



Case Report

Simultaneous Contralateral Vestibular Schwannoma and Middle Ear Paraganglioma Tumor

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To the best of our knowledge, only 2 cases of a simultaneous contralateral vestibular schwannoma (VS) and middle ear paraganglioma (MEP) have previously been reported in literature. We report the third case observed in a 43-year-old male, who presented with an 11-year history of right-sided hearing loss and a 1-year history of left-sided pulsatile tinnitus. A magnetic resonance imaging (MRI) showed a VS on the right side and computer tomography (CT) identified a Fisch type A1 paraganglioma on the left side. The VS was treated using a translabyrinthine approach and the MEP was kept under radiological observation for 1 year. Due to the growth of the MEP (Fisch type A2), it was treated with excision via a retroauricular approach. Our case was very challenging because there was a different and important pathology on each side, both carrying a risk of deafness as a consequence of the disease and/or the treatments.

KEYWORDS: Vestibular schwannoma, paraganglioma, translabyrinthine, retroauricular approach

INTRODUCTION

Vestibular schwannomas (VS) arise from the Schwann cells on the branches of the vestibulocochlear nerve, most commonly the superior vestibular nerve. The incidence is 2 in 10,000^[1]. VS account for approximately 6% of all intracranial tumors^[2]. Paragangliomas arise from glomus bodies or paraganglionic tissue in the adventitia of the jugular bulb, usually in relation to the auricular branches of the vagus and glossopharyngeal nerves. MEPs are rare tumors, representing less than 0.5% of all head and neck tumors. Approximately, 3% of them occur in the head and neck area; other estimates suggest that they are 1 in 30,000 head and neck tumors^[3]. To the best of our knowledge, only 5 cases of simultaneous VS and MEP have been previously reported in literature. In the first 3 cases the lesions were located on the same side, but in the fourth and fifth they were located contralaterally (Table 1). Here, we report a new case of simultaneous contralateral VS and MEP and present a review of the literature.

CASE PRESENTATION

A 43-year-old male presented with an 11-year history of right-sided hearing loss and dizziness and a 1-year history of left-sided pulsatile tinnitus. Speech audiometry showed a maximum discrimination score on the right side of 30% at 80 dB (Figure 1). Otoscopy revealed the presence of a red pulsatile mass behind the left tympanic membrane (Figure 2a). Pure-tone audiometry revealed sensorineural, down-sloping hearing loss on the right side and low-tone conductive hearing loss on the left side. The patient underwent an axial high-resolution computed tomography (HRCT), which showed a 4.9 mm round lesion of the same density as soft tissues adherent to the promontory wall of the left ear (Figure 2b). Turbo spin echo magnetic resonance imaging (MRI) detected a hyper-intense nucleus at the base of the basilar turn of the cochlea (Figure 2c) of the left ear on T2 in the coronal image. A T1-weighted three-dimensional gradient-echo technique with gadolinium injection showed a marked increase of the signal at the level of the lesion in the hypotympanum in contact with the promontory wall. When analyzing the lesion using a dynamic sequence turbo fast field echo, an enhancement in the arterial phase was found during gadolinium injection, permitting the identification of the lesion as a 4.9 mm type A1 MEP of the left temporal bone (Figure 2d-f). T1 images with gadolinium also showed a 17 mm VS on the right temporal bone, which did not reach the fundus of the internal auditory canal (Figure 3a). The right VS was removed with a translabyrinthine approach that resulted in total hearing loss on the same side. For this reason, it was temporarily decided not to treat the MEP surgically, but to observe it with planned radiological examinations. After 1 year, an exacerbation of pulsatile tinnitus was reported by the patient and both HRCT and MRI were repeated. The images showed that the MEP had increased in size from 4.9 mm to 6.2 mm (Figure 3b, c), thereby becoming

