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Small vestibular schwannoma presented with trigeminal neuralgia: illustrative case

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BACKGROUND A vestibular schwannoma (VS) presenting with paroxysmal facial electric shock pain, that is, trigeminal neuralgia (TN), is relatively rare. Furthermore, TN is extremely rare in small VSs.

OBSERVATIONS Herein, the authors report the case of a 52-year-old woman with a complaint of right TN. Magnetic resonance (MR) imaging revealed a right VS of 12-mm diameter that compressed the trigeminal nerve. Although she did not report any hearing impairment, audiometry revealed decreased high-frequency range on the right side. The tumor was excised using the right retrosigmoid approach, and TN was confirmed to be caused by direct compression of the trigeminal nerve by the VS. Sufficient decompression of trigeminal nerve was done. The proximity of the trigeminal nerve root to the vestibular nerve root was the cause of TN. TN disappeared immediately after surgery, and there was no worsening of hearing impairment and facial paralysis.

LESSONS It is important to remember that TN may occur with direct tumor compression, even in small VSs. A preoperative 3-dimensional MR cisternogram/angiogram fusion image clearly showed direct tumor compression of the trigeminal nerve and the absence of responsible vessels, which was useful for surgical planning.

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KEYWORDS vestibular schwannoma; trigeminal neuralgia; surgery; trigeminal nerve

Trigeminal neuralgia (TN) is a paroxysmal electric shock pain on one side of the face triggered by washing the face, eating, and speaking and is often caused by vascular compression of the trigeminal nerve.[1] TN can also be caused by compression of the trigeminal nerve by a tumor of the cerebellar pontine angle, but this is relatively rare.[2] Epidermoid is the most commonly found cerebellar pontine angle tumor causing TN, and vestibular schwannomas (VSs) are relatively uncommon,[2] occurring in only about 1.2%–3.3% of VSs.[3–5] There are 2 etiologies: compression of the TN by the tumor alone and a combination of compression by the tumor and by the blood vessels.[3,5,6] However, in either case, the onset of TN is considered as dependent on the large size of the VS.[3,5,6] There are no reports of TN due to direct compression by tumor in a small VS. In this report, we describe a case of VS as small as 12 mm in diameter, thereby showing severe TN, and discuss the causes.

Illustrative Case

History and Examination

A 52-year-old woman visited our department complaining of severe right facial pain and slight numbness. Five years earlier, she had been experiencing electric shock pain in the third branch of the right trigeminal nerve, which was triggered by washing her face or eating, whereas she began feeling slight numbness in the same area. The symptoms had worsened over the past 6 months, and she was referred to our department. At the time of the initial visit, there were no obvious

ABBREVIATIONS ABR = auditory brainstem response; MR = magnetic resonance; MRA = magnetic resonance angiogram; MRC = magnetic resonance cisternogram; MRI = magnetic resonance imaging; NIM = nerve integrity monitor; TN = trigeminal neuralgia; VS = vestibular schwannomas.

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complaints of hearing disturbance. Pregabalin administered by another family doctor had mild effects but was no longer effective at this time. No special notes were found in the patient's past medical history or family history.

Neuroimaging Findings

Magnetic resonance (MR) imaging (MRI) showed a solid tumor measuring 12 mm in diameter with a contrast effect in the right cerebellopontine angle (Fig. 1, left). Slight enlargement of the right internal auditory canal was observed. A 3-dimensional MR cisternogram/angiogram (3D-MRC/MRA) fusion image showed that the tumor was compressing the trigeminal nerve (Fig. 1, right). There were no obvious responsible vessels for the compression of the trigeminal nerve. The distance between the trigeminal nerve and the vestibulocochlear nerve was as short as 3.5 mm. A diagnosis of VS with direct compression of the trigeminal nerve was made. Surgical treatment was decided on the basis of the patient's wishes.

Neuro-Otological Findings

Audiometry showed a 50-dB reduction at high frequencies (data not shown).

Neurological Findings on Admission

Paroxysmal electric pain in the region of the third branch of the right trigeminal nerve was observed on facial contact. Objective neurological findings revealed no sensory abnormalities on the right face. The Weber test[7] was lateralized to the left ear, and the Rinne test[7] result was positive, thereby suggesting right sensorineural hearing disturbance. No facial nerve palsy was noted, and no tinnitus or dizziness was observed.

