Chances of Improvement in Cases of Vestibular Schwannoma Presenting with Facial Nerve Weakness: Presentation of Two Cases and Literature Review

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Abstract

Background and Study Aim Facial nerve (FN) weakness as a presenting feature in vestibular schwannoma (VS) is extremely rare. We are presenting two different cases of VS with significant facial weakness and reviewed the literature for similar cases.

Methods and Results We are presenting two cases of VS with significant facial weakness. The first case was a 63-year-old male patient presented with 3 weeks' history of severe left-sided facial weakness (House–Brackmann [HB] grade V) and hearing loss. Magnetic resonance imaging (MRI) of the brain showed a tumor in the left internal auditory canal. Gross total removal with anatomical and physiological FN preservation was performed through a retrosigmoid approach under neurophysiological monitoring. FN function improved postoperatively to HB grade II after 16 months. The other case was 83-year-old male patient presented with sudden left-sided hearing loss and severe facial weakness (HB grade V). MRI of the brain 2.5 years before showed a left-sided (Class-T3A) cystic VS. The tumor was asymptomatic; wait-and-scan strategy was advised by the treating neurologist. Recent MRI of the brain showed approximately three times enlargement of the tumor with brain stem compression, extensive cystic changes, and suspected intratumoral hemorrhage. Surgery was performed; the tumor was subtotally removed through a retrosigmoid approach with intraoperative FN monitoring. The FN was anatomically preserved; however, physiological preservation was not possible. Severe facial weakness with incomplete lid closure persisted postoperatively.

Conclusion Surgical treatment could be offered to cases of suspected VS presenting with facial weakness, as these cases may still have a chance for improvement especially in laterally located tumors.

Keywords

- ▶ vestibular schwannoma
- ▶ facial nerve weakness
- ▶ cystic VS
- ▶ intrameatal
- ▶ intratumoral hemorrhage

Introduction

Vestibular schwannomas (VSs) are benign neoplasms that arise from Schwann cells at the neuroglial–neurilemmal junction (Obersteiner–Redlich's zone)¹ of either the inferior^{2,3} or superior vestibular nerve.⁴ They comprise 8 to 10% of all intracranial tumors and more than 80% of cerebellopon-

tine angle tumors.⁵ VS usually presents with tinnitus, hearing loss, and/or disequilibrium; however, large tumors may also present with facial hypoesthesia, facial nerve (FN) weakness, lower cranial nerve dysfunction, ataxia, or hydrocephalus.⁵ Incidences of postoperative FN weakness as well as FN preservation after VS treatment have been thoroughly discussed in the literature. However, facial weakness as a

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presenting feature in VS is rarely discussed. Only 4 to 8% of VS patients presents with facial weakness.^{6–8} The optimal surgical management of these cases is a challenge to the surgeons. We are presenting two different cases of VS with significant facial weakness, and we reviewed the literature for similar cases.

Case 1

A diabetic 63-year-old male patient presented with 3 weeks' history of severe left-sided facial weakness. Two years ago, the patient had a similar attack of facial weakness on the right side that improved after corticosteroid therapy. Neurological examination revealed in addition to severe left-sided facial palsy (House–Brackmann [HB] grade V), hearing loss on the left side. Cerebrospinal fluid examination showed no manifestation of inflammatory process. Magnetic resonance imaging (MRI) of the brain showed a tumor in the left internal auditory canal (IAC) with slight extrameatal extension and no extension into the fallopian canal (→Fig. 1). The lesion was

isointense in T1, slightly hyperintense in T2-weighted images, with homogenous enhancement after contrast injection. Because of the previous history of contralateral Bell's palsy, the patient was initially treated with corticosteroids. After failed improvement of the facial weakness under corticosteroids for 3 weeks, surgical removal of the tumor was offered to the patient. Gross total tumor removal was performed through a retrosigmoid approach with intraoperative neurophysiological monitoring. Intraoperatively, the FN could be stimulated with a good response from both orbicularis oculi and orbicularis oris following bipolar stimulation starting at 0.1 mAmp. The tumor was involving the vestibular division of the eighth cranial nerve and was extrinsic to the FN. Facial and cochlear nerves were anatomically preserved during surgery. At the end of surgery, FN stimulation at the exit zone from the brain stem was effective at 0.1 mAmp. The pathological examination revealed schwannoma of Antoni-A category with significant fibrotic and regressive changes. The last follow-up examination (after 16 months) showed improvement of the FN function to HB grade II, and cranial imaging using MRI of the brain

Fig. 1 Pre- and postoperative images of the first case: T1-weighted MR imaging before and after gadolinium shows left intrameatal tumor with slight extension into the cerebellopontine cistern but not reaching the brain stem (upper images). CT imaging depicts widening of the internal auditory canal (lower left). Postgadolinium T1-weighted MR imaging performed 16 months after surgery showing no tumor remnant or recurrence (lower right). CT, computed tomography; MR, magnetic resonance.

with and without contrast showed no evidence of tumor remnant or recurrence (►Fig. 1).