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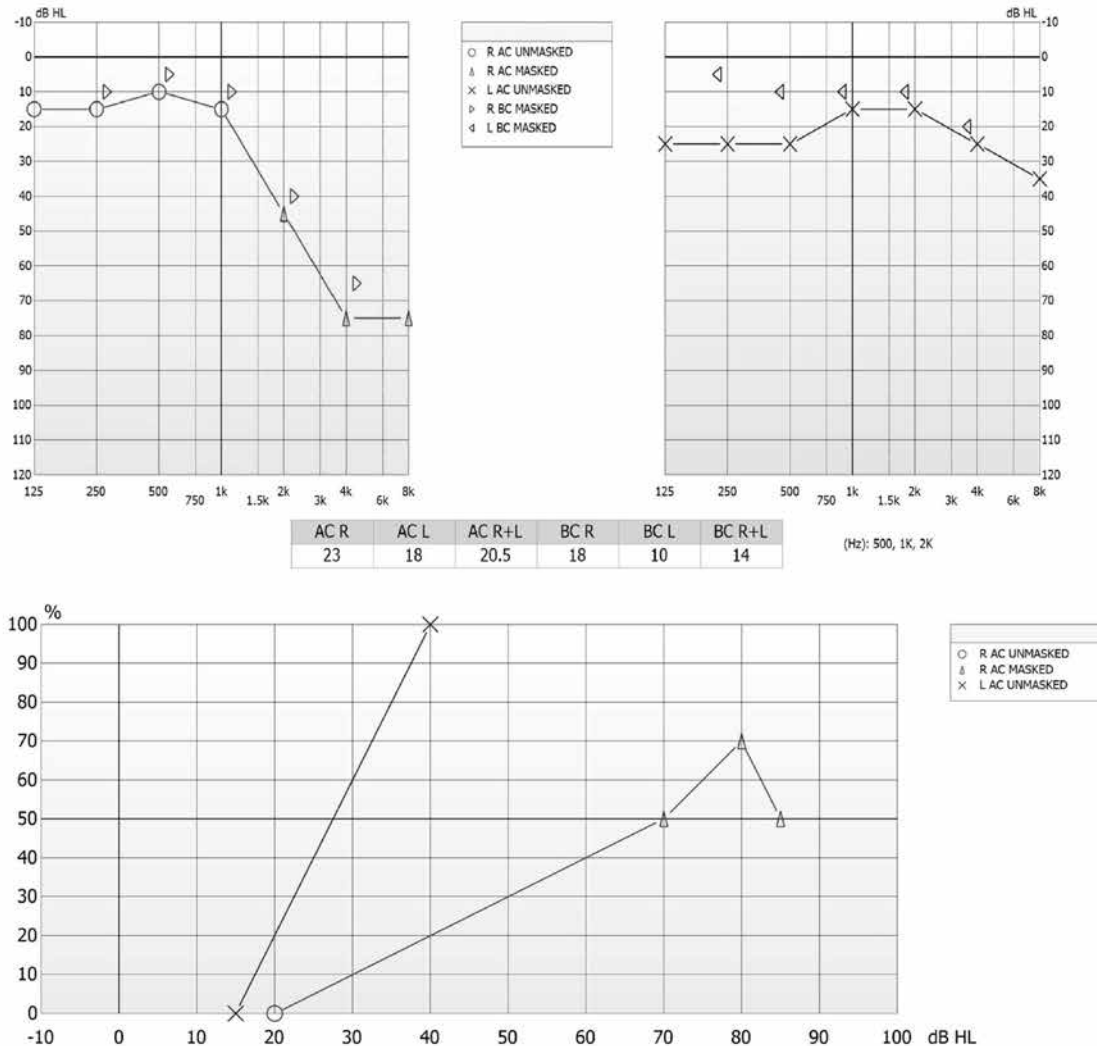
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Table 1. Characteristics of the 3 cases of simultaneous contralateral VS and Paranglioma

Author	Age/Sex	VS	GJT	Management
Moumoulidis et al. ^[5] 2005	66/Female	Right 15 mm	Left Fisch type D	Schwannoma observation Glomus tumor embolization radiotherapy
Ceylan et al. ^[6] 2007	63/Female	Right 22 mm	Left Fisch type C2	Schwannoma gamma knife stereotactic Glomus tumor embolization radiotherapy
The present study	41/Male	Right 17 mm	Left Fisch type A2	Schwannoma surgery Glomus tumor surgery

VS: vestibular schwannoma

**Figure 1.** Pre-operative pure-tone audiometry (right ear: down-sloping curve; left side: conductive hearing loss on the low frequencies) and speech audiometry showed a maximum discrimination score of 70% at 80 dB

a type A2 paraganglioma ^[4]. Because of the worsening of symptoms (tinnitus) and more importantly, because of tumor growth, after an appropriate and thorough counselling with the patient, the MEP was surgically removed via a retroauricular, laser-assisted approach. Both the preoperative diagnoses were confirmed by pathologic examination of the specimens. One month after surgery, the patient reported disappearance of pulsatile tinnitus with preservation of the preoperative auditory threshold. All the described diagnostic and therapeutical procedures were performed after obtaining a signed informed consent by the patient.

