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Glomus Tympanicum Paraganglioma in Middle Aged Adult Female: CT & MRI Imaging Findings

Dr. Tejal Patel[1], Dr. Hinal Bhagat[2]

Resident Doctor

Associate Professor

Abstract: ***Glomus tympanicum paraganglioma tumor is the most common primary neoplasms of the middle ear and the second most common tumor of the temporal bone.[1] We report a case of 42-year-old female patient with glomus tympanicum. She had symptoms for the last 7 years. She was diagnosed with the help of radiological investigative modalities. The clinical feature and it's radiographic computed tomography and magnetic resonance imaging [CT & MRI] features are mentioned in literature.***

Keywords: glomus tympanicum, temporal bone tumor, primary neoplasm, middle ear, computed tomography and magnetic resonance imaging [CT & MRI]

1. Introduction

Glomus tympanicum is a benign vascular tumor and the most common primary neoplasm of the middle ear[1].Glomus tympanicum paragangliomas arise from the Jacobson nerve at the cochlear promontory. They usually present in 40-60 years of age group with most common symptoms of hearing loss and tinnitus. There is a female predominance (M:F = 1:3).

Although histologically benign, glomus tympanicum is slow growing, locally destructive and spreading along paths of least resistance. History and proper clinical examination followed by radiological investigations are required for diagnosis and management.

CT and MRI scans are the primary imaging modalities used in evaluating the size and extent of glomus tympanicum tumour, but confirmation can only be done by histopathology. Surgery and radiotherapy are the two modalities of treatment available.Complete surgical excision is the treatment of choice followed by regular follow-ups to prevent and manage recurrences. The treatment goal was to improve her quality of life and control the disease.

2. A Case Report

A 42-year-old female patient presented to new civil hospital, Surat in otorhinolaryngology department with history of right ear dischargehearing loss and bleeding. The discharge and hearing loss were there for 7 years; however, recently, the right ear started bleeding.

There was no history of vertigo. She was a known case of hypertensionand on antihypertensive medication since four years. There was no history of diabetes mellitus or ischaemic heart disease (IHD).

Clinical examination revealed stenosis of the right external auditory canal and presence of a pulsatile mass behind the right tympanic membrane. On otoscopy, a reddish mass was protruding through external auditory canal was seen. There was no evidence of seventh or other cranial nerve deficit or vestibular finding. The systemic examination was essentially normal. She has mild to moderate conductive hearing loss in the right ear in pure tone audiometry.

With a diagnosis of pulsatile vascular mass of the right middle ear, she was subjected to further evaluation by imagingCT scan with contrast examination of the temporal bone performed on 22 September 2022, which demonstrated vividly enhancing mass occupying the middle ear cavity and encasing all ear ossicles without it's erosion **[Figure. 2, 3]**. The lesion appears to protrude into right external auditory canaland shows extension into mastoid, infiltrating tegmen plate. Inferiorly, the mass was eroding the floor of the middle ear cavity with extension into the hypotympanum with involvement of right eustachian tube was seen. Superiorly in contact with the dura with thinning and erosion of the tegmen plate and was in close proximity to the jugular plate, jugular vein, sigmoid sinus, seventh nerve and labyrinth.

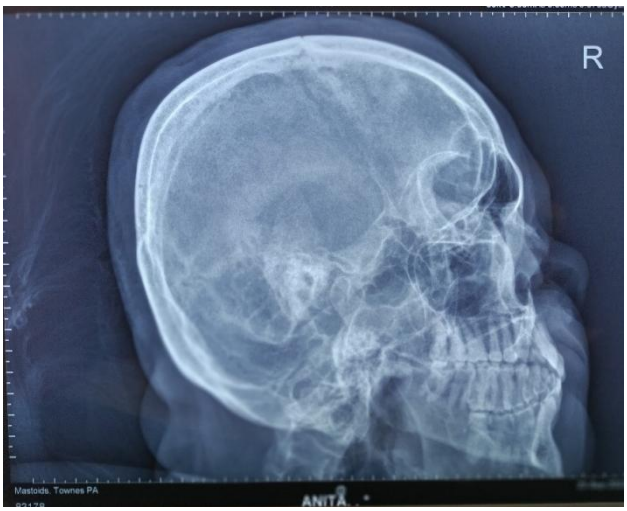


Figure 1: X-ray right lateral mastoid view showing mastoid air cell sclerosis – suggestive of changes of mastoiditis

