Hemifacial Spasm Associated with Intraparenchymal Brain Stem Tumor

Athanassios Leonardos, MD, Paul E. Greene, MD, Louis H. Weimer, MD, Alexander G. Khandji, MD, and Pietro Mazzoni, MD, PhD*
The Neurological Institute, Columbia University, New York, New York, USA

Hemifacial spasm (HFS) is characterized by paroxysmal bursts of involuntary clonic and tonic activity in muscles innervated by the facial nerve.[1] Idiopathic HFS is hypothesized to be a result of microvascular compression of the facial nerve at its root exit zone in the brain stem.[2] Symptomatic (secondary) HFS is usually caused by compression of the facial nerve's root exit zone.[3] Whether HFS can also be caused by intraparenchymal brain stem lesions remains unclear.

A 58-year-old woman, who had been experiencing slowly progressive left facial weakness, diplopia, and dysphagia for 7 years, developed intermittent facial twitches around her left orbicularis oculi, lower face, and platysma. On examination, she had severe left facial weakness, left abducens palsy, moderate dysarthria, tongue atrophy, and mild right hemiparesis. Intermittent contractions were visible in left frontalis, orbicularis oculi, and zygomaticus muscles. These occurred as single twitches and as brief series of contractions (Video). Magnetic resonance imaging revealed a nonenhancing lesion (maximum diameter, 22 mm) in the left lower pons and the central and left upper medulla, with increased signal on T2-weighted and proton density images, consistent with a low-grade pontomedullary glioma or hamartoma. Notably, the lesion involved the facial nucleus and spared the root exit zones of both facial nerves (Fig. 1).

Nerve conduction studies showed significantly reduced amplitudes in the left facial nerve, especially in the orbicularis oris and mentalis branches; however, responses were evident in all facial muscles tested. In blink reflex studies, left supraorbital nerve (SON) stimulation produced normal-latency ipsilateral R1 and significantly prolonged ipsilateral and normal contralateral R2 latencies (55.3 and 39.9 ms, respectively). Right SON stimulation produced normal ipsilateral R1 and R2 latencies, but absent contralateral R2 response. These findings suggest interruption of normal inputs to the left facial nucleus. They are also evidence of cross-communication that could originate from the intramedullary portion of the facial nerve or from the facial nucleus. Electromyography revealed evidence of chronic but not acute neurogenic changes in affected facial muscles. Discharges consistent with HFS were recorded from the left orbicularis oris during an episode of repeated spasm-like contractions, typical of HFS, of the left cheek and perioral muscles. The left orbicularis oris had 1+ fasciculations, as well as intermittent single discharges associated with clonic spasms, typical of HFS. No myokymic discharges were observed.

Although involuntary facial muscle contractions have been reported in association with intraparenchymal lesions,[4,5] these conditions may have represented "spastic paretic hemifacial contracture" (SPHC), which is characterized by sustained facial muscle contractions and continuous muscle activity.[6,7] In 1 case[4] electromyography showed high-frequency bursts, but clonic spasms were not mentioned. In the other case[5] contractions were sustained, and electrophysiology showed tonic, nearly continuous activity of the facial nerve and high-frequency discharges in its innervated muscles. Both cases may thus have represented SPHC rather than HFS. To our knowledge, therefore, the present report is the first description of facial twitching with the clinical and electrophysiologic hallmarks of idiopathic HFS, associated with an intraparenchymal brain lesion affecting the facial nucleus but not the facial nerve's root.

Legend to Video. Hemifacial spasm and signs of brain stem dysfunction in the patient described in the present report. The video shows intermittent involuntary muscle contractions mainly involving the zygomaticus and to a lesser degree the frontalis and orbicularis oculi on the left side of the face. These occur at variable intervals, at times as single twitches and other times as brief trains of contractions.

Additional Supporting Information may be found in the online version of this article.

***Correspondence to:** Dr. Pietro Mazzoni, Neurological Institute, 710 W. 168th St., Box NI-175, New York, NY 10032, USA; pm125@columbia.edu

Funding agencies: Supported by a research grant from the Parkinson Disease Foundation.
Relevant conflicts of interest/financial disclosures: Nothing to report. Full financial disclosures and author roles may be found in the online version of this article.

