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Glomus Tympanicum: Diagnosis and Management

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Abstract

Glomus tympanicum is the most common middle ear tumour. They are often revealed by pulsatile tinnitus and hearing loss. Imaging is crucial in establishing the diagnosis and permits clear analysis of its extension. They should be differentiated from jugular tumours as they require a different surgical approach. We report the example of a female patient who is 57 years old, presenting progressive hearing loss of the left ear. MRI demonstrated a vascular mass occupying the left middle ear with no evidence of jugular bulb involvement. Surgery was recommended and the pathology findings confirmed a glomus tympanicum tumour.

Keywords: Glomus tympanicum; Hypotampanicum; Radiotherapy

Introduction

Glomus tympanicum represents the most frequent middle ear tumour with a slow-growing evolution. Imaging plays an important role in diagnosis and to distinguish them from jugular tumours. Hypotampanicum involvement requires MRI to exclude jugular bulb invasion. The main treatment option is surgery. Resorting to embolization or radiotherapy is also possible [1-2].

Case Report

Our patient is a lady in her 57th year, who reported a left-sided and progressive loss of hearing, otorrhea, with pulsatile tinnitus during the previous 10 months. No other comorbidities were found. Acoustic testing revealed conductive hearing loss. Otologic examination showed a reddish lesion beneath an intact tympanic membrane. A glomus tumour was suspected based on clinical examination. MRI revealed a left middle ear mass occupying the middle ear and the hypotympan, lateral to the cochlea and extending anteriorly into the Eustachian tube (Figure 1). The mass showed an intense enhancement with no evidence of jugular or intracranial involvement (Figure 2). The preoperative aspect was suggestive of a vascular mass. Surgery was then performed. And histopathological examination with immunohistochemistry was consistent with a glomus tympanicum tumour.

Discussion

Two different types of glomus tumours can develop inside the temporal bone. These are the glomus tympanicum and the glomus jugular [1]. Glomus tympanicum tumours (GT) also called paragangliomas and chemodectomas develop from non-functioning chemoreceptors within the temporal bone named the glomus bodies and are histologically similar to pheochromocytoma. They can be found along Jacobson's nerve, a tympanic branch of the glossopharyngeal nerve that receives sensations from the middle ear and the bony Eustachian tube, and also provides parasympathetic fibres to the parotid gland. They are rich in blood vessels, both capillary and precapillary intermingled with epithelioid cells making them hypervascular. The inferior tympanic artery, a branch of the ascending pharyngeal artery provides blood supply [2-3]. Regardless of being an uncommon tumour, GT is the most frequent middle ear tumour [4], and the temporal bone's second most prevalent tumour. The annual incidence of GT is estimated around at 1 case per 1.3 million people [1]. The fifth and sixth decades have the highest rates of occurrence. There is a female predominance (F:

M 2:1 to 5:1 from literature) [4], and a hormonal (estrogen) influence has been suggested [3]. Both ears are equally likely to be affected [4]. Guild, in 1941, identified "glomic tissue" in the temporal bone. Glomus tissue is a form of vascular tissue that may be found in the dome of the jugular bulb and middle ear promontory. The term glomus tumour was used by Rosenwasser in 1945 to describe mastoid and middle ear "carotid body tumour". Following this, the name glomus tympanicum was used by Guilford and Alford to refer to tumours that are only contained within the middle ear [1]. The name "glomus" is vague; it was assumed to derive from actual glomus complexes. However, paraganglions that are often adjacent to sympathetic ganglions are the origin of glomus tumours [1].

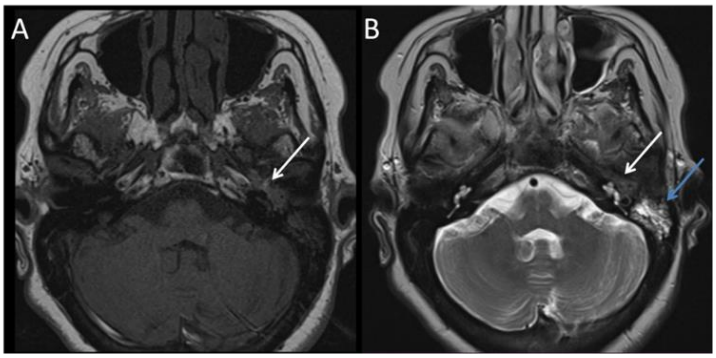

Figure 1: *A: Axial T1WI (A) and T2WI (B) revealing a left middle ear lesion slightly hyperintense on T1 and T2 with anterior extension along the Eustachian tube (white arrow). Note the fluid retention within the left mastoid cells (blue arrow).*

