



Case Report

PARAGANGLIOMA OF THE MIDDLE EAR – CASE REPORT

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ABSTRACT

Paraganglioma or glomus tumours are named according to their origin. Glomus tympanicum tumours are small-sized tumours originating in the middle ear. Benign tumours in general grow slowly and do not spread to other organs. Their malignant counterparts have a faster growth pattern and can spread to other parts of the body. Usually, paragangliomas behave as benign tumours. Symptoms of these tumours include pulsatile tinnitus or ringing in the ears that is heard with each heartbeat. We report the case of a 48-year-old female with large middle-ear paraganglioma. We describe the diagnostic steps and surgical treatment by the retroauricular (tympanomastoid) approach.

Key Words: glomus tympanicum; paraganglioma; middle-ear tumours

INTRODUCTION

Paragangliomas are benign, slow growing tumours that arise from neuroectodermal tissues. They can occur anywhere along the sympathetic paraganglial chains from the neck to the pelvis, but they occur most often intra-abdominally in the periaortic region and the organ of Zuckerkandl (1). They also have been reported in other unusual sites, such as the gallbladder, the biliary duct system, and the urinary bladder. In the head and neck, two anatomic groups of paragangliomas can be differentiated: cervical paragangliomas and temporal bone (jugulotympanic) paragangliomas. The cervical group includes carotid body tumours and glomus vagale tumours, while the jugulotympanic comprises glomus jugulare and glomus tympanicum tumours. Glomus tympanicum tumours are more common than glomus tumours around the jugular vein, and are the most common primary neoplasm of the middle ear and the second most common tumour of the temporal bone (2).

CASE REPORT

A 48 year-old female presented to the ENT clinic Saint Marina University Hospital –

Varna with a history of pulsatile tinnitus of three years and intermittent otorrhea from the left ear since one year. There was no history of vertigo. Clinical examination revealed stenosis of the left external auditory canal and presence of a pulsatile mass behind and through the left tympanic membrane. Conductive hearing loss has been determined. CT examination of the temporal bones demonstrated an enhancing mass occupying the cavum tympani, the mastoid air cells with extension to external auditory canal (**Figure 1**). The ossicles were missing and the tumour mass engulfed the tympanic membrane.

The glomus tympanicum was removed via a retroauricular approach. We provided transmeatal, transmastoid excision of the glomus tumour by the post-aural route. Tumour was present in the external auditory meatus and filling the middle ear. No intraoperative complications occurred, and gross removal of the tumour was achieved. The intraoperative blood loss was in the normal frontiers. In the postoperative period the cavity was clean and dry. A transient facial nerve weakness was present for two weeks, which subsided spontaneously. Two years after tympanomastoidectomy there were no recurrences. Findings on postoperative audiometry were unchanged from preoperative levels.

