



# Vertigo as the sole complaint of tympanomastoid paraganglioma

Xi-Xing Li <sup>a</sup>, Wei-Na Cui <sup>a,\*</sup>, Guo-Dong Gao <sup>b</sup>

<sup>a</sup> Department of Otorhinolaryngology Head and Neck Surgery, The Second Hospital of Hebei Medical University, Shijiazhuang, China

<sup>b</sup> Department of Medical Radiology, The Second Hospital of Hebei Medical University, Shijiazhuang, China

## ARTICLE INFO

### Article history:

Received 15 April 2019

Received in revised form

30 August 2019

Accepted 12 September 2019

### Keywords:

Paraganglioma

Glomus tympanicum

Vertigo

Tympanomastoid

## ABSTRACT

**Background:** Tympanomastoid paragangliomas are usually benign, slowly growing, painless tumors. The common presenting symptoms of this tumor are pulsatile tinnitus and conductive hearing loss. Vertigo as the cardinal or initial symptom is extremely rare, especially in the early stages of the disease.

**Case presentation:** A 53-year-old female patient presented only with intermittent recurrent vertigo and was later found to have a tympanomastoid paraganglioma. Her symptoms disappeared completely after resection of the tumor. This is the first report in literature of a case of tympanomastoid paraganglioma with vertigo as the single symptom.

**Conclusion:** The tympanomastoid paraganglioma is rare and its clinical symptoms are nonspecific, so it is easy to be misdiagnosed or missed. It is worth noting that although clinically uncommon, vertigo can also be the first or sole symptom of tympanomastoid paraganglioma. Detailed physical examination and imaging examination of the ear are necessary and should be carried out meticulously.

© 2019 PLA General Hospital Department of Otolaryngology Head and Neck Surgery. Production and hosting by Elsevier (Singapore) Pte Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Paragangliomas, also named glomus tumors or chemodectomas, are rare highly vascular neuroendocrine neoplasms arising from specific neural crest components, known as paraganglion cells (Ludman, 1998). Tympanomastoid paragangliomas (TMPs), also commonly known as “glomus tympanicum”, which are tumors that originate from the glomus bodies that lie along the Arnold’s nerve and the Jacobson’s nerve (Patnaik et al., 2015). They are usually benign, slowly growing, painless tumors with the potential to remain stable over years.

The common presenting symptoms of this tumor are pulsatile tinnitus and conductive hearing loss. However, in those cases of TMP, vertigo as the cardinal or initial symptom is extremely rare, especially in the early stages of the disease. We described a patient with a diagnosis of TMPs who was presented with recurrent vertigo rather than the usual clinical manifestations such as tinnitus and hearing loss. Surgical treatment was chosen by the patient and she had no recurrence of vertigo after operation.

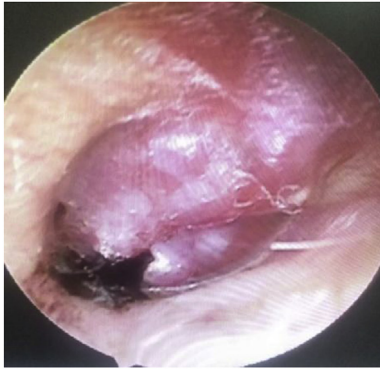
## 2. Case presentation

A 53-year-old female patient told us that she had a 4-month history of recurrent dizziness. She claimed that dizziness seemed to occur easily when lying flat and it was more of a pinning type of sensation. It was episodic and each episode lasted for several minutes. There was also history of nausea and vomiting when the dizziness was severe. She hasn’t felt any noticeable pulsatile tinnitus or hearing loss. There was no history of ear discharge, ear pain or any other ear symptoms. Also there were no nasal or throat or laryngeal symptoms. Half a month ago, in view of the swelling of the right tympanic membrane revealed by physical examination, according to the patient, doctors at the local community clinic performed a tympanocentesis on her. At that time, a little blood flowed out of perforation of the tympanic membrane.

Otосcopy showed the hyperemia and swelling of the right tympanic membrane, and the blood scab which attached to the lower part of the tympanic membrane (Fig. 1). Pure tone audiometry showed mild conductive hearing loss (15 dB) on the affected side. High resolution computerized tomography (HRCT) of the temporal bone showed soft tissue density shadow occupied the mesotympanum and hypotympanum of the right ear. The structure and position of auditory ossicles were basically normal (Fig. 2). The soft tissue lesion in the tympanic cavity demonstrated equal T1WI and long or equal T2WI signal on MRI plain scan and obvious

\* Corresponding author. No.215, Heping West Road, Shijiazhuang, 050000, China.  
E-mail address: [iamcuiweina@126.com](mailto:iamcuiweina@126.com) (W.-N. Cui).

