

GLOMUS TYMPANICUM PARAGANGLIOMA IN A 63-YEAR-OLD MALE - A RARE ENTITY IN THE MIDDLE EAR: A CASE REPORT

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Glomus tympanicum paraganglioma arise from the glomus bodies that run with the tympanic branch of the glossopharyngeal nerve (Jacobson nerve). Although *glomus tympanicum* tumours are the most common primary neoplasms of the middle ear, these tumours are the rarest of head and neck paragangliomas. In most cases these benign tumours grow slowly, but can be locally aggressive. We present the case of small *glomus tympanicum* in a 63-year-old male patient with one year long history of pulsatile tinnitus and ear fullness as well as decreased hearing. On otoscopy, a pulsatile reddish mass was seen behind the tympanic membrane and Brown sign was elicited. Imaging revealed middle ear cavity mass on the cochlear promontory with strong post contrast enhancement. The knowledge of the clinical presentation, the imaging features and the differential diagnosis of the middle ear masses is necessary to establish the correct diagnosis. This case illustrates small size *glomus tympanicum* paraganglioma with typical clinical and imaging findings. Imaging and imaging based classification play most important roles in diagnostic and treatment planning in patient with *glomus tympanicum* paragangliomas.

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Key words: *glomus tympanicum paraganglioma, imaging, classification*

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Introduction

Head and neck paragangliomas (also commonly referred to as glomus tumours or chemodectoma) are highly vascular lesions originating from the paraganglionic tissue, located at four typical locations: the carotid bifurcation (carotid body tumours), along the vagus nerve (vagal paragangliomas), and in the jugular fossa and tympanic cavity (jugular and tympanic paragangliomas) (1, 2, 3). These tumours originate from the chief cells of the paraganglia, or glomus bodies, located within the wall (adventitia) of the jugular bulb, and can be associated with either the auricular branch of the vagus nerve (Arnold nerve) or the tympanic branch

of the glossopharyngeal nerve (Jacobson nerve). Paraganglia develop from the neural crest and are believed to function as chemoreceptors (4).

Paragangliomas account for 0.6% of all neoplasms of the head and neck and 0.03% of all neoplasms (5).

These lesions are histologically similar to the pheochromocytomas that may develop in the adrenal medulla, but unlike pheochromocytomas, head and neck paragangliomas rarely secrete catecholamines (6).

In most cases they are benign neoplasms; overall less than 10% of all paragangliomas have been cited to be malignant (5).

Tumours in the area of the middle ear cavity are called *glomus tympanicum* paragangliomas; they arise from the glomus bodies that run with the tympanic branch of the glossopharyngeal nerve (Jacobson nerve). Although *glomus tympanicum* tumours are the most common primary neoplasms of the middle ear, these tumours are the rarest of head and neck paragangliomas (7, 8, 9, 10).

Case report

We present a 63 year old male patient with one year long history of pulsatile tinnitus in the right ear. The tinnitus was accompanied by ipsilateral ear fullness. Last few months patient noted decreased hearing on the right side. There was no history of

headache, vertigo and facial weakness, as well as otorrhea and ear pain. No other comorbidities were present.

On otoscopy, a pulsatile reddish mass was seen behind the tympanic membrane and Brown sign was elicited. Audiometry revealed conductive hearing loss in the right ear. The left ear had normal audiometric finding.

The patient came to our department after clinical examination which was made few days before imaging. The first diagnostic method in our department was computed tomography (CT).

CT demonstrated oval middle ear cavity mass on the cochlear promontory. The diameter of the mass was 8 mm. There were no ossicular chain destruction and no evidence of bony erosion. The vestibule, cochlea and semicircular canals had normal morphology. The carotid canal had normal configuration and it was intact. There was no evidence of dehiscence of the internal jugular vein bulb. No abnormalities were seen along the course of the facial nerve canal. The postcontrast CT study resulted in intense enhancement of the cochlear promontory mass, which was well-defined and slightly bulged the tympanic membrane.

The next diagnostic tool was magnetic resonance imaging (MRI). The mass demonstrated low T1 weighted and high T2 weighted signal intensity on MR images. It presented intense enhancement after contrast administration.

According to the CT and MR presentation of the middle ear cavity mass, as well as clinical presentation, otoscopic examination and audiometric findings, we made the diagnosis of glomus tympanicum paraganglioma.

The patient underwent excision of the glomus tumour via a transcanal approach. Follow up few months after surgery showed the absence of tinnitus and improvement of hearing.

Histopathological examination showed branching vascular canals separated by stroma. In the stroma, grouped glomus cells surrounding blood vessels were seen. Nuclei of these cells were small and showed low mitotic activity. Final histopathology was consistent with glomus tumour.

Discussion

Glomus tumours have an annual incidence of 1 per 1.3 million people (11). These tumours are commonly seen in 5th and 6th decade of life, with a female predominance (12).