Case 2

A 83-year-old male patient presented with sudden left-sided hearing loss and facial weakness (HB grade V). MRI of the brain 2.5 years earlier showed a left-sided (Class-T3A) cystic VS. The tumor was asymptomatic; wait-and-scan strategy was advised by the treating neurologist. Recent MRI of the brain showed approximately three times enlargement of the tumor (Class-T4A) with brain stem compression, extensive cystic changes, and suspected intratumoral hemorrhage (►Fig. 2). The patient was taking antiplatelet drug (aspirin and clopidogrel) due to recent coronary stents. After holding aspirin and clopidogrel for 5 days, surgery was performed within 3 weeks of the onset of facial weakness; the tumor was subtotally removed through a retrosigmoid approach with intraoperative FN monitoring. The tumor was cystic with areas of old bleeding and the FN was extremely stretched anterosuperior to the tumor. To preserve

the integrity of the FN, a small part of the tumor was left at its entrance into the IAC. Intraoperative FN stimulation with up to 1 mAmp gave a very weak response so that functional preservation was not sure. Pathological examination revealed Antoni-A schwannoma with signs of old and fresh intratumoral bleeding as well as regressive changes. Severe facial weakness (HB grade V) with incomplete lid closure persisted postoperative, and tarsorrhaphy was performed to avoid corneal ulceration. MRI of the brain with contrast was performed 6 months postoperatively and showed tumor remnant at the IAC (►Fig. 2). The clinical examination after 6 months revealed severe facial weakness so that hypoglossofacial anastomosis was offered to the patient.

Discussion

Facial weakness is reported as a rare presentation in cases of VS, and usually occurs with large tumors especially those undergoing rapid expansion or cystic degeneration.⁹ Matthies and Samii reviewed the presenting symptoms of

Fig. 2 Pre- and postoperative images of the second case: T2-weighted MR imaging shows left cystic cerebellopontine angle tumor with brain stem compression (upper left). ADC image demonstrates foci of reduced signal indicated restricted diffusion due to intratumoral hemorrhage (one indicated by arrow in the upper right image). CT imaging confirmed the intratumoral hemorrhage (lower left). Postgadolinium T1-weighted MR imaging performed 6 months after surgery shows small tumor remnant (lower right). ADC, apparent diffusion coefficient; CT, computed tomography; MR, magnetic resonance.

1,000 VS and found facial weakness in 6% of the cases.⁷ Wiegand et al reviewed 62 cases of intracanalicular VS, of whom 2 (3.2%) had presented with facial weakness.¹⁰ Therefore, significant facial weakness in the setting of a cerebello-pontine tumor should raise the suspicion for a tumor rather than VS, like FN schwannoma,^{11,12} meningioma,^{9,13} or other less common pathologies as malignancy or cavernous hemangioma.^{14–16}

Important factors that influence the management of facial weakness include the duration and severity of weakness as well as the nature of the underlying pathology.¹⁷ Patients with FN schwannomas usually present with facial weakness either in the form of sudden paralysis or recurrent attacks of facial palsy with recovery in between the attacks.¹⁸ Extension of FN schwannomas beyond the fundus of the IAC and involvement of other segments of the intratemporal FN are diagnostic in most cases.^{11,19} There are previous reports of cases of FN schwannomas not extending into fallopian canal and associated with normal FN function, and these cases were suspected to be VSs and discovered to be FN schwannomas during surgery.^{20,21} In cases of FN schwannomas involving the IAC and associated with severe facial weakness, surgery may be considered and FN reanimation is usually included.²² The mechanism of FN weakness in cases of VS is usually due to direct compression¹⁷ rather than infiltrative growth as in cases of FN schwannoma;¹³ therefore, anatomical preservation of the FN could be possible during surgical removal of these tumors.

The other factor that could influence facial function recovery after surgery is the timing of intervention.^{23–26} Several reports showed that surgical treatment within 3 months of facial paralysis in cases of benign temporal bone lesions is associated with significant recovery of function.^{17,26} Our first patient was suspected to have Bell's palsy due to previous history of facial weakness on the contralateral side; however, the patient did not show any improvement under corticosteroids for 3 weeks. In this case, the tumor was intracanalicular with no involvement of other segments of the intratemporal FN, and total tumor removal was possible with anatomical and physiological FN preservation, and the facial function improved postoperative to HB grade II.

Reviewing the literature, we found five cases of intracanalicular VS presented with significant facial weakness. The authors described five patients presented with 1 to 3 months' history of moderate or severe facial weakness (HB grades III–V) in the setting of small intracanalicular VSs, the tumors were removed with significant postoperative improvement of facial function 3 months after surgery.^{6,17,27} Translabyrinthine approach was performed in three cases,^{6,17} whereas in two cases,²⁷ the authors used the retrosigmoid approach. The tumors were totally removed in four cases; however, in one case, near-total removal was performed. The authors reported anatomical preservation of the FN in all cases, interestingly intraoperative stimulation of the proximal part of the FN was possible in spite of significant facial weakness. The same in our first patient, tumor was totally removed and stimulation of the proximal part of the FN with 0.1 mAmp associated with good response, the facial function improved postoperative to HB grade II. We agree with other authors^{17,27} that anatomical

preservation of the FN should be the aim during removal of these tumors even at the expense of total tumor removal.