Peer review under responsibility of PLA General Hospital Department of Otolaryngology Head and Neck Surgery.



**Fig. 1.** Otoscopy showed the hyperemia and swelling of the right tympanic membrane, and the blood scab attached to the lower part of the tympanic membrane.

enhancement on the enhanced scan. But there also was some dotted or striped low signals scattered init and not enhanced under the enhanced scan. So it presented a typical “salt and pepper” sign (Fig. 2). Combined with the specialist examination and imaging examination, we diagnosed it as TMP. Because the differential diagnosis of vertigo is very extensive, at the same time, we have also done bedside examination and vestibular function examination related to vertigo and no obvious abnormality was found in these examinations.

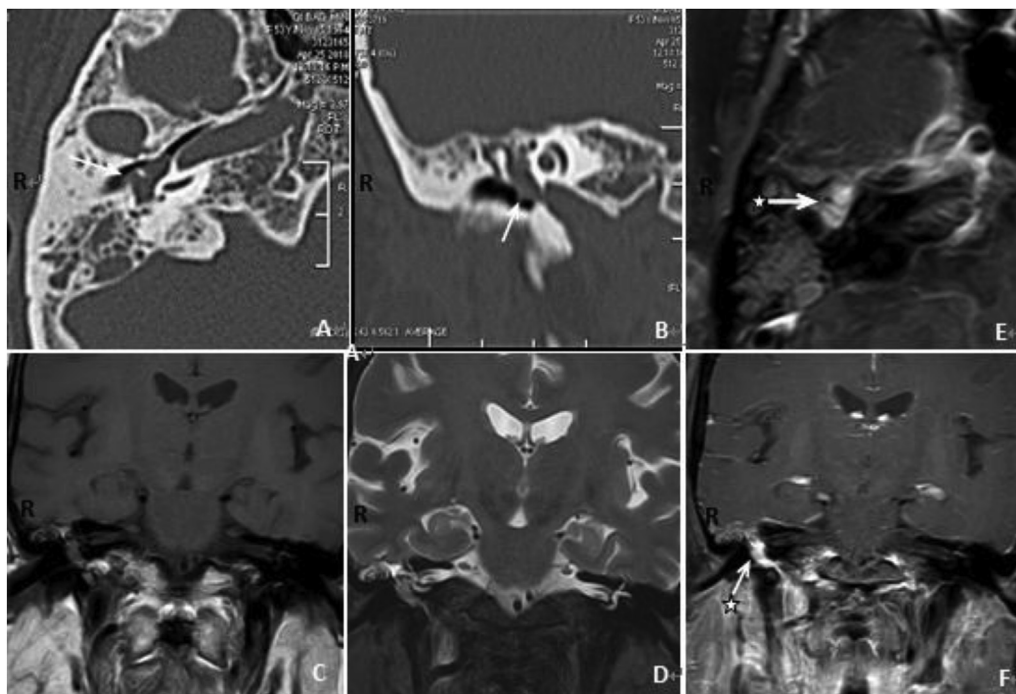
There are three treatment options for this condition: observation, surgical excision, and radiotherapy. After fully informed, the patient chose surgical treatment. We performed transmeatal, transmastoid excision of the TMP via a retroauricular approach. The tumor filled the middle ear and extended towards the eustachian tube. Partial bone in the anterior inferior wall of the promontory of the tympanum was invaded. After the complete excision of the tumor, we used autogenous cartilage for tympanoplasty.

Histopathological examination of the mass revealed features consistent with that of TMP (Fig. 3). At her latest review (6 months post-operative), vertigo didn't recur, and she had only a mild ipsilateral conductive hearing loss (10 dB) without self aware hearing loss.

