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Hemifacial Spasm Due to a Large Distant Ipsilateral Posterior Fossa Meningioma

ABSTRACT—A rare case of hemifacial spasm due to an ipsilateral foramen magnum/clival meningioma is described. Magnetic resonance imaging demonstrated that the tumor was located distant to the cranial nerve VII/VIII complex. Resolution of the ipsilateral hemifacial spasm was noted after complete resection of the tumor. The mechanism of hemifacial spasm was likely due to displacement and distortion of the brain stem from the lesion distant to the cranial nerve VII/VIII complex. In our review of the literature this is the first reported case of an ipsilateral posterior fossa meningioma causing hemifacial spasm from indirect mass effect.

Hemifacial spasm (HFS) is a clinical syndrome characterized by unilateral facial nerve dysfunction that begins with spontaneous paroxysms of eye twitching and progresses to involve other muscles innervated by the facial nerve. The paroxysms characteristically begin with a series of twitches, increasing in frequency and intensity, followed by a sustained spasm. Hemifacial spasm may be intensified by stress, anxiety, or fatigue. After some time, a mild ipsilateral facial weakness may be noted by the clinician.¹⁻³

In 1970, Jannetta⁴ popularized the concept that the most common etiology of HFS is compression of the facial nerve at its exit from the brain stem by one or more arteries or veins. The most common arteries being, in order, AICA, posterior inferior cerebellar artery (PICA), and the vertebral arteries. Other less frequent etiologies have been described to cause hemifacial spasm, including: aneurysm,

arteriovenous malformation (AVM), tumor, ectatic vertebral arteries and Paget's disease of the skull.

Tumors have been described to cause hemifacial spasm via direct or indirect mass effect. These include meningiomas, acoustic schwannomas, dermoids, epidermoids, and lipomas.^{1,5} Contralateral pathology has been implicated in causing HFS in the setting of numerous etiologies. Reported cases include meningioma, acoustic schwannoma, dermoid, lipoma, blood vessels, aneurysm, and AVM.^{6,12} An ipsilateral supratentorial falcine meningioma and an ipsilateral cerebellopontine angle lipoma have been reported to cause HFS.^{13,14} No case of an ipsilateral posterior fossa mass causing HFS from distant mass effect has been described in the literature.

We report a unique case of a large foramen magnum/clival meningioma causing ipsilateral HFS. The mass was distant to the cranial nerve VII/VIII complex

on preoperative magnetic resonance imaging (MRI) and the patient's hemifacial spasm resolved after removal of the tumor. We present the clinical features, radiological studies, operative findings, and a review of the literature on HFS caused by tumors.

CLINICAL PRESENTATION

A 63-year-old man presented with a one-and-one-half-year history of right-sided HFS of insidious onset involving the orbicularis oculi and orbicularis oris. The patient had undergone several botulinum toxin injections and, following the last one, developed paresis of

the right orbicularis oculi muscle resulting in incomplete eye closure. The patient had no hearing difficulties, dysphagia, or hoarseness and denied headaches or dizziness.

The neurological examination demonstrated a normal cranial nerve exam save the previously noted paresis of the right orbicularis oculi. The remainder of the neurological examination was unremarkable.

MRI with gadolinium contrast (Fig. 1) demonstrated a large mass involving the foramen magnum and lower clivus that compressed the brain stem dorsally and to the left. The cranial nerve VII/VIII complex was noted to exit the brain stem above and distinctly separate from the tumor.

Figure 1. A and B: Preoperative MRI with gadolinium demonstrates a large meningioma of the lower clivus and foramen magnum compressing the brain stem. The tumor is notably distant from the VII/VIII complex. C and D: Postoperative MRI with gadolinium demonstrates gross total resection of the tumor.

SURGICAL INTERVENTION

A far lateral approach was selected for surgery. Tumor was noted to displace the brain stem and vertebral artery toward the left. Cranial nerves XI and XII were sacrificed during the procedure to achieve gross total resection of the tumor. The VII/VIII cranial nerve complex was not observed at the time of surgery. The postoperative course was uneventful and the patient's HFS resolved within several days. A postoperative MRI with gadolinium confirmed gross total resection of the tumor (Fig. 1). Pathological analysis later confirmed meningotheiomatous (syncytial) meningioma. Nine months after surgery the patient was tolerating the right XI and XII cranial nerve deficits well, with rare mild facial twitches on the right occurring less than once per week.

DISCUSSION

The vast majority of HFS is thought to be due to vascular compression of the facial nerve as it exits the brain stem.^{4,15,16} Tumors have been cited to cause HFS in 0.3 to 0.6% of reported cases.^{5,15,17}

The pathogenesis of HFS remains a matter of some debate. Gardner and Sava¹ popularized the concept that HFS results from a false synapse, or "ephapse," formed when nerve fibers are pinched together causing a local demyelination within the facial nerve. This permits transaxonal "short circuiting" of the action potential. The "ephaptic theory" was later supported in the literature by other authors.^{18,19} Moller²⁰ postulated that vascular irritation of the facial nerve causes abnormal retrograde activity in the facial nucleus similar to the kindling phenomenon.

The mechanism of HFS from tumor may be due to direct or indirect mass effect. Masses may directly impinge on the facial nerve depending on the size and location of the tumor. The particular anatomy of the facial nerve and the surrounding posterior fossa structures may also play a role.^{7-9,12,21} Operative cases have been described where the facial nerve was compressed against bony structures or tethered to arachnoid adhesions.^{6,8,9,11,22} Similarly, a tumor may cause kinking of the brain stem and indirectly result in stretching of the facial nerve.

CONCLUSION

In our review of the literature, this is the first reported case of an ipsilateral posterior fossa meningioma causing HFS from distant mass effect. The precise mechanism cannot be definitively known in this case,

though relief from the HFS in the early postoperative setting strongly supports our assertion that distant mass effect played a primary role in this case.

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