Our patient is male in the 7th decade of life, with right sided glomus tympanicum paraganglioma.

Paragangliomas are typically described as benign and slow growing. Glomus tympanicum may be confined to the middle ear or may spread along the paths of least resistance to the mastoid or even up to the nasopharynx. In our patient the tumour was on cochlear promontory in the middle ear cavity, without spread to surrounding structures.

Glomus tympanicum may present with pulsatile tinnitus and hearing loss, and our patient had these symptoms. Other symptoms may include head-

ache, vertigo and facial weakness, as well as otorrhea and ear pain. Tachycardia, hypertension, tremors and headaches can indicate the possibility of a functional tumour. None of these symptoms were present in our patient.

On otoscopy, a pulsatile reddish mass was seen behind the tympanic membrane and Brown sign was elicited. Brown's sign is the pulsations elicited by pneumatic compression of the tumour. All these signs were present in our patient.

Imaging is the primary modality of investigation for glomus tympanicum tumours. A combination of contrast enhanced computed tomography, magnetic resonance imaging and angiography is ideal for the diagnosis and localisation of glomus tumours.

Strongly enhancing mass at its site of origin is typical in the diagnosis of glomus tympanicum tumours (13).

CT is best for evaluation of bony erosion and destruction which is demonstrated in larger glomus tympanicum. Similar to CT, contrast enhanced MRI shows strongly enhancing soft tissue mass, which is typical for diagnosis, at characteristic locations (13, 14).

Multiple areas of high and low signal intensity on MRI, the so-called "salt and pepper appearance", originally described by Olsen et al. (15) can be seen within the larger lesion on T1 weighted and T2 weighted images. The "pepper" component represents the multiple areas of signal void interspersed with the "salt" seen as hyperintense foci (due to slow flow or hemorrhage) on both T1 weighted and T2 weighted images (15, 16). This feature is especially seen in larger paragangliomas, and was not seen in our patient, who had small glomus tympanicum tumour.

Angiography identifies the primary feeding vessels to the lesion, helps in detecting multicentric tumours and allows for possible preoperative embolisation. Angiography is imaging procedure which can confirm the diagnosis of a head and neck paraganglioma and some authors report that it is still the gold standard in detection of small paragangliomas (15, 17, 18). Angiography shows the specific vascular supply of the paraganglioma (19). The ascending pharyngeal artery can be considered "the artery of the paraganglioma", because its branches can supply tympanic, jugular, vagal, carotid, and even laryngeal paragangliomas (20). Embolisation can be performed with angiography for preoperative devascularisation of the large tumour.

This diagnostic procedure was not performed in our case.

Surgical resection is the treatment of choice especially in small paragangliomas. The surgical approach depends on the size and extent of the tumour. Preoperative image based classification of glomus tympanicum paragangliomas is essential, since the operative approach will be chosen depending on the tumour stage (21).

The tympanic paragangliomas present usually like a small mass at the cochlear promontory, ossicular destruction is not common and is especially present in larger lesions. The extension of temporal bone destruction is important to classify those tu-

mours. The two established classifications of temporal bone paraganglioma are based mainly on tumour size with special emphasis on intracranial extension as a decisive factor for resectability. Paragangliomas of the temporal bone, as well as the glomus tympanicum paragangliomas are usually staged according to the classification by Fisch (22) and Glasscock and Jackson (23).

In the present case, the tumour was limited on the cochlear promontory, without any destruction of the ossicular chain and surrounding bone, and it was classified as Fisch type A and Glasscock – Jackson type 1, with recommendation for surgical treatment. Type 1 glomus tympanicum tumours are generally approached by transcanal tympanotomy (24).

Surgery remains the treatment of choice in an otherwise healthy patient who desires the immediate cure of disease provided by a total resection. Preoperative embolisation is necessary in patients especially when a tumour diameter is larger than 3 cm (4). It can reduce intraoperative blood loss significantly.

Paragangliomas may be treated with either surgery or radiotherapy. The goal of any treatment is to control tumour growth and prevent further neurological compromise and it is important to select the intervention with the greatest chance of tumour control and the lowest risk of complications (25).

However, radiotherapy is often used for larger slow growing tumours with an aim to restrict growth in unresectable tumours. Preoperative evaluation with CT, MRI and angiography has been proposed as essential for optimal planning (26).

The differential diagnosis considerations for the middle ear mass include benign neoplasms and diseases (adenoma, endolymphatic sac tumours, choristoma, cholesteatoma, cholesterol granulomas) and malignant neoplasms (squamous cell carcinoma, adenocarcinoma, sarcoma). Adenomatous tumours (mixed pattern type) of the middle ear manifest as soft-tissue masses in the tympanic cavity, usually without destruction of surrounding osseous structures. These tumours may enhance intensely following the intravenous administration of contrast material (27). Endolymphatic sac tumours of the temporal bone arise from the region of vestibular aqueduct. Although they may closely resemble glomus tumours, their origin from the vestibular aqueduct is a useful discriminating characteristic. They are frequently quite large at the time of presentation, demonstrate lytic changes in the temporal bone and characteristically have regions of hyperintensity on T1-weighted sequences on MRI(28).

