

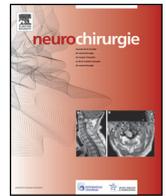


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Letter to the editor

Trigeminal neuralgia in vestibular schwannoma: Atypical presentation and neuroanatomical correlations



1. Introduction

Vestibular schwannomas (VS) are benign tumors of the myelin-forming Schwann cells of the vestibular branch of the vestibulocochlear nerve. In the vast majority of cases, the most common to least common symptoms at diagnosis are unilateral hearing loss, tinnitus, paresthesia of the posterior ear canal (i.e. Hitselberger's sign), headache, vertigo, hemifacial spasm and weakness [1]. In addition, some patients may experience facial hypoesthesia or pain, yet these are rarely encountered [1]. Importantly, the presenting symptoms have not been correlated with tumor extension [2]; nevertheless, some association has been suggested with tumor size [3].

2. Clinical Case

A 63-year-old female patient presented in our outpatient department for a regular annual evaluation of her acoustic neuroma. She complained of progressively worsening hearing loss and gait imbalance, as well as recent onset of intermittent shock-like pain distributed along the course of the ophthalmic branch of the left trigeminal nerve. In addition, the mass was found to have increased in size – tumor equivalent diameter:

$$TED = \sqrt[3]{D1 \times D2 \times D3} \approx 1.87\text{cm}$$

where D is the maximum diameter in each of the three planes. Consequently, surgical treatment was suggested.

Upon admission, she had hemianalgesia of the left side of her face, dysidiadochokinesia of the left upper limb and dysmetria (undershooting) bilaterally, as well as a positive left Babinski's sign and Romberg's sign (sway to the left side). Her preoperative workup was unremarkable except for her audiogram being suggestive of declining auditory acuity. Magnetic resonance imaging (MRI) scan of the brain with intravenously administered contrast agent revealed the presence of a mass with inhomogeneous enhancement originating from the left vestibulocochlear nerve (Fig. 1).

The patient underwent elective retrosigmoid craniotomy and the extrameatal portion of the mass was excised. Macroscopically, the tumor was round, lobulated and greyish in color (Fig. 1f), arising from the vestibulocochlear nerve. The postoperative course was uneventful. No change in her neurological condition was noted at that point. At a routine re-evaluation 6 months postoperatively, she had complete remission of the cerebellar symptomatology, while the severity and frequency of the intermittent shock-like pain had decreased. No change was found during the sensory examination of the face.

3. Comments

Here, we report the case of a vestibular schwannoma with an atypical neurological presentation.

The incidence of symptoms secondary to trigeminal or cerebellar compression appears to increase with increased tumor grade and size [3], but does not always reach statistical significance [4]. On the other hand, some authors argue the presenting symptoms are not exclusively dependent on the diameter of the mass [5], but rather consider the final neurological manifestation to be multifactorial.

Some of the factors suggested are the variable morphology of the posterior fossa among individuals, the direction of tumor growth and the extension of brainstem compression [5]. Indeed, in our patient, while the mass was classified as medium in size [6], it appears to greatly compress the brain stem and cerebellum (Fig. 1c), whereas it leaves adjacent structures unaffected, most importantly the trigeminal nerve (Fig. 1d). Another possible pathogenetic mechanism is direct or indirect compression of a nerve [7]. Here, neither macroscopic infiltration nor compression of the trigeminal nerve or adjacent vessels was identified radiologically (Fig. 1d, e) or intraoperatively.

As none of the above could explain the symptomatology in our patient, we speculate that a different mechanism was at play, which involves insults to the spinal trigeminal nucleus. This nucleus, which extends up to the main sensory nucleus in the pons, and particularly its caudal part, has been found to be responsible for the conduction of pain and temperature from the ipsilateral half of the face [8], while structural changes at the cellular level have been identified in chronic cases of neuropathic pain [9]. Therefore, it is possible, given the position of the mass in our patient, that her symptomatology was secondary to direct compression of this nucleus. Similarly, cerebellar symptomatology (e.g. ataxia, dysmetria) can be explained by damage to the superior and middle cerebellar peduncles [10] found in the same region. Both conclusions are further supported by the observed remission of symptoms following tumor removal.

The clinical manifestation of vestibular schwannomas—and cerebellopontine angle tumors in general—may be the result of compression of deeper brainstem structures. This is especially the case for small to medium size tumors that exert their mass effect on the brainstem and could potentially explain why existing tumor grading systems do not reliably correlate with the neurological symptomatology.