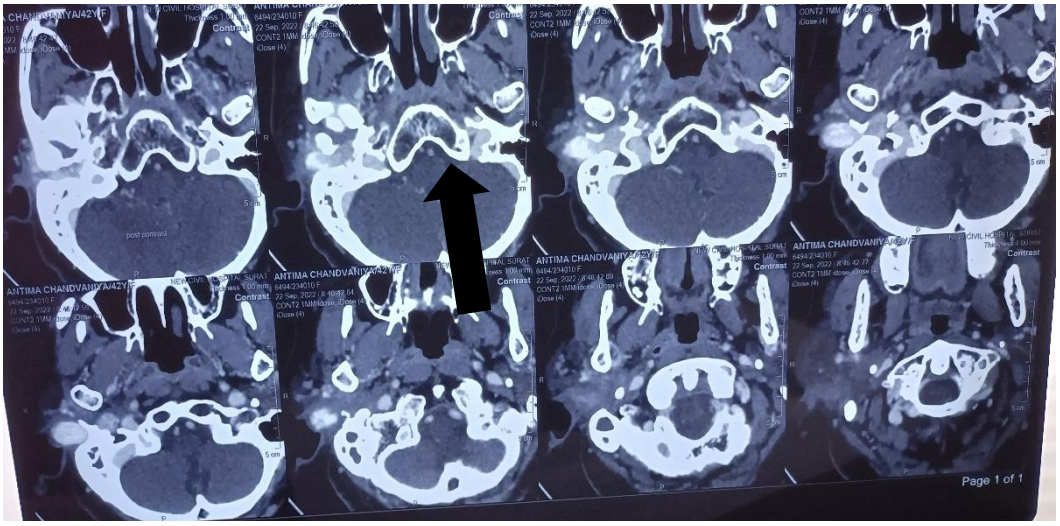


Figure 2: Axial contrast enhanced CT scandemonstrated vividly enhancing right middle ear cavity tumor extending into right external auditory canal

