Simultaneous Presentation of Contralateral Vestibular Schwannoma and Glomus Jugulare Tumour

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Introduction

Vestibular schwannomas arise from Schwann cells on branches of the vestibulocochlear nerve, most commonly the superior vestibular nerve, and have an annual incidence of 1 in 100 000\(^1\). Glomus jugulare tumours arise from glomus bodies or paraganglionic tissue in the adventitia of the jugular bulb, usually in relation to auricular branches of the vagus and glossopharyngeal nerves, and have an annual incidence of 1 in 1.3 million\(^2\). Simultaneous presentation of vestibular schwannoma and glomus jugulare tumour has been reported in only three cases in the literature and in all three of these cases the tumours presented on the same side (Table 1). We report this unique case of vestibular schwannoma and glomus jugulare tumour presenting simultaneously on opposite sides.

Case Report

A 66 year old female presented with a 6 year history of right sided hearing loss and a 2 year history of left sided pulsatile tinnitus. Other symptoms included unsteadiness and left sided headaches. Past medical history included right sided chronic suppurative otitis media. Otoscopy revealed a central perforation of the right tympanic membrane and a red pulsatile mass behind the left tympanic membrane. Neurological examination demonstrated normal cranial nerves and cerebellar function. Romberg’s test was negative and Unterberger’s stepping test was normal but the patient was unsteady on tandem gait with the eyes open.
Pure tone audiometry showed a mixed hearing loss in both ears with air conduction thresholds averaging 82dB in the right ear and 44dB in the left ear (Figure 1). Speech audiometry showed a maximum discrimination score of 0% at 80dB in the right ear (representing socially useless hearing) and 100% at 80dB in the left ear. An MRI scan revealed a 15mm maximum medio-lateral diameter vestibular schwannoma on the right (Figure 1) and a Fisch type D glomus jugulare tumour on the left (Figure 2). Carotid angiography demonstrated the appearances typical of a large glomus tumour supplied mainly by branches of the external carotid artery (Figure 3).

The management planned was embolisation followed by radiotherapy for the left-sided glomus jugulare tumour and observation of the right-sided vestibular schwannoma.

Discussion

Simultaneous presentation of a vestibular schwannoma and a glomus jugulare tumour has been reported in only three cases in the literature (Table 1). All of these patients were females and were aged between 65 and 79. In all three cases the tumours presented on the same side. We report a case of simultaneous presentation of a vestibular schwannoma and a glomus jugulare tumour in a 66 year old female. Our case is unique in the literature as the tumours presented on opposite sides.

All three previous cases presented with symptoms in one ear only. A vestibular schwannoma was diagnosed incidentally on radiological investigation for a glomus
jugulare tumour in two cases \(^3,4\) and a glomus jugulare tumour was diagnosed incidentally at operation for a vestibular schwannoma in one case\(^5\). In contrast, our case presented with symptoms in both ears and the vestibular schwannoma and glomus jugulare tumour were diagnosed simultaneously on radiological investigation.

Management options for both glomus jugulare tumours and vestibular schwannomas include observation, surgery and radiotherapy. In the three previous cases of simultaneous presentation of a vestibular schwannoma and a glomus jugulare tumour the management was more straightforward as the tumours were ipsilateral. All three cases were managed differently - with surgery in one case\(^5\), radiotherapy in one case\(^4\) and observation in one case\(^3\).

In contrast, in our case the management was less straightforward as the tumours were contralateral, with a Fisch type D glomus jugulare tumour in a better hearing left ear and a 15mm diameter vestibular schwannoma in a right ear with no useful hearing. The main priority was to manage the disease in both ears whilst preserving the hearing in the better hearing ear. In view of its size, the glomus jugulare tumour clearly required treatment whereas for the relatively small vestibular schwannoma a watch, wait and rescan policy was adopted after careful consideration. It was decided that embolisation followed by radiotherapy was preferable to surgery as this would be likely to control the disease without the need for major surgery and probably with a lower risk to the hearing.
References


Legends

Figure 1
Axial MRI scan showing right-sided vestibular schwannoma.

Figure 2
Axial CT scan showing left-sided glomus jugulare tumour.
Figure 3

Carotid angiogram showing left-sided glomus jugulare tumour.

Table 1

Previous cases.
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<th>Case</th>
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Table 1.