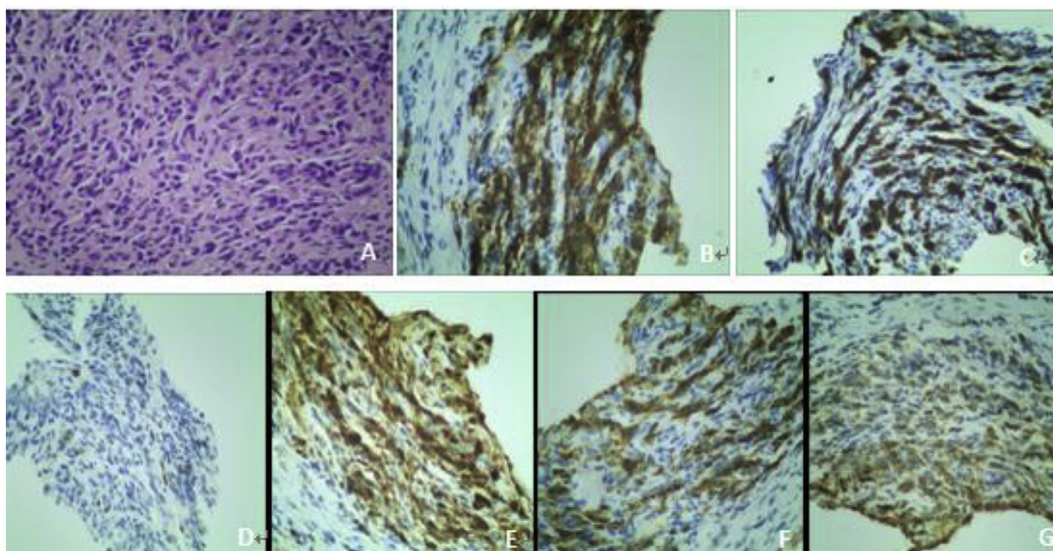
### 3. Discussion

Paraganglioma is a rare neoplasm originating from neuroectodermal tissue. It has been described in many anatomic locations, most commonly in the abdomen. In the head and neck, paragangliomas are divided into three types: carotid body tumors (the most common), vagal body tumors and jugulotympanic tumors (Pellitteri et al., 2004). The jugulotympanic paragangliomas include TMP and tympano jugular paraganglioma (TJP) or “glomus jugulare” (Scherner et al., 2015). TJPs are the most common head and neck paragangliomas while the TMPs are rare (Reddy, 2011).

TMPs are pathologically benign, locally invasive middle ear tumors. They grows lowly, along the path of least resistance around them, and can also cause local damages. It should be noted that TMPs are more common in women, with a peak age of 40–70 years (Carlson et al., 2014). The most common clinical symptoms include pulsating tinnitus and conductive hearing loss (Teixeira et al., 2005). Given their anatomic predisposition, it is perhaps unsurprising that patients most commonly present with hearing loss and pulsatile tinnitus. Conductive hearing loss, which is significantly more common, is often caused by ossicular mass loading, ossicular discontinuity, as well as tympanic membrane or round window obstruction (Walker et al., 2019). It rarely causes sensorineural hearing loss and/or dizziness, only when the tumor invades the inner ear structure. Other symptoms may include otorrhea or ear hemorrhage, ear pain and facial paralysis. Signs and symptoms of TMPs may vary on an individual basis for each patient. It is worthy of note that this particular patient who presented only



**Fig. 2.** HRCT Scan of the temporal bone (Fig.2A, Fig.2B) showing a soft tissue density occupying the mesotympanum and hypotympanum of the right middle ear (→). The lesion demonstrated equal T1WI and long or equal T2WI signal on MRI plain scan (Fig.2C, Fig.2D) and obvious enhancement on enhanced MRI scan (Fig.2E, Fig.2F). But there were also some dotted or striped low signals scattered init and not enhanced under enhanced scan. It presented a typical “salt and pepper” sign (\*→).



**Fig. 3.** Consistent with TMP (Combining with histomorphology and immunophenotype). A: HE ( $\times 200$ ). (B–G): Phenotype of immunohistochemistry ( $\times 200$ ), (B) CD56 (+), (C) CgA (+), (D) Ki-67 (+1%), (E) S-100 (+), (F) Syn (+), (G) Vimentin (+).

intermittent vertigo did not have the most common symptoms of TMP. We considered that her vertigo was related to the lesion that invade the promontory of the tympanum and thus affected the inner ear.

