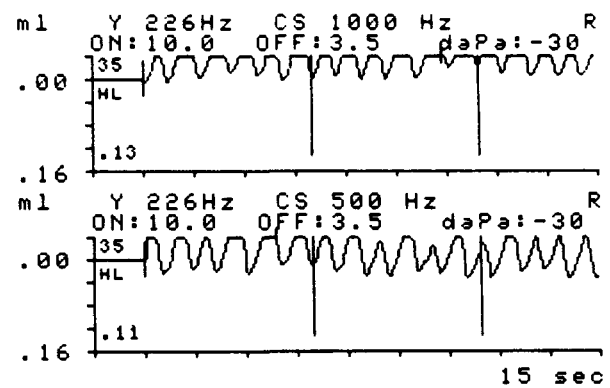
### Case 2

The patient, a 27-year-old man, presented with a four-month history of intermittent vertigo, left tinnitus and side headache. The tinnitus in the left ear was described as pulsatile at a rate consistent with the heart beat, and occasionally associated with nausea and vomiting. The patient also reported a decreased hearing in the left ear.

On examination, the left TM was intact with a reddish mass bulging from the floor of the external canal near the anterior-inferior part of the TM. The



(a) Normal ear



(b) The ear with glomus tumour

FIG. 4

The baseline of impedance measurement shows the waveform during 15 second interval recording at atmospheric pressure on both ears. The right ear with glomus tumour demonstrates a pulsating baseline compared to the flat baseline on the left ear. Note that the frequency of the pulsating peaks is synchronized with the patient's heart beat (68/min).

mass in the middle ear was barely visible through the TM. Examination of the right ear was unremarkable. MRI of the brain showed a glomus tumour near the jugular foramen. The tumour was bounded by the tympanic cavity laterally and the petrous ridge posteriorly. The patient underwent left sub-craniotomy, and mastoidectomy with excision of the glomus tumour. The surgery revealed that the glomus tumour extended to the cerebellopontine angle (CPA) and the posterior aspect of the petrous ridge (Figure 5).

Pure-tone thresholds ranged from 10 to 35 dB HL with 15 HL of SRT and 96 per cent of WRS for the left ear. Hearing sensitivities and WRS were within normal limits for the right ear. Tympanometric findings were normal for both ears. ARTs were normal for the right ear and elevated at 500, 1000 and 2000 Hz with absent response to 100 dB HL tone at 4000 ipsilaterally on the left ear. The measurement of the impedance baseline showed a pulsating waveform for the left ear. TEOAEs were absent for the left ear and mildly reduced with 8.7 dB SPL of the response level and 89 per cent reproducibility for the right ear. Auditory brainstem responses (ABR)



FIG. 5

Axial T1 weighted MRI showed the glomus tumour extended to the cerebellopontine angle and the posterior aspect of the petrous ridge in a 27-year-old man.

revealed a delayed wave V (latency: 6.21 ms) on the left ear and normal latency of wave V (5.68 ms) on the right ear upon response to the clicks at 80 dB nHL.

**Discussion**

The diagnosis of glomus tumours may challenge even the most experienced otolaryngologists. Radiological studies including CT, MRI and angiography are essential to determine the extent, limitation and vascularity of the tumour and differentiate glomus tympanicum from glomus jugulare (Probst *et al.*, 1991; DeAndrea and Josephson, 1992; Glasscock and Jackson, 1993; Stewart, 1993). Audiological data are also helpful to evaluate the effects of tumour on the auditory system. Pure-tone audiometry in glomus tympanicum tumours may show a conductive, sensorineural or mixed hearing loss of varying degree (Alford and Guilford, 1962; Bratt *et al.*, 1979). In 170 patients with tumours of the glomus jugulare associated with hearing loss, 65 per cent revealed sensorineural loss, 32 per cent demonstrated conductive loss and four per cent had mixed loss (Alford and Guilford, 1962). A conductive hearing loss is a typical manifestation in glomus tympanicum tumour. The presence of a sensorineural hearing loss or mixed hearing loss may indicate that the tumours have invaded the cochlea or the eighth nerve as shown in Case 2 (Table I). Impedance audiometry provides unique findings dependent on the nature and extent of pathological involvement. If the tumour involves the tympanic membrane or ossicular chain, tympanometry or the baseline measure of the acoustic reflex may show pulsating curves which are synchronous with the patient's heartbeat. A typical waveform is shown in Figure 4. Three of the five cases in this study revealed these pulsating waveforms. Note that the frequency of pulsating waveforms (17 peaks per 15 seconds) is synchronized with the patient's heart rate (68/min). Evoked otoacoustic emissions (EOAEs) are low-intensity sounds that are produced by the outer hair cells of the cochlea and recorded from the external canal (Kemp, 1978). EOAEs are present in the ear with hearing thresholds better than 30–35 dB HL and normal middle ear function (Probst and Harris, 1993). In Case 1, glomus tympanicum tumour resulted in the absence of EOAEs in the right ear. In Case 2, EOAEs were absent in the left ear with a mild SNHL, suggesting a possible cochlear involvement as confirmed by the subsequent exploratory surgery (Figure 5). It is believed that combination of pure-tone and impedance audiometry with EOAEs and ABR is valuable in the case of glomus tumours which involve the cochlea or retrocochlea. If impedance audiometry reveals normal middle ear function, the absence of EOAEs associated with SNHL may indicate that cochlear damage has been caused by involvement by tumour unless special audiological and radiological tests demonstrate a retrocochlear involvement.

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