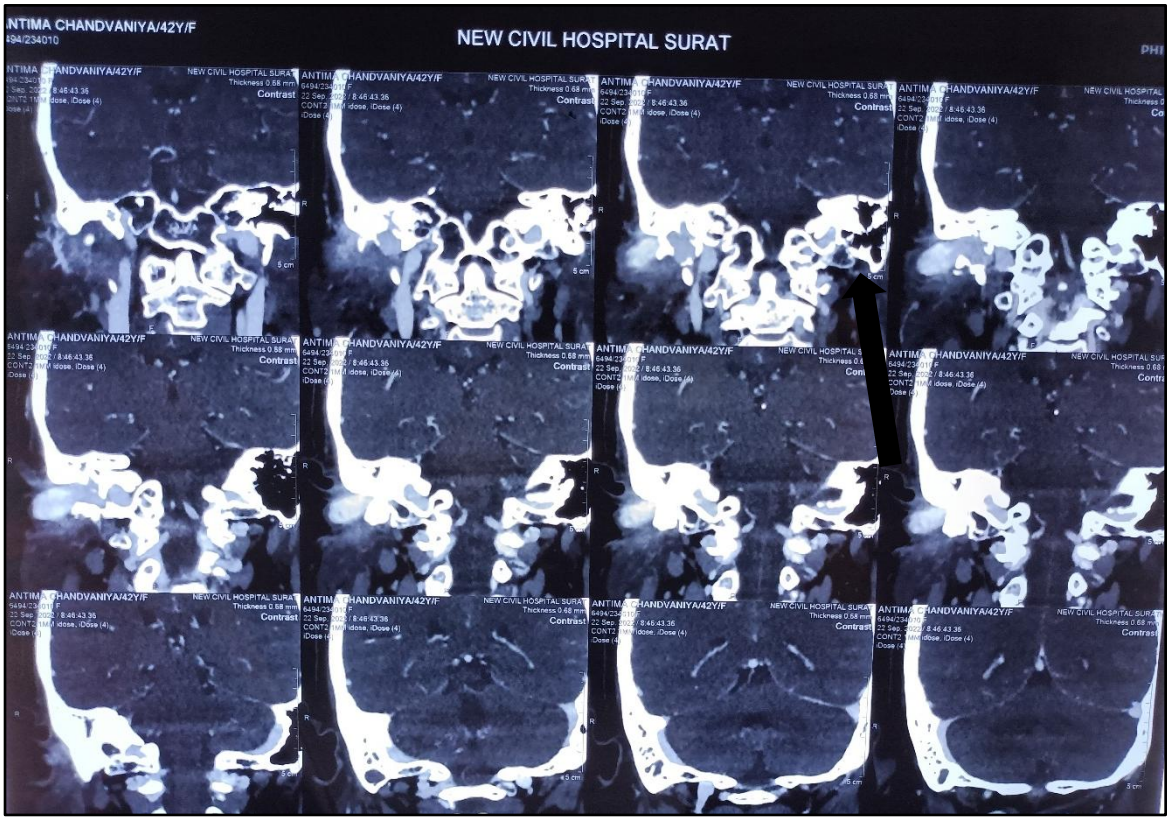


Figure 3: Coronal contrast enhanced CT scan demonstrated enhancing right middle ear cavity tumor with it's extensions

Magnetic resonance imaging (MRI) scan of the temporal bone performed on 10th November, 2022, which showed well defined lobulated exophytic altered signal lesion with epicentre in right middle ear cavity with T1 hypo intense, T2& FLAIR hyperintense signal. The lesion shows moderate contrast enhancement and similar extension and relation as mentioned above.