Received: 8 February 2011; **Revised:** 12 May 2011; **Accepted:** 23 May 2011
Published online in Wiley Online Library (wileyonlinelibrary.com).
DOI: 10.1002/mds.23877

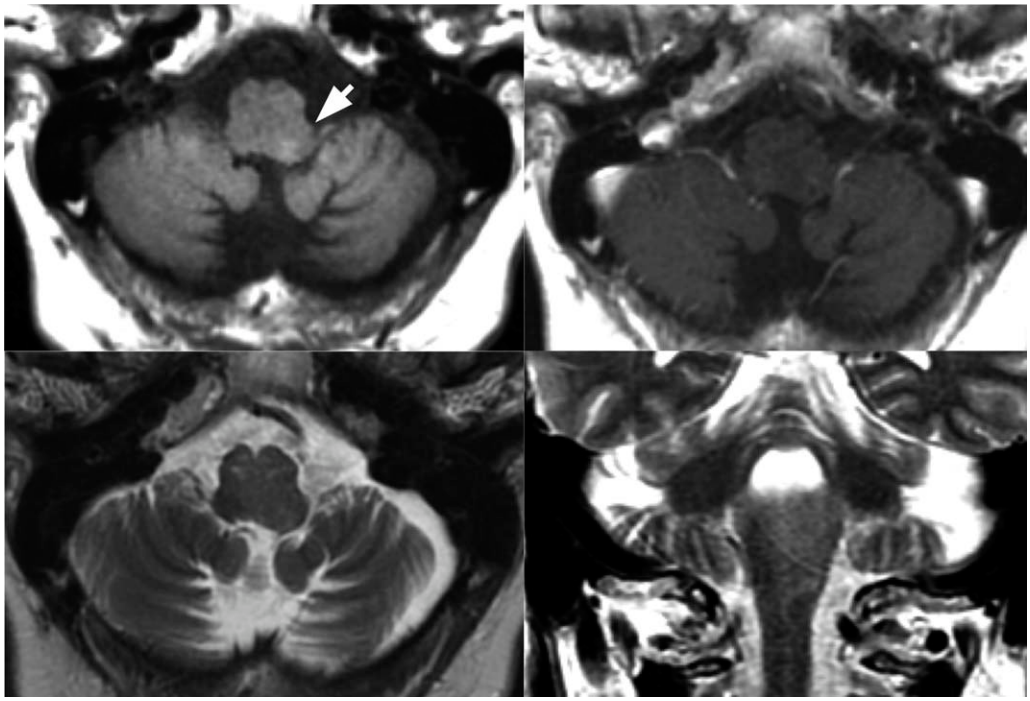

FIG. 1. Magnetic resonance images illustrating intramedullary mass lesion (arrow) in the left dorsal lower pons. The lesion extends into the central and left upper medulla and affects the facial nucleus but spares the root exit zones of both facial nerves. Clockwise from top left: axial T1-weighted image; axial T1-weighted, postintravenous gadolinium injection; coronal T2 weighted; axial T2 weighted.

Acknowledgments: We thank Dr. Toni Pearson for comments on this article and David Payne for assistance with video editing.

References

1. Greene P. Hemifacial spasm. In: Gilchrist J, ed. Prognosis in Neurology. Boston: Butterworth-Heinemann; 1998:173–175.
2. Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS. Etiology and definitive microsurgical treatment of hemifacial spasm. Operative techniques and results in 47 patients. J Neurosurg. 1977;47: 321–328.
3. Nagata S, Matsushima T, Fujii K, Fukui M, Kuromatsu C. Hemifacial spasm due to tumor, aneurysm, or arteriovenous malformation. Surg Neurol. 1992;38:204–209.
4. Westra I, Drummond GT. Occult pontine glioma in a patient with hemifacial spasm. Can J Ophthalmol. 1991;26:148–151.
5. Elgamal EA, Coakham HB. Hemifacial spasm caused by pontine glioma: case report and review of the literature. Neurosurg Rev. 2005; 28:330–332.
6. Sogg RL, Hoyt WF, Boldrey E. Spastic paretic facial contracture: a rare sign of brain stem tumor. Neurology. 1963;13: 607–612.
7. Koutsis G, Kokotis P, Sarrigiannis P, Anagnostouli M, Sfagos C, Karandreas N. Spastic paretic hemifacial contracture in multiple sclerosis: a neglected clinical and EMG entity. Mult Scler. 2008;14: 927–932.