CASE REPORT

Glomus jugulare: high index of clinical suspicion is important for optimal management

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SUMMARY

Glomus tumour is a type of extra-adrenal chemodectoma or paraganglioma, originating within the wall of jugular bulb. We report a case of a 60-year-old woman who presented with hearing loss and pulsatile tinnitus. High index of suspicion and appropriate use of imaging led to early diagnosis. However, the patient did not give consent for surgery and was managed satisfactorily with radiotherapy.

BACKGROUND

Glomus jugulare tumours are rare, slow-growing, hypervascular tumours that often go unnoticed because of the insidious onset of symptoms.^{1 2} They originate from the paraganglia called as the glomus bodies, located within the wall of the jugular bulb.³ These tumours present a significant challenge owing to their location and large size at presentation.⁴ The common presenting symptoms are tinnitus, decreased hearing, ear pain and vertigo.⁵ In some cases extensive tumours may also involve various cranial nerves.⁶

CASE PRESENTATION

A 60-year-old woman presented with symptoms of hearing loss and pulsatile ringing sound in the right ear for the last 4 years. The patient also had difficulty in swallowing for the last 9 months. No history of dizziness or pain in ear was present. On examination, she was of average build, height and well oriented to time, place and person. On protrusion of the tongue, it deviated to right side suggesting lingual weakness. There was also right palatal paresis and absent gag reflex on right posterior pharyngeal wall, which may

account for patient's symptoms relating to regurgitation of liquids from the nose. The patient's vitals were within normal limits. On otoscopic examination, a pulsatile reddish mass was seen behind the intact pars tensa, which blanched and stopped pulsating on applying pressure with Siegel's pneumatic speculum. Tuning fork tests and audiometric examination showed hearing loss of mixed nature.

INVESTIGATIONS

MRI examination was performed, which showed evidence of an expansile, well-defined mass lesion in the region of right jugular fossa of approximate size 3.0×2.5 cm (figure 1). The lesion eroded petrous part of temporal bone and extended into the ipsilateral middle ear cavity, mastoid air cells eustachian tube and petrous part of the carotid canal. However, there was no evidence of intracranial extension. Bony window of non-contrast enhanced CT showed similar findings (figure 2). The lesion showed variable signal intensity of different MRI sequence. On postcontrast fat suppressed T1-weighted sequence, the lesion demonstrated avid contrast enhancement with mixed areas of flow voids consistent with typical 'salt and pepper appearance' of paragangliomas. Twenty-four hours urine vanillylmandelic acid level was within normal range.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses considered were glomus jugulare, jugular Schwannoma, neurofibroma, meningioma, metastasis and primitive neuroectodermal tumour.

Figure 1 (A and B): Contrast enhanced axial T1-weighted MR scans at the level of external acoustic meatus showing an enhancing mass in right jugular fossa with erosion and destruction of petrous part of temporal bone and extending to middle ear, Eustachian tube and right petrous carotid canal with involvement of the mastoid air cells. Note the typical 'salt and pepper appearance' of the lesion.

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Figure 2 (A and B) Axial CT scans showing bone window images at the same level of external acoustic meatus as in MR images, demonstrating erosion and destruction of petrous part of temporal bone with collection in the middle ear and mastoid air cells.

OUTCOME AND FOLLOW-UP

The patient was diagnosed as a case of glomus jugulare and was advised surgery. But she did not give consent for surgery and consequently radiation therapy was given. The patient responded to the treatment and remains stable during 6 months follow-up.

DISCUSSION

Glomus tumours are benign, slow-growing tumours that arise from neuroectodermal tissues. In the head and neck, two anatomic groups of glomus tumour can be divided into two main categories cervical 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 jugulare, and are the most common neoplasms of the middle ear.⁷ Microscopically, they consist of clusters of type I or catecholamine containing chief cells ('zellballen') and type II or sustentacular cells (modified Schwann cells), intimately interlaced with a rich network of capillaries and venules.⁸ Histology shows nests of ovoid cells having ovoid to round basophilic nuclei and moderately eosinophilic cytoplasm. There are network of thin-walled capillaries within the surrounding fibrous stroma.⁹

Glomus jugulare tumour is traditionally considered as surgically inaccessible tumour. Because of recent advancement in skull base surgery in terms of surgical techniques and instrumentations, it is now possible to safely remove these tumours without significant morbidity and mortality. However, they still pose a significant surgical challenge owing to slow growth and resultant large size at presentation.¹⁰ Although rare with an estimated annual incidence of one case per 1.3 million people, glomus tumours are common tumours of the middle ear cavity and the temporal bone. They occur commonly in adults and elderly population with significantly higher incidence in women. A left-sided preponderance is also reported. Multiple tumours are more common in familial cases.^{11–13}

As glomus jugulare tumour originates in the region of the jugular bulb, it sometimes extends into the posterior fossa with involvement of the lower cranial nerves. Despite extensive improvement in surgical and radiation therapy management of this tumour, what constitutes an optimal treatment, is still a matter of debate. Most authors agree that high index of suspicion for early diagnosis along with early appropriate treatment of this tumour can reduce the morbidity and mortality in majority of cases.

Learning points

- Glomus jugulare tumour is one of the causes of pulsatile tinnitus.
- It is frequently associated with multiple cranial nerve involvement.
- MRI is very helpful in diagnosis and defining the exact extent of tumour.
- Early diagnosis is based on recognition of clinical symptoms and maintaining a high index of suspicion for this disease.

Contributors IMM and AMM were involved in patient evaluation, treatment and follow-up. MA and AS were involved in the investigation and follow-up of the patient. All authors were involved in the preparation and approval of the manuscript.

Competing interests None.

Patient consent Obtained.

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