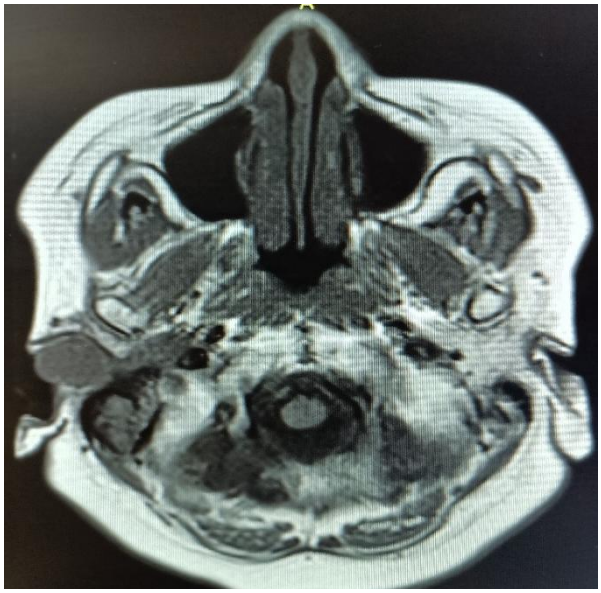
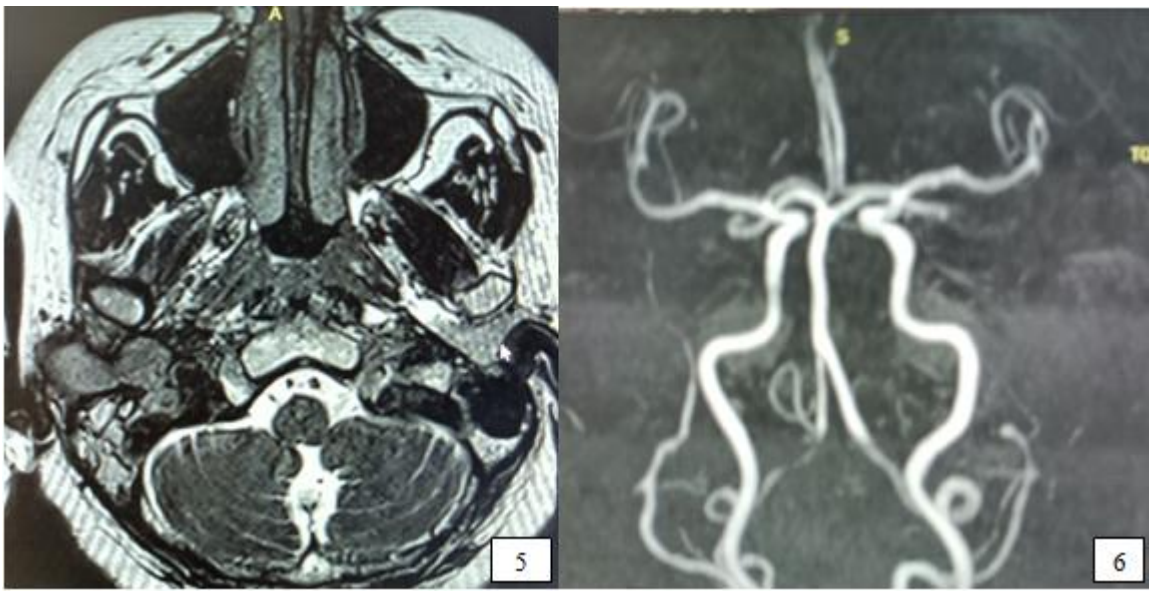


Figure 4: Well-defined T1 lobulated hypo intense lesion in right middle ear cavity



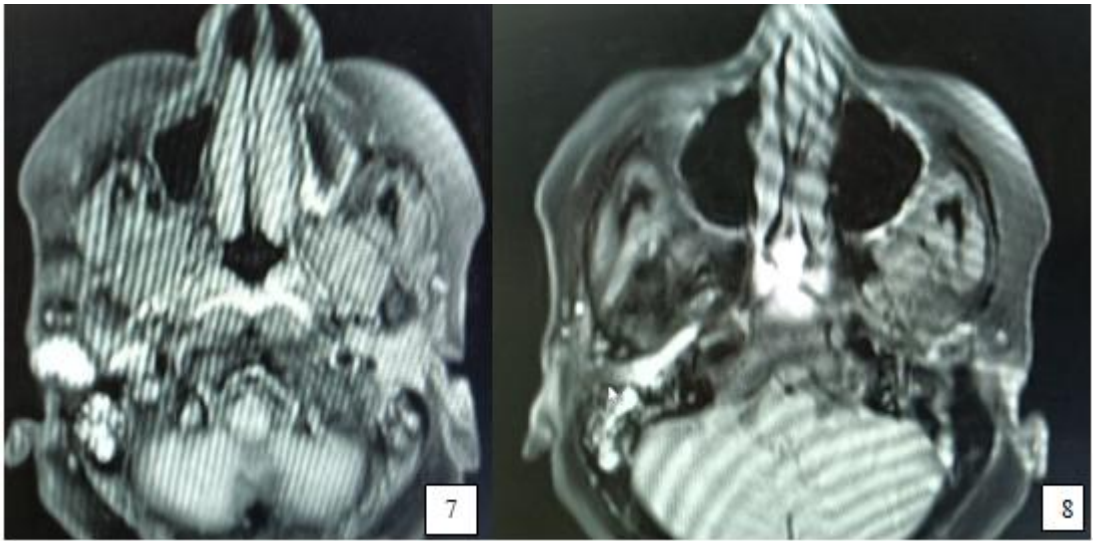


Figure 5, 6, 7, 8: T2 hyperintense, MRA shows no vessel invasion, MRI contrast showed enhancing right middle ear mass with extension into eustachian tube and external auditory canal.