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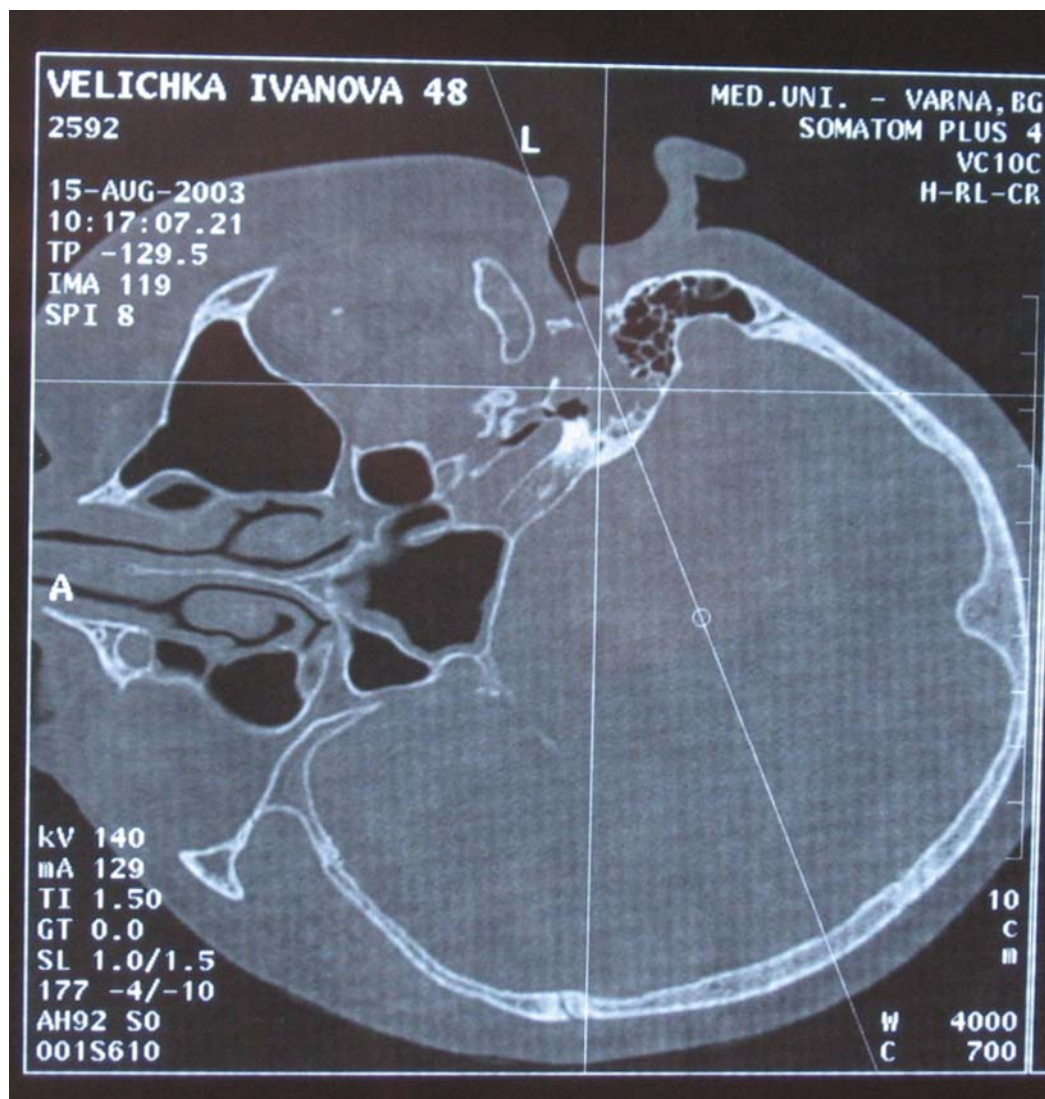


Figure 1. CT examination of the temporal bones demonstrated the tumour occupying the cavum tympani, the mastoid air cells with extension to external auditory canal

DISCUSSION

Paragangliomas are rare neuroendocrine tumours derived from the extra-adrenal paraganglia. Head and neck paraganglia are associated with the parasympathetic nervous system, the largest being the carotid body, with others found at the vagus nerve, the jugular bulb, the tympanic branch of the ascending pharyngeal artery, the larynx, and other sites. Head and neck paraganglia have the same histological characteristics as those in the abdomen, but their role, well defined in the carotid bulb, while less so elsewhere, is more likely to be sensory than secretory. Paraganglia are aggregations of cells derived from the embryological neural crest and are located throughout the body in the vascular and neuronal adventitia (3). Paraganglia of the temporal bone are ovoid, lobulated bodies, usually found accompanying Jacobson's nerve (inferior tympanic branch of the glossopharyngeal nerve) or Arnold's nerve

(mastoid branch of the vagus nerve) or in the adventitia of the jugular bulb. Serving as baroreceptors, paraganglia sense and regulate oxygen pressure in middle ear and mastoid.

Microscopically, they consist of clusters of Type I or catecholamine-containing chief cells and Type II or sustentacular cells (modified Schwann cells), intimately interlaced with a rich network of capillaries and venules (4). Paragangliomas are strongly vascularised. From a histological view, they have a thin capsule and are composed of round polygonal chief cells arranged in compact cell nests ('Zellballen') or trabecular formations. Spindle-like or supporting (sustentacular) cells are situated peripherally. In benign paragangliomas pleomorphism of the nucleus, necrosis, mitosis and some local invasion may be observed (5).

In the 1990s, there were published studies investigating a five-generation

pedigree with evidence of familial paragangliomas employed linkage analysis to map a 'susceptibility locus' (i.e. a chromosomal region likely to harbour the gene responsible for the condition studied) on chromosome 11 (band 11q23), labelled 'paraganglioma locus 1' (PGL1) (6). All patients presenting a paraganglioma with a family history of paraganglioma or pheochromocytoma and those with no family history but multiple paragangliomas or a paraganglioma and a pheochromocytoma should be offered genetic testing (7).