Cholesterol granuloma occurs secondary to nonspecific chronic inflammatory changes and it can be seen in the middle ear cavity. Hemorrhage, cholesterol, and granulation tissue are the hallmarks of the lesion and account for its appearance on MRI (29). Patients most often present with conductive

hearing loss and a vascular appearing retrotympenic mass on physical examination. On CT, the imaging appearance is nonspecific with a soft tissue middle ear mass. Findings on MRI are more characteristic, demonstrating a nonenhancing middle ear mass which is hyperintense on both T1 and T2 sequences secondary to hemorrhagic blood products (30). Care must be taken to compare pre and postcontrast T1 sequences, since the lesions are hyperintense on both which may be mistaken for enhancement. With adjacent inflammation, there may be thin peripheral enhancement.

Cholesteatoma also can be found in the middle ear cavity, but on the MRI it exhibit specific findings with restriction of diffusion and does not demonstrate the postcontrast enhancement. Cholesterol granuloma and cholesteatoma usually are not located at cochlear promontory.

Glomus tympanicum tumours are usually described as arising on the promontory (31).

Cavernous hemangioma is a rare benign neoplasm of the middle ear, which can rarely cause tinnitus and mimic a glomus tympanicum clinically, radiographically and otoscopically (32). Middle ear hemangiomas should be differentiated from the more common geniculate ganglion hemangiomas (33), which usually present with facial nerve palsy (34) but may rarely cause pulsatile tinnitus (35, 36). The diagnosis is often possible only at surgery. The literature review revealed one case of hemangioma localized on the promontory of the middle ear confirmed by histopathological study as capillary hemangioma (37).

In our case the diagnosis is made after clinical examination and imaging, and confirmed after histopathological examination.

Conclusion

In conclusion, we report the case of a rare entity - glomus tympanicum paraganglioma in a 63 old male patient with one year long history of pulsatile tinnitus and ear fullness as well as decreased hearing. Imaging revealed middle ear cavity mass on the cochlear promontory with no ossicular chain destruction and no evidence of bony erosion. On otoscopy, a pulsatile reddish mass was seen behind the tympanic membrane and Brown sign was elicited. The imaging features demonstrated strongly enhancing mass. All these signs are important and can help to make the diagnosis of the glomus tympanicum tumour. The knowledge of the clinical presentation, the imaging features and the differential diagnosis of the middle ear masses is necessary to establish the correct diagnosis. Imaging based classification is essential in management of the treatment choice.

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Prikaz bolesnika

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doi:10.5633/amm.2019.0314**GLOMUS TYMPANICUM PARAGANGLIOM KOD 63 GODINE STAROG MUŠKARCA – REDAK ENTITET SREDNJEG UVA: PRIKAZ SLUČAJA**Filip Petrović^{1,2}, Dragan Stojanov^{1,2}, Nikola Živković^{2,3}, Jovana Zdravković²,
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Glomus tympanicum paragangliomi potiču od glomus tela lokalizovanih duž timpanične grane glosofaringealnog nerva (Jacobsonov nerv). Iako su *glomus tympanicum* tumori najčešće primarne neoplazme srednjeg uva, ovi tumori najređi su paragangliomi regije glave i vrata. U najvećem broju slučajeva ovi benigni tumori imaju spor rast, ali mogu biti lokalno agresivni. Prezentujemo slučaj malog *glomus tympanicum* paraganglioma kod 63 godine starog bolesnika muškog pola, sa jednogodišnjom istorijom puslatilnog tinitusa i osećajem punoće u uvu, kao i sa oslabljenjem sluha. Otoskopija je pokazala puslatilnu crvenu masu iza timpanične membrane kao i Brownov znak. Imidžing je pokazao masu srednjeg uva na kohlearnom promontorijumu sa izraženim postkontrastnim pojačanjem. Poznavanje kliničke prezentacije, imidžing karakteristika, kao i diferencijalne dijagnoze masa srednjeg uva, neophodni su za postavljanje tačne dijagnoze. Ovaj slučaj pokazuje *glomus tympanicum* paragangliom malih dimenzija sa tipičnim kliničkim i imidžing karakteristikama. Imidžing i imidžing klasifikacije igraju najvažniju ulogu u dijagnostičkom i terapijskom planiranju kod bolesnika sa *glomus tympanicum* paragangliomima.

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Ključne reči: *glomus tympanicum* paragangliom, imidžing, klasifikacija