Surgical Findings

A right retrosigmoid approach was used with the patient under general anesthesia in the lateral recumbent position with continuous auditory brainstem response (ABR) and nerve integrity monitor (NIM) system monitoring. A liner skin incision was made in the right retroauricular region, and a small craniotomy was performed. The arachnoid membrane on the tumor surface was dissected; the presence of facial nerves was evaluated using NIM; the tumor capsule was incised, presumably without facial nerves; and internal decompression was applied. The root exit zone of the facial nerve could then be visualized, and the facial nerve was expected to run dorsal to the tumor. The trigeminal nerve could be seen dorsal and cranial to the tumor and was compressed by the tumor (Fig. 2). The area between the trigeminal nerve and the tumor was dissected, and the tumor was removed to ensure sufficient decompression to the trigeminal nerve. The external lateral wall of the internal auditory canal was drilled out, the internal auditory canal dura was incised, and the tumor in the internal auditory canal was removed as far as possible. The cochlear nerve was identified on the cranial surface of the tumor, and this part was untouched, including the surrounding tumor, to preserve hearing function. Facial nerve travels were evaluated using NIM and were found to run on the dorsal surface of the tumor. The tumor around the facial nerve was allowed to remain to preserve facial nerve function. No adhesions of the tumor to the brainstem were identified. After tumor removal, ABR showed the same finding as before surgery. In histopathological examination, the tumor was composed of tumor cells with cone-shaped nuclei. We confirmed a classic pattern of mixed distribution of Antoni type A dense tissue arranged in fascicular bundles and Antoni type B sparse tissue arranged in reticular bundles.[8] Consequently, schwannoma was diagnosed (World Health Organization grade I; data not shown).

Postoperative Course

TN and facial numbness disappeared immediately after surgery. No facial paralysis and preservation of the right-side effective hearing were confirmed. Postoperative MRI revealed the removal of the tumor and sufficient decompression of the trigeminal nerve (Fig. 3). The patient was discharged 8 days after surgery without the appearance of any new neurological deficits.

Discussion

Observations

Of 1,205 surgical cases of TN, 25 cases (2%) had tumors in the cerebellar pontine angle.[9] Of these, only 8 cases (0.7%) were VS.[9] In 1,000 surgical cases of VS, 3.3% had TN,[5] and another paper indicated that even in Koos grade IV[10] and large schwannomas, which present

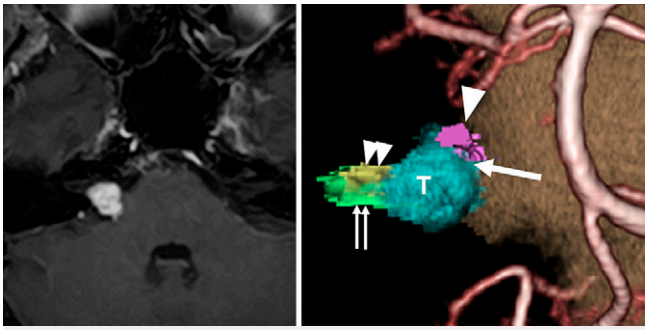

FIG. 1. Preoperative axial gadolinium T1-weighted MRI (**left**) showing a tumor 12 mm in diameter in the right cerebellopontine angle with contrast effect by gadolinium. A 3D-MRC/MRA fusion image (**right**) showing compression of the trigeminal nerve by the tumor. The distance between the trigeminal nerve and the vestibulocochlear nerve is as short as 3.5 mm. *Large arrowhead* indicates the trigeminal nerve; *long arrow*, the compression site; *double arrowheads*, the facial nerve; and *double arrows*, the vestibulocochlear nerve. T = tumor.

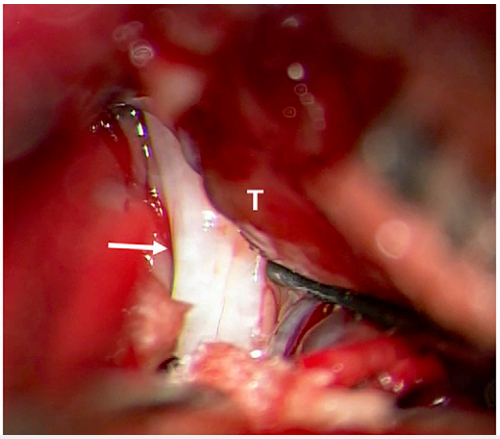

FIG. 2. After internal decompression of the tumor, the trigeminal nerve, which was compressed by the tumor (T), could be observed on the dorsal and cranial sides of the tumor. *Arrow* indicates the trigeminal nerve.

