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Case Report

Altered clinical course of glomus tympanicum - a case report

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Abstract Glomus tumours of temporal bone are rare and usually present with symptoms of hearing loss and tinnitus. Diagnosis is often delayed due to the slow growth of the tumour. Here we present a case report of a patient diagnosed as glomus tympanicum who presented only with unilateral progressive hearing loss for the past one year and rapidly detoriating hearing loss since two months who was managed successfully.

Keyword Glomus tympanicum · Hearing loss · Magnetic Resonance Imaging (MRI)

Introduction

Glomus tumours of temporal bone are rare and glomus jugulare is the most common type encountered clinically with symptoms of hearing loss and pulsatile tinnitus. We profile a patient diagnosed as glomus tympanicum who presented only with unilateral progressive hearing loss of short duration which is a variation of the normal presentation of glomus tumours as they usually present with tinnitus, hearing loss and may have a long natural history.

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Case report

A 40-year-old lady presented with history of unilateral progressive hearing loss for the past one year and rapidly deteriorating hearing loss since two months. She had history of giddiness and headache. Otoscopic examination revealed reddish mass occupying the postero-superior and partly postero-inferior quadrant. There was a rim of normal tympanic membrane near the floor. Cranial nerve functions were normal.

Pure tone audiogram revealed 30db conductive hearing loss on right ear with normal hearing on the left. MRI of temporal bone including neck showed circumscribed enhancing space occupying lesion in medial aspect of the right middle ear along the cochlear promontory suggestive of glomus tympanicum without any evidence of extension into jugular fossa and carotid canal (Fig. 1). A week later the patient underwent Right posterior tympanotomy, atticotomy and complete excision of the mass in toto with Type III tympanoplasty under general anaesthesia. The polypoidal mass was extending from the Eustachian tube orifice to the mastoid antrum through the mesotympanum proper including the ossicular chain except the footplate of stapes. Defect in the posterior canal wall was reconstructed with autograft (conchal cartilage). Histopathological examination of the mass revealed features consistent with that of glomus tympanicum (Paraganglioma Fig. 2). Postoperatively patient made a good recovery without any complications. At her latest review (3 months post-operative) her symptoms improved and she had only a mild ipsilateral conductive hearing loss (15db) with minimal affection of speech frequencies.

Discussion

The glomus jugulare is a collection of paraganglionic cells, derived from the neural crest, which are found in close association with the jugular bulb. Paraganglionic cells are

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found widely distributed within the autonomic system, and are divided into two groups on the basis of their endocrine activity. The paraganglia of the adrenal medulla secrete adrenaline and noradrenaline, and on histological examination stain chromaffin positive. Non-physiologically active paraganglia have a negative chromaffin reaction, and the glomus jugulare falls into this group. It is from this group of cells that glomus tumours arise. Thus, only a very small proportion (two percent [5]) of glomus tumours have any endocrine activity, which can be diagnosed by measurement of 24-hour urinary vanillylmandelic acid (VMA) levels.

The histological appearance of glomus jugulare tumours is similar to that of normal glomus jugulare tissue, with only rare mitotic figures and a well-defined capsule. Malignant tranformation and metastatic spread are rare, but these tumours are locally destructive and invasive of surrounding bone and neural tissue. The majority of glomus tumours arise sporadically, but there are literature reports of familial glomus tumours, with an autosomal dominant pattern of inheritance and variable penetrance [3].

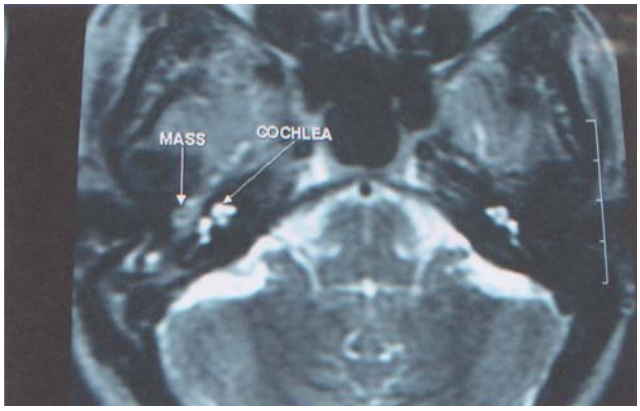

Fig. 1 MRI of temporal bone showing circumscribed enhancing space occupying lesion in medial aspect of right middle ear along the cochlear promontory suggestive of glomus tympanicum without any evidence of extension into jugular fossa and carotid canal

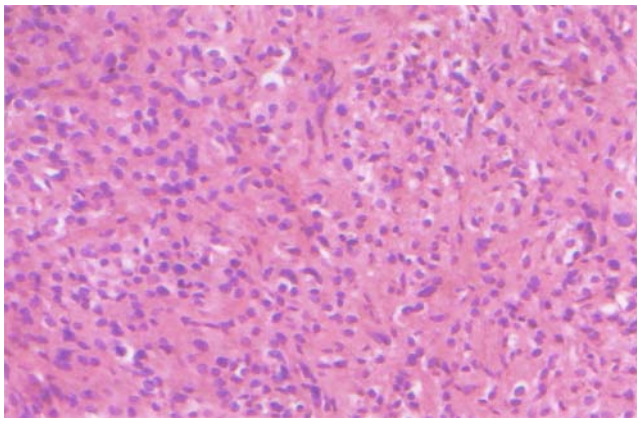

Fig. 2 Photomicrograph of glomus tympanicum showing cells arranged in nests separated by delicate fibrovascular septa x400 (H&E stain)

There are several different classification systems for glomus jugulare tumours;the most widely used is that by Oldring and Fisch which is based on tumour site(Table 1) [2]. Although there have been subsequent modifications to this system, the original classification as reproduced here is still the most useful.

Classic presenting features of glomus jugulare tumours include hearing loss (69 percent) and pulsatile tinnitus (55 percent). Examination may reveal a middle-ear mass, the so called 'rising sun sign' (75 percent) [2]. Cranial nerve deficits most commonly affect the vagus (16 percent) [1], glossopharyngeal (16 percent) [1], facial nerves (8 percent) and headache (15 percent). Glomus tumours are usually slow growing tumours and may have a long natural history. They usually present in the middle age groups. But this particular patient presented with unilateral progressive hearing loss of short duration which is a varied presentation of glomus tumours, without the classical diagnostic clinical signs.

Conclusion

The diagnosis of glomus tumours is frequently delayed as a result of the characteristically slow growth of these tumours. The classical features of a middle ear mass, pulsatile tinnitus and loss of hearing occur in the majority of patients. Here it is noteworthy to mention that this particular patient did not have the classical signs of glomus tumour. Also, it should be borne in mind that all patients presenting with unilateral progressive hearing loss should be investigated thoroughly for identifying any rare pathology in the middle ear or retrocochlear tract. Hence a careful clinical assessment of the ear should be carried out meticulously in all cases so that diagnosis is clinched and intervention at the earliest leads to a good prognosis.

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