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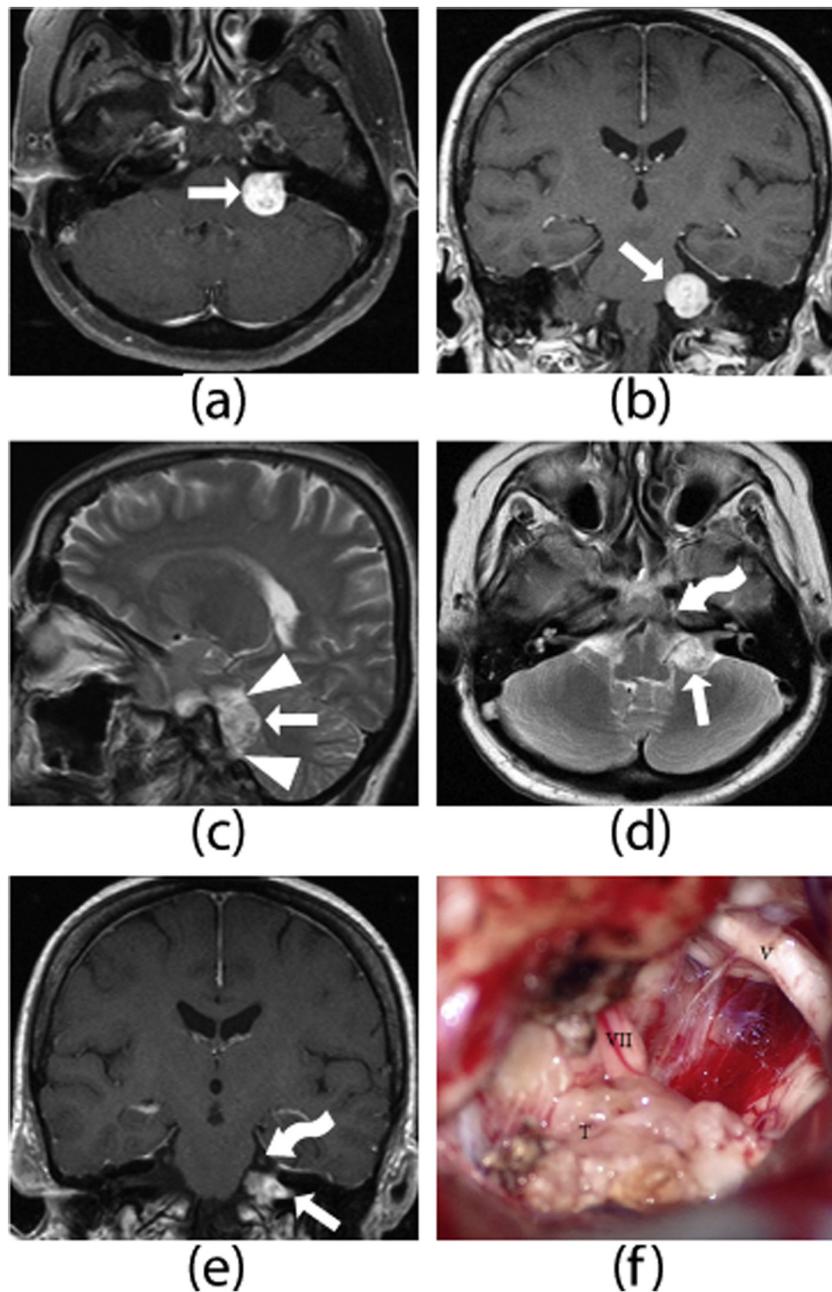


Fig. 1. T1W MRI with intravenously administered contrast agent shows a mass with inhomogeneous enhancement (arrow) originating from the left vestibulocochlear nerve, (a) axial view, (b) coronal view. The tumor exerted a mass effect (arrowhead) on the neighboring structures, as shown by the T2W images (c), however no direct compression of the trigeminal nerve was observed (image d T2W, image e T1W contrast enhanced, curved arrows). Intraoperative photograph showing the tumor (T) and its position relative to the facial (VII) and trigeminal (V) nerves (f).

Disclosure of interest

The authors declare that they have no competing interest.

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S. Apostolakis*
A. Karagianni
A. Mitropoulos
P. Filias
K. Vlachos

*Department of Neurosurgery, KAT General Hospital,
2, Nikis street, 145 61 Kifisia, Greece*

* Corresponding author.

E-mail address: sotapostolakis@gmail.com (S. Apostolakis)

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