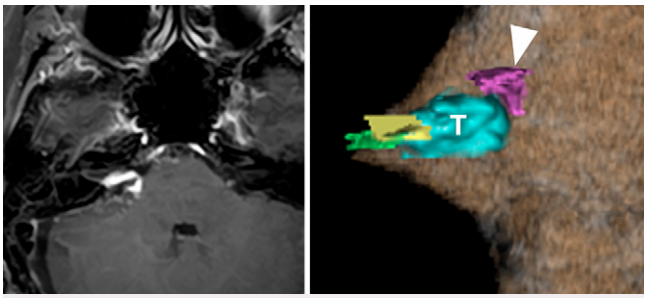

FIG. 3. Postoperative axial gadolinium T1-weighted MRI (**left**) showing partial residual tumor capsules. 3D-MRC/MRA fusion image (**right**) showing sufficient decompression of the trigeminal nerve. *Arrowhead* indicates trigeminal nerve. T = tumor.

deformity of the brainstem and shift of the fourth ventricle, only 1.2% of cases showed TN.[3] The onset of TN is due to compression by a large VS.[3–6] In particular, the trigeminal nerve is compressed when the tumor grows in an upward direction.[5] It is extremely rare for a VS as small as 12 mm in diameter, as in our case, to present with TN. Samii and Matthies[5] reported TN in 9 (0.9%) of 1,000 operated cases of VS, despite the small size of the tumor, which approximately corresponded to Koos grade II.[10] The surgical findings described in all 9 cases of small VS confirmed vascular compression of the trigeminal nerve without direct tumor compression.[5] The pathogenesis of TN in VS has been shown to involve direct compression of the nerve not only by the VS but also by the blood vessels.[3,5,6] Additionally, 75% of cases of large VS presenting with TN had vascular compression involvement.[6] When approaching surgery for VS presenting with TN, care should be taken not only to remove the tumor but also to identify the area surrounding the trigeminal nerve and to move the responsible vessels to ensure decompression of the trigeminal nerve.[3–6]

Here, preoperative 3D-MRC/MRA clearly confirmed compression by the tumor, whereas no vascular compression could be noted. The surgical findings were identical to preoperative 3D-MRC/MRA, thereby making this case a rare case of TN caused by direct compression of small VS. Even small-sized VS can present with TN, which, in our case, may have been related to the anatomical characteristic of the close distance between the trigeminal and vestibulocochlear nerve origins. The distance between the vestibulocochlear nerve and the trigeminal nerve was as short as 3.5 mm on MRI, thereby suggesting that the compression of the trigeminal nerve had occurred despite the tumor's small diameter. This case is the first demonstration that even a small VS can directly compress the trigeminal nerve and cause TN.

This case requires attention for diagnosis because the patient complained only of TN but not of hearing disturbance. Neuroimaging using MRI was necessary for an accurate diagnosis. Especially when the tumor is small, it is important to consider the existence of blood vessels compressing the trigeminal nerve. We have reported that 3D-MRC/MRA was useful in preoperative evaluation in surgical decision making and simulation.[11,12] The 3D-MRC/MRA is created by superimposing the MRA image on the structures in the brain chamber imaged by MRC to create a 3D anatomical structure around the trigeminal nerve. 3D-MRC/MRA can accurately identify the responsible vessel in TN cases and can also identify nerve deformities.[11,12] Here, preoperative 3D-MRC/MRA allowed us to confirm direct compression of the trigeminal nerve by the tumor and the absence of responsible vessels, which permitted precise surgical planning.

Lessons

We describe a case of TN caused by small VS. TN was provoked by direct compression by the tumor, and an anatomical characteristic of close distance between the trigeminal nerve root and vestibulocochlear nerve root was suspected to be involved. Preoperative 3D-MRC/MRA confirmed that the TN was induced by direct VS compression and not by vascular compression, which was very useful in planning the surgery.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Onoda, Ogasawara, Shimoji. Acquisition of data: Onoda, Ogasawara, Sashida, Fujiwara, Michiwaki. Analysis and interpretation of data: Onoda, Ogasawara, Fujiwara, Wakamiya, Yamane. Drafting the article: Ogasawara, Sashida, Fujiwara, Michiwaki. Critically revising the article: Ogasawara, Hirokawa, Michiwaki, Tanaka, Shimoji, Kawashima. Reviewed submitted version of manuscript: Ogasawara, Hirokawa, Michiwaki, Shimoji, Suehiro, Kawashima, Matsuno. Administrative/technical/material support: Onoda, Hirokawa, Shimoji, Yamane. Study supervision: Kawashima, Matsuno.

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