Hence, the provisional diagnosis was glomus tympanicum paraganglioma (Fisch Type D) in the right ear.

3. Discussion

Paragangliomas are benign and slow growing tumors that arise from neuroectodermal tissues. Glomus tympanicum tumorsare the most common primary neoplasms of the middle ear and the second most common tumor of the temporal bone. [1]

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. The incidence of malignancy in glomus tumors is believed to be low (<5%).

Although histologically benign, glomus tumors are slow growing, locally destructive non-metastasizing, spreading along paths of least resistance. Spread is multidirectional and simultaneous. The main routes of spread are the air cell tracts of the temporal bone, but spread through and beyond the temporal bone is not uncommon, via eustachian tube, vascular lumens, and neurovascular foramina.

Patient age averages 40 to 60 years at presentation. The female: male incidence ratio is 4:1. The most common presenting symptoms include conductive hearing loss and pulsatile tinnitus. [1] Conductive hearing loss occurs when tumor impairs the normal vibration of the ossicles or bones behind the eardrum. A sensorineural hearing loss and/or dizziness can result rarely, if the tumor has invaded the inner ear. Other symptoms may include aural hemorrhage or otorrhea, otalgia and facial palsy. Brown's sign describes the pulsation elicited by pneumatic compression that is abolished with further compression. On physical examination, the hallmark of a jugulo-tympanic glomus tumor is a reddish-blue mass seen behind the tympanic membrane. Hypertension, tachycardia, tremor, or complaints of vascular headaches alerts the possibility of a functional tumor.

The Glasscock and Jackson's system [2] classifies glomus tympanicum by area and degree of involvement into four types

Type 1 glomus tumors are limited to the promontory,

Type 2 denotes tumor completely filling the middle ear.

Type 3 indicates tumor extending further into mastoid

Type 4 glomus tumors spread into external auditory canal and may have intracranial extension.

On CT scan, glomus tympanicum appears as a soft tissue mass abutting the promontory of the middle ear [3]. There may be displacement of ossicles or bony erosion of the tympanic cavity. The finding of air or bone between the tumor and the jugular bulb virtually assures the diagnosis of a tympanicum. CT scans are best for evaluating bony destruction and erosion, which is a hallmark of jugulo-tympanic glomus tumors. MRI is usually better than CT for delineating tumor edges and intracranial extent. It is also better for evaluating the relationship of the tumor to adjacent jugular vein, carotid artery, membranous labyrinth and cranial nerves [4].

Angiography recognizes the primary feeding blood supply to the lesion, helps in detecting multicentric tumors, identifies intrasinus and intravenous extension, provides further information on flow in contralateral sigmoid and internal jugular vein and allows for possible pre-operative embolization. Pre-operative embolization of feeding vessels is useful, if performed within 48 hours of planned surgery thereby greatly reducing the blood supply to the tumor, diminishing the need for transfusion, allowing the procedure to be performed with greater safety.

4. Conclusion

Glomus tissue in temporal bone is a vascular tissue in the dome of the jugular bulb and the promontory of the middle ear [5].With reference to our case, it is important to tailor the management of glomus tympanicum based on the patient's co-morbidity. The treating physician should also be aware of such a diagnosis, and probing should not be attempted as it could cause a disastrous bleeding event. Physicians should also be open to other modalities of treatment, including use of the gamma knife and radiotherapy, which has shown promising results. It is important to consider the proper approach to and planning of treatment for this disease.Hence, in any patient presenting with tinnitus, reduced hearing, and vertigo, an otoscopic examination is mandatory during a clinical office examination by all treating physicians.CT and MRI scans are the primary imaging modalities used in the evaluation and management of glomus tympanicum paraganglioma[6].

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