If the studies confirm that the tumour is very small and limited to the space behind the eardrum, the tumour is classified as a glomus tympanicum. If the tumour extends from the ear down into the internal jugular vein in the jugular foramen, preparations must be made for a larger type of surgical procedure to remove the tumour completely. Tumours in this location are referred to as glomus jugulare and tend to be larger at the time of diagnosis than other types of glomus tumours. The Glasscock and Jackson's system classifies glomus tympanicum by area and degree of involvement into four types. While Type 1 glomus tumours are limited to the promontory, Type 2 denotes tumour completely filling the middle ear. Type 3 indicates tumour extending further into mastoid, whereas Type 4 glomus tumours spread into external auditory canal and may have intracranial extension.

The most common presenting symptoms include conductive hearing loss and pulsatile tinnitus. Conductive hearing loss occurs when tumour impairs the normal vibration of the ossicles or bones behind the eardrum. A sensorineural hearing loss or dizziness can result rarely, if the tumour has invaded the inner ear. Other symptoms may include aural haemorrhage or otorrhea, otalgia and facial palsy.

On CT scan, glomus tympanicum appears as a soft tissue mass abutting the promontory of the middle ear. There may be displacement of ossicles or bony erosion of the tympanic cavity. The finding of air or bone between the tumour and the jugular bulb virtually assures the diagnosis of a tympanicum. As the tumour enlarges, bone destruction in the middle ear cavity and/or extension into the external auditory canal may occur. CT scans are best for evaluating bony destruction and erosion, which is a hallmark of jugulotympanic glomus tumours. MRI is usually better than CT for delineating tumour edges and intracranial extent. It is also better

for evaluating the relationship of the tumour to adjacent jugular vein, carotid artery, membranous labyrinth and cranial nerves. Angiography recognizes the primary feeding blood supply to the lesion, helps in detecting multicentric tumours, identifies intrasinus and intravenous extension, provides further information on flow in contralateral sigmoid and internal jugular vein and allows for possible pre-operative embolisation. Paragangliomas and other neuroendocrine tumours are hypervascular and, therefore, have hyperintense signals on T2-weighted MR-images (8).

The differential diagnosis of benign tumours of the middle ear includes middle ear adenoma, paraganglioma, schwannoma, and schneiderian-type mucosal papilloma. The differential diagnosis of middle ear lesions also includes other benign lesions such as cholesteatoma, chronic otitis media, choristoma, and hamartoma. Primary malignant tumours of the middle ear include squamous cell carcinoma, rhabdomyosarcoma, and papillary adenocarcinoma (aggressive papillary tumour). Some other tumours, such as meningioma and jugular paraganglioma, can grow into the middle ear from the surrounding tissue. A single case of middle ear invasion by a central nervous system yolk sac tumour has also been reported (8). Metastasis to the middle ear and temporal bone from sites such as the breast, lung, kidney, stomach, larynx, parotid, and external ear and from melanomas has been reported (9).

The treatment of glomus tympanicum, largely takes into account the patient's age, the site, size, and extent of the tumour, the rate of symptom progression, the preoperative cranial nerve status, the possibility of multicentricity and neurosecretory status and lastly the patient preference. Treatment of small tumours involves less risk than larger ones. For that reason, periodical examination and early treatment are recommended. Knowledge about the mutation and expression of the disease within the family is desired (10). Surgery and radiotherapy are the two modalities of treatment available (5). In larger paragangliomas, embolisation of the main arterial blood supply preoperatively may reduce the risk of haemorrhage or other complications during surgery.

The risks of radiotherapy include tumour re-growth, late-onset cranial nerve defects and osteoradionecrosis of temporal bone. The surgical goal is total or near-total removal. Surgical risks include cranial nerve

defects, vascular injury and bleeding and cerebrospinal fluid leak. The size and the extent of the glomus tumour determine the surgical procedure needed (5). Type 1 glomus tympanicum tumours are generally approached by trascanal tympanotomy. The Type 3 tumours require mastoidectomy with extended facial recess approach. Some authors have reported that facial nerve involvement is a poor prognostic indicator (11). However, others have indicated that facial nerve involvement is most likely related to nerve compression and not invasion. Facial nerve symptoms may resolve after resection (12).

CONCLUSION

All patients with a family history of paraganglioma should be offered genetic testing. Cases presenting paraganglioma require careful consideration and early surgical removal. Residual tumours may require postoperative radiotherapy or observation with MRIs with long term follow up with ENT examinations. The well-timed surgery of the paraganglioma is a factor for the successful treatment.

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