DISCUSSION

To our knowledge, only 5 cases of simultaneous VS and MEP have been reported in literature in the same patient: 3 cases being localized on the same temporal bone and the other 2 cases in the contralateral ear. As in the present case, the therapeutic approach for such a situation could be controversial, considering many treatment possibilities for both lesions, including observation, radiotherapy, and surgery. Moumoulidis et al. ^[5] described the first case of simultaneous VS and MEP affecting both sides and proposed a wait-and-scan

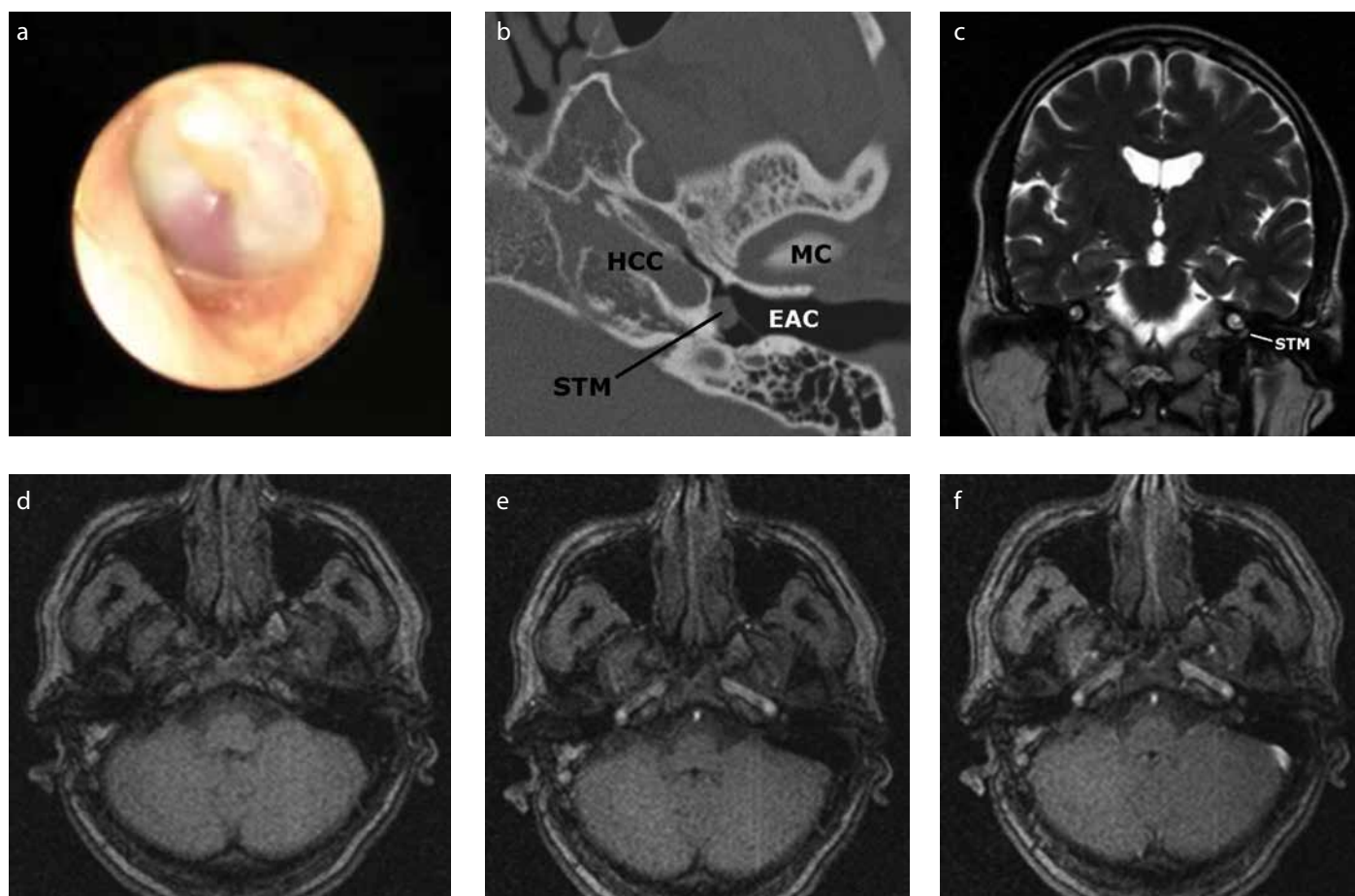


Figure 2. a-f. (a) Otoscopy on the left tympanic membrane revealed the presence of a red pulsatile mass behind, (b) axial section HRCT scan of the GJT located anteriorly in the middle ear cavity; the bone of the carotid canal is intact over the carotid artery, (c) turbo spin echo T2 on the coronal image shows a hyperintense nucleus inferiorly at the base of the basilar turn of the cochlea, (d) MRI dynamic sequence turbo fast field echo during gadolinium injection; image: the contrast has not yet arrived, (e) MRI dynamic sequence turbo fast field echo during gadolinium injection; the image is in the arterial phase with precocious lesion enhancement, (f) MRI dynamic sequence turbo fast field echo during gadolinium injection; the image is in venous phase showing the venous sinus
HRCT: high-resolution computed tomography; MRI: magnetic resonance imaging; GJT: glomus jugular tumor