The mechanism of facial weakness in these cases is unclear, as previously mentioned direct compression of the nerve may be a possible explanation, edema and/or ischemia of the FN fibers may result from compression of the nerve against the borders of the internal auditory meatus.²⁷ Although the FN is frequently crushed against the borders of the internal auditory meatus in cases of VS, FN weakness is extremely rare; therefore, other possible mechanisms such as inflammation should be considered.

Our second case represents a different scenario; a case of cystic VS that showed progress in size with signs of intratumoral hemorrhage (ITH) and resulted in sudden facial weakness in an old patient who took antiplatelet drugs due to recent cardiac stents. Sudden deterioration of symptoms in a case of cystic VS is usually due to ITH.^{28,29} The average growth rate in VS is 2 to 2.5 mm/year.¹ This slow growth rate represents an acceptable explanation of normal facial function even in cases of large VSs. Niknafs et al reviewed the literature (from 1990 to 2014) for cases of VS associated with ITH; they reported 39 cases with average tumor size 3.11 ± 1.12 cm. The most common clinical presentation was unilateral hearing loss (75%), followed by headache (40.6%), facial weakness (31.3%), gait disturbance (28.1%), trigeminal nerve dysfunction (25.0%), and nausea (15.6%).⁸ Recent prospective studies suggested that ITH might be part of the natural history of VS.^{30–32} Sughrue et al analyzed prospectively the histopathological findings in 274 VS, almost all tumors showed evidence of hemorrhage, but in 23% of the cases, hemorrhage involved more than 25% of tumor volume.³⁰ Another prospective study found evidence of hemorrhage, based on T2*-weighted recalled echo MR imaging, in 15 out of 16 cases of VS.³¹ Large tumor size, abnormal thin-walled vasculature, mixed Antoni A and B cellularity on histopathology, as well as cystic tumors were reported in association with ITH in VS.^{5,33–35} In addition, the use of antiplatelet or anticoagulant drugs might predispose to bleeding into tumors, Carlson et al reported 25-fold increase in the frequency of ITH among patients receiving anticoagulation was 5.6% (2 of 36), compared with 0.2% (3 of 1,356) in nonanticoagulated patients.³⁶

Surgery for cystic VS represents a challenge due to extreme adhesions with the FN. Piccirillo et al observed unfavorable FN outcome in cases of medially located thin-walled cysts.³⁷ Xia et al studied the outcome predictors in cases of cystic VS, and they divided them according to the presence of fluid–fluid level into two groups. They found more peritumoral adhesions in the fluid–fluid level group; in addition, this group has worse outcome regarding rate of total tumor removal and FN preservation. They concluded that repeated ITH with subsequent inflammatory reaction resulted in destruction in the tumor–nerve barrier and consequently difficult dissection.³⁸ In addition, the FN is extremely thinned and stretched over the tumor in these cases, and this makes anatomical preservation extremely difficult. In cases of acute tumor expansion with acute presenting symptoms such as facial weakness, the condition

is more difficult. Severe facial weakness in these cases is usually a result of significant damage of the nerve. Even if anatomical preservation of the nerve could be possible, functional recovery may not occur. FN killed with reinnervation could be considered in these cases, and the results of FN grafting showed recovery to HB grade III or IV.²³ Surgical tumor removal with the aim of FN decompression should be offered to these cases. Careful dissection of the FN under optimal intraoperative neurophysiological monitoring techniques is recommended. If dissection between the tumor capsule and the FN is not possible, a thin sheath of tumor could be left over the FN. Considering preoperative severe facial weakness, complete tumor removal with FN reanimation could be a valuable option in some case.^{29,39} In our second case although direct FN stimulation resulted in a weak response, we preferred to leave a small part of the tumor at the FN entry to the IAC to preserve the anatomical integrity of the nerve; nevertheless, the facial function did not improve postoperative. Therefore, we could recommend total tumor removal with FN reanimation (using sural nerve graft) if the nerve could not be stimulated intraoperatively.

Conclusion

We discussed two cases of VS presenting with severe facial weakness. The cases represent different scenarios, one case of small tumor that compressed the FN near the IAC and the facial weakness improved after surgery. The other case represented a sudden deterioration due to hemorrhage in a cystic VS, with no improvement of facial function after subtotal tumor removal. Surgical treatment could be offered to cases of suspected VS presenting with facial weakness, as these cases may still have a chance for improvement especially in laterally located tumors. Intraoperative FN monitoring is very important in these cases, if the nerve is still stimulated proximally, functional recovery usually occurs. In cases where proximal FN stimulation was ineffective, complete tumor removal followed by FN reanimation should be performed.

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