

LETTER TO THE EDITOR

Two Cases of Secondary Hemifacial Spasm: Pathophysiology and Management

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Dear Editor,

Classical hemifacial spasm (HFS) has been attributed to an atraumatic pulsatile vascular compression of the facial nerve leading to hyperactivity within the central components of the nerve, alleviated via mobilization of the culprit vessel.¹ In a small subset of patients the cause may be attributed to lesions involving the facial nerve, most commonly benign cerebello-pontine angle (CPA) tumors.^{2,3} In such secondary HFS the clinical manifestation is usually a tonic facial contraction rather than the intermittent twitching characteristic of common HFS.^{4,5} There may, however, also be rare patients with a common presentation of HFS but with an associated CPA tumor;^{6,7} the role of neurovascular compression (NVC) in these cases is uncertain.

We present 2 cases of secondary HFS, both related to benign CPA tumors.

Case 1

A 56-year-old female with a known left sided vestibular schwannoma (VS) that was treated with fractionated stereotactic radiotherapy 5 years prior, presented with a 12 month history of acute onset and progressive HFS, characterized by periorbital twitching. At the time of radiosurgery the tumor measured 2.5 cm in diameter. Post treatment, she developed gradual sensory neural hearing loss, leaving her with a 50% reduction in hearing in the left ear. Brain magnetic resonance imaging (MRI) displayed no change in size since radiosurgery. It was elected to undertake a surgical debulking of the tumor combined with microvascular decompression (MVD) for HFS.

Intra-operatively a left retro-sigmoid approach to the CPA was taken. The inferior margin of the tumor was then exposed at the level of the pontomedullary sulcus, identifying an ectatic

vertebral artery and the left posterior inferior cerebellar artery (PICA) grooved into the caudal pons at the region of the root exit zone (REZ) of the facial nerve. The PICA was then elevated and mobilized from the REZ of the facial nerve and shredded Teflon felt was placed to maintain the vessel away from the nerve. Internal debulking of the tumor was performed, and the facial nerve was dissected off of the tumor capsule.

The bone overlying the internal acoustic canal (IAC) was removed exposing the dura of the IAC, which was then incised allowing tumor to spontaneously herniated through the opening. This decompressed the intra-cannicular portion of the facial nerve. Intra-operative monitoring demonstrated electromyographic lateral spread that was successfully abolished with the decompression. Brainstem auditory evoked potentials (BAEP) remained stable. Intra-operative illustrations of the lesion and MVD of the facial nerve can be seen in Figure 1.

Post-operatively her HFS improved and eventually abated, and hearing remained unchanged. She encountered no post-operative complications. She remains spasm free since 2003. The final pathology of the tumor returned as VS.

Case 2

A 51-year-old male, actor by profession, presented with a history of 3 episodes of right tonic facial contraction lasting several minutes. Further investigation he revealed decreased hearing in the right ear over the last 12 months and subtle balance difficulties. Brain MRI demonstrated a right CPA angle tumor filling the IAC, with mild compression of the middle cerebellar peduncle.

It was elected to perform a surgical debulking of the tumor and facial nerve decompression. Complete tumor resection was

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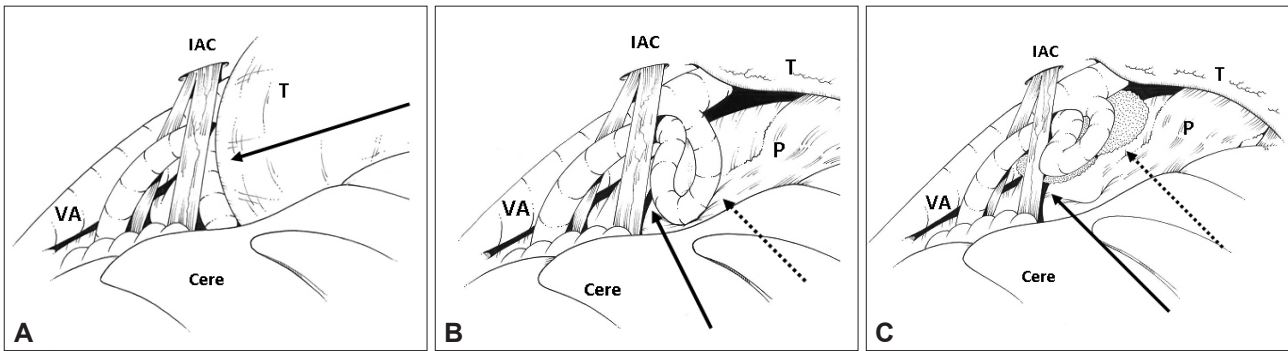