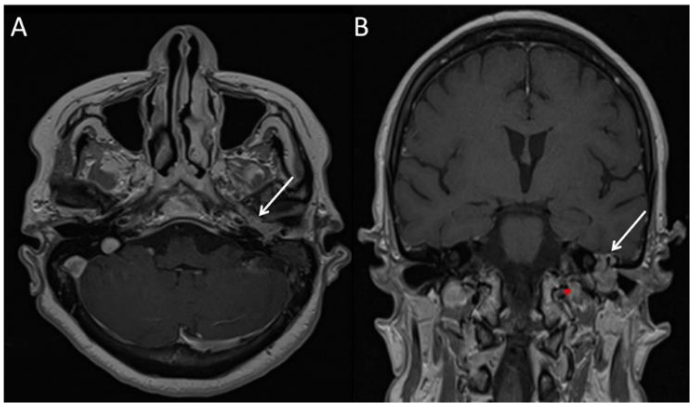

Figure 2: *A: Axial T1WI after gadolinium injection revealing middle ear enhancing lesion with anterior extension along the Eustachian tube (arrow). B: Post-contrast coronal T1WI demonstrating the vascular mass (arrow) and no extension into the jugular bulb (arrowhead).*

During otoscopy, GT appears as a pulsating reddish-bleu mass behind an intact tympanic membrane also named the ''vascular tympanic membrane'' [3,4]. These tumours have a slow growth rate that manifest through mass effect or by eroding nearby structures. The most frequent sign includes pulsating tinnitus which affects 82% of patients while 56% experience hearing loss. Rarely, patients may present vestibular signs, otalgia, or otorrhagia [1-4]. When the tumour prevents the ossicles from vibrating normally, conductive hearing loss occurs. In rare cases, sensorineural hearing loss and/ or vertigo might occur if the tumour spreads to the inner ear [1]. Upon clinical suspicion, imaging is required to confirm the diagnosis. For initial imaging, a CT scan is typically preferred [4]. GT are recognized by its distinctive placement in the tympanic cavity, lateral to the cochlear promontory. They may also indicate the presence of aberrant blood arteries, particularly an aberrant internal carotid artery [2]. To rule out the possibility of a glomus jugular tumour, it is crucial to determine whether or not the jugular bulb is intact [2]. GT are poorly destructive; can grow along pathways of low resistance [1-3], invade the eustachian tube, and dissect along fascial planes [2]. On CT scans, they should be divided into those with or without hypotympanic involvement. This sub-classification would determine the surgical approach to achieving complete tumour excision [4]. They appear as a mass of soft tissue adjacent to the middle ear's promontory. The diagnosis is almost certain if there is bone or air between the jugular bulb and the tumour [1]. A further MRI investigation is needed to evaluate hypotympanic extension. It excludes tumour extension into the jugular bulb wall and outside the middle ear [4]. Dynamic contrast-enhanced MRI distinguishes the tumour from the jugular bulb's blood flow presenting signal-void. On T1, GT typically show hypointense or intermediate signal (salt and pepper appearance), while on T2 they appear hyperintense. T1-weighted images are helpful in ruling out intracranial extension, whereas T2-weighted images distinguish GT from cholesteatoma/middle ear fluid [1-4]. Angiography, if performed will show an early, strong, and inhomogenous blush of the lesion connected to Jacobson's nerve [3]. GT should be distinguished from facial nerve neuroma, middle ear adenocarcinoma, aberrant internal carotid artery, and high jugular bulb. Despite the advanced imaging, a correct diagnosis prior to surgery could be challenging [4]. Nasopharyngeal and middle ear pathologies that spread along the Eustachian tube should also be considered in the differential diagnosis [2]. Various classifications exist. Fisch's classification which was later revised by Oldring and Fisch is the one frequently used. Type A represents a tympanicum tumor, one of the four distinct categories defined. Glasscock and Jackson suggested a second classification separating glomus tympanicum from glomus jugular tumours [4]. Neither of these systems is optimal in providing the best surgical indications for tympanicum tumours in this regard [4]. It is prudent to presume that the jugular bulb is involved if there is any uncertainty and to treat it like a jugular tumour [4]. It is essential to accurately stage the tumour before surgery so that appropriate treatment can be planned. The available treatments could be radiation therapy, surgery, or a combination of the two. Observation may also be an option [4].

The choice of therapy is heavily influenced by the stage of the tumour, the patient's age, and his general well-being. Follow-up is recommended only for small asymptomatic lesions [2]. Approximately 40% of irradiated tumours keep growing following radiation therapy, despite the fact that radiotherapy can momentarily stop tumour growth. Thus, surgical removal is the best treatment option because of its minimal risk and favourable outcomes over the long term [4]. Feeding vessel embolization if done less than 48 hours prior to surgery is beneficial [1].

Conclusion

Glomus tympanicum is the most common middle ear tumour; it should be subdivided into two categories depending on the extension to the hypotympanum. MRI permits the differentiation between large tympanicum tumours from jugular tumours requiring two different surgical approaches.

Author Contributions

All authors contributed equally to this work.

Patient consent

Informed consent for publication was obtained

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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