In order to reduce misdiagnosis and missed diagnosis, detailed physical examination and imaging examination of the ear are necessary. The first step in investigation requires a careful clinical examination. Observation of the tympanic membrane under the endoscope will occasionally reveals a pulsating mass which can be soft and often turns white on palpation. CT scan remains the most valuable method of imaging giving information about the extent of bone destruction caused by the tumor. MRI is usually superior to CT in describing the extent and margin of intracranial lesions. It is also better for evaluating the relationship of the tumor to adjacent carotid, artery jugular vein, cranial nerves and membranous labyrinth (Vogl et al., 1993). MRA has also demonstrated its value in diagnosis. In addition, histopathological examination is necessary to differentiate this tumor from others.

For the TMPs, the preferred treatment is surgical removal of the tumor. They have historically been addressed via a microscopic approach through either the canal or a postauricular incision. Recently, some authors describe a successful resection method of endoscopic resection of TMPs in some adults who had undergone endoscopy assisted resection (Noel et al., 2018). The endoscope provides superior visualization of these spaces and may, in some instances, allow surgeons to avoid an incision. Furthermore, adding high-definition imaging to the procedure has transformed the applications of endoscopic surgery, making it a viable and comparable alternative to traditional microscopic techniques. In addition, advances in radiotherapy, especially gamma knife radio surgery, have been shown to control tumor growth and reduce the risk of treatment-related cranial nerve injury (Jacob et al., 2015). Radiotherapy has been performed and can be offered in select patients with significant comorbidities and unfavorable preoperative risk stratification. Recently, some scholars reported the long term surgical outcomes in TMPs in the largest series published to date. The report points out that surgery is the best treatment option for TMPs as it achieves total tumor removal, improved postoperative hearing and low rates of recurrence and complications. Furthermore, they pointed out that radiation or wait-and-scan has no role in the treatment of TMPs (Patnaik et al., 2015).

#### 4. Conclusion

TMP is rare and its clinical symptoms are nonspecific. It is easy to be misdiagnosed because some clinicians are inexperienced in the diagnosis and treatment of this disease or patients refuse to undergo further detailed examination. It's remarkable that this particular patient did not have the classical signs of TMP such as a middle ear mass, pulsatile tinnitus and conscious hearing loss. All-round examination of the ear should be carried out carefully in all cases so as to make a definite diagnosis earlier.

#### Declarations of interest

All authors have no conflicts of interest relevant to this paper.

#### Funding

None.

#### References

- Carlson, M.L., Sweeney, A.D., Pelos, S., et al., 2014. Glomus Tympanicum: a review of 115 cases over 4 decades. *Otolaryngol. Head Neck Surg.* 152 (1), 136–142.
- Jacob, J.T., Pollock, B.E., Carlson, M.L., et al., 2015. Stereotactic radiosurgery in the management of vestibular schwannoma and glomus jugulare: indications, techniques, and results. *Otolaryngol. Clin. N. Am.* 48 (3), 515–526.
- Ludman, H., 1998. Tumors of middle ear and mastoid. In: *Diseases of the ear*, 6th Edition, pp. 463–472.
- Noel, J.E., Sajjadi, H., 2018. KTP-laser-assisted endoscopic management of glomus tympanicum tumors: a case series. *Ear Nose Throat J.* 97 (12), 399–402.
- Patnaik, U., Prasad, S.C., Medina, M., et al., 2015. Long term surgical and hearing outcomes in the management of tympanomastoid paragangliomas. *Am. J. Otolaryngol.* 36 (3), 382–389.
- Pellitteri, P.K., Rinaldo, A., Myssiorek, D., et al., 2004. Paragangliomas of the head and neck. *Oral Oncol.* 40 (6), 563–575.
- Reddy, R., 2011. An unusual cause of vertigo and syncope: a case report. *Hawaii Med. J.* 70 (1), 16–17.
- Schermer, K.L., Pontius, E.E., Dziabis, M.D., et al., 2015. Tumors of the glomus jugulare and glomus tympanicum. *Cancer* 19 (9), 1273–1280.
- Teixeira, A., Al-Mefty, O., Husain, M.M., 2005. Paragangliomas of the skull base. Chapter 49. In: *Textbook of Neuro-Oncology*, 5, pp. 366–370, 4.
- Vogl, T.J., Mack, M.G., Juergens, M., et al., 1993. Skull base tumour gadodiamide injection-enhanced MR imaging. *Radiology* 188 (2), 339–346.
- Walker, D.D., Babu, S., 2019. Temporal bone paraganglioma: hearing outcomes and rehabilitation. *J. Neurol. Surg. Part B Skull Base* 80 (2), 209–213.