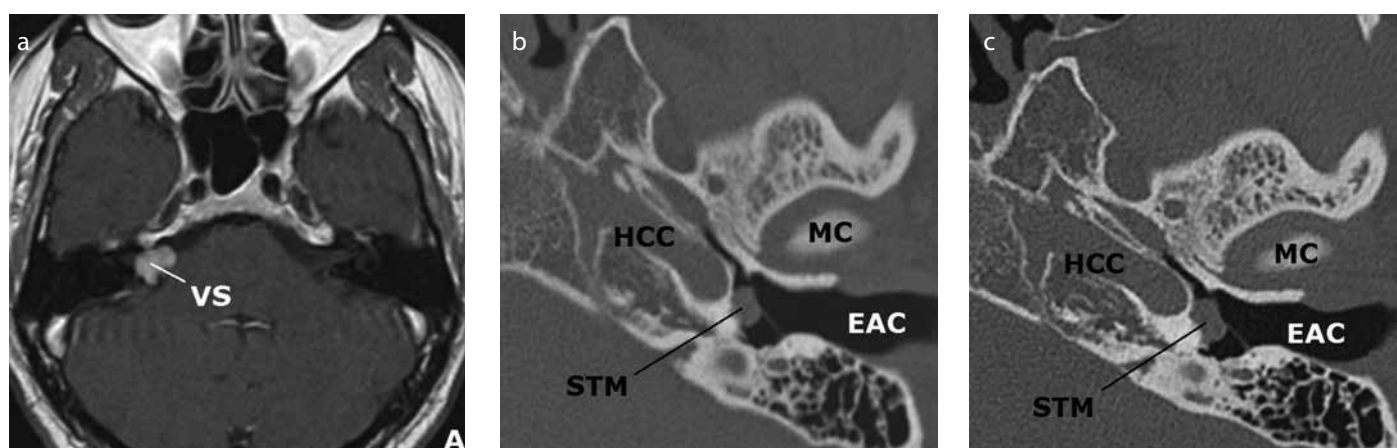


Figure 3. a-c. (a) Axial MRI shows right-sided VS, (b) coronal section HRCT scan at the moment of diagnosis, (c) coronal section HRCT scan follow-up after 2 years
HRCT: high-resolution computed tomography; MRI: magnetic resonance imaging

policy for hearing preservation purposes also due to the limited size of the VS, while the larger paraganglioma was treated with embolization and radiotherapy. The authors chose these approaches instead of surgery for a safer hearing control [5]. Ceylan et al. [6] reported on a 63-year-old woman presenting with a 22 mm VS on the right side,

which they decided to treat with gamma knife stereotactic radiosurgery, while the left-sided type D paraganglioma was treated with embolization and radiotherapy (Table 1). These two different approaches were therefore motivated by the relatively large dimensions of both lesions and by the intention of hearing preservation.

In the present report, both lesions were relatively small initially. The young age of the patient was the main motivation for the choice to first treat the VS surgically via a translabyrinthine approach due to the poor speech discrimination in the affected ear. Due to the resulting total hearing loss, it was at first decided not to treat the GJT on the left side, but to wait and scan it regularly. Due to the worsening of symptoms (pulsatile tinnitus) and to lesion growth, surgery was instead planned 2 years later to prevent possible complications in the event of additional growth. The decision to perform surgery on GJT was consented by the patient, explaining the possible postoperative hearing issues. Surgery was performed through a retroauricular approach, with complete lesion removal and no effects on residual hearing.

One month after surgery, the patient reported the disappearance of pulsatile tinnitus without deterioration of hearing in the left ear. If surgical removal of the glomus had led to a worsening auditory threshold of the left ear, a conventional or semi-implantable hearing aid would have been recommended.

CONCLUSION

The concomitant presence of tumoral lesions affecting both temporal bones deserves caution, as far as their treatment is concerned, due to the likelihood of bilateral hearing damage. The decision of a surgical removal should thoroughly be discussed with the patient in accordance with the clinical behaviour of these lesions.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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