Figure 1. Intra-operative illustration of tumor and vascular compression. A: Lateral view of the cerebellopontine angle via a retro-sigmoid approach. Cerebellum retracted gently to expose the T compressing the posterior inferior cerebellar artery (PICA) (solid black arrow) and displacing it towards the cranial nerve (CN) VII and VII complex. B: T has been debulked, exposing the P and the loop of PICA contacting both the pons near the facial root entry zone (dotted arrow) and CN VII (solid arrow). C: PICA has been mobilized from the P and CN VII and padded with Teflon felt at both sites (dotted and solid arrows). IAC: internal acoustic canal, VA: vertebral artery, T: tumor, Cere: cerebellum, P: pons.

not planned, in order to minimize risks of cranial nerve injury.

A right retro-sigmoid approach was taken, with the origin of the facial nerve identified below the lower pole of the tumor, as confirmed with stimulation. There was no evidence of NVC on the REZ of the facial nerve. The lateral surface of the tumor was noted to be free of vessels and nerves, it was subsequently incised and internal debulking of the tumor occurred. Debulking stopped once tumor attached to the vestibule-cochlear nerve was noted to be adherent.

The IAC was opened as described in case 1 causing the tumor to herniate through the opening. This decompressed the intra-cannicular portion of the facial nerve. Dissection of the tumor off of the facial nerve was not conducted given adherence. Facial nerve electromyography did not display any injury potentials. The BAEP could not be obtained at the onset of surgery.

Post-operatively the tonic facial contractions completely resolved and there were no complications. Pathology returned as a World Health Organization grade I meningioma. Residual tumor will be followed radiographically with a plan for radiosurgery if growth is demonstrated.

Discussion

A few important points can be taken from these cases. First, the atypical clinical presentation (case 2) highlights the difference between classic primary HFS (case 1), related to pulsatile vascular compression of the facial nerve, and compression secondary to a CPA/IAC tumor. Second, the potential revers-

ible nature of the primary or secondary HFS related to tumor CPA tumors has been displayed in both cases. For the classical HFS symptoms (case 1), alleviation of NVC at the REZ of the facial nerve was effective while the tonic facial contractions of secondary HFS (case 2) were successfully alleviated with tumor decompression, particularly in the IAC. As a result, when presented with CPA tumors and facial twitching one must consider whether extensive tumor debulking versus alleviation of potential NVC is warranted. Symptoms of HFS secondary to NVC should be treated via standard MVD techniques, with elevation of the culprit vessel from the facial nerve and maintenance of separation with Teflon felt. Fourth, we do not believe that radiosurgery would be effective to treat either of these conditions, and was in fact utilized previously in our case 1. Smaller CPA tumors such as those found in our 2 cases are now often treated with stereotactic radiosurgery (SRS). This offers excellent tumor growth control and often regression in size over a couple of years. There is some data in the literature to suggest that SRS for tumor related HFS may lead to symptom resolution,⁸ with a latency period of 12 months or more until symptom improvement. In cases such as presented here, however, the most prominent clinical manifestations were HFS and best treated with decompression of the relevant point of symptomatic compression. In our case 1 where SRS was previously utilized, the patient had 5 years of follow-up and 12 months of progressive HFS symptoms. Given previous SRS and a lack of change in tumor on MRI, we suspected NVC of the facial nerve and elected for surgical exploration rather

than repeat SRS. Finally, complete surgical resection of these tumors was not necessary to achieve resolution of symptoms. Only decompression and alleviation of the associated NVC were required. This concept is important, as aggressive surgical removal of these tumors is associated with a higher risk of facial and vestibular-cochlear nerve impairment.

Conflicts of Interest

The authors have no financial conflicts of interest.

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