LETTER TO THE EDITOR

To The Editor

Brainstem Developmental Venous Anomaly Causing Hemifacial Spasm – Case Report and Review of the Literature

Hemifacial spasm (HFS) is considered to be a form of segmental myoclonus of muscles innervated by the facial nerve (CN VII), with unilateral twitching beginning in the orbicularis oculi, and spreading to the other ipsilateral muscles of facial expression over several years. Over time, patients may present with sustained or tonic contractions.[1] The prevalence of HFS is 14.5 per 100,000 women and 7.4 per 100,000 men, with an average age of onset of 44 years.[1,2] Primary HFS is triggered by vascular compression whereas secondary HFS comprises all other causes of CN VII damage, thus, magnetic resonance imaging is recommended in all patients with HFS.[3] Primary HFS most commonly occurs secondary to arterial compression of the CN VII at the root exit zone (REZ).[4] Venous compression is rare.[4-6] Secondary causes include brainstem tumours, stroke, demyelinating plaques, trauma or Bell's palsy.[4] We describe a rare case of HFS associated with a pontine developmental venous anomaly (DVA).

A 32 year-old otherwise healthy woman presented with left HFS since the age of five years. The spontaneous twitching initially involved the left eye; subsequently progressing to become tonic spasms of the left face and anterior neck over several years. These were exacerbated by voluntary facial movements and stress; eventually increasing in frequency and persisting for a longer duration, lasting over one minute (Video 1). There were no speech changes, ocular deviation or other abnormal movements. The remainder of her neurological examination was normal. Electroencephalogram (EEG) was normal. Magnetic resonance imaging brain showed a DVA in the left pons in proximity to the left CN VII REZ (Figure 1). Carbamazepine and phenytoin were tried, but discontinued early due to adverse reactions (flu-like illness and vomiting, respectively). She responded to botulinum toxin (onabotulinumtoxinA) injections in the affected left facial muscles.

Hemifacial spasm is thought to result from hyperexcitability of the CN VII nucleus due to antidromic feedback from peripheral lesions compressing the nerve or ephaptic transmission within the proximal segment of CN VII, leading to excessive firing.[2] Hemifacial spasm is most commonly caused by vascular compression of CN VII by the anterior inferior cerebellar artery (43%), posterior inferior cerebellar artery (31%), vertebral artery (23%), or a large vein (3%).[4,5] Untreated, HFS can lead to social embarrassment and, in severe cases, involuntary eye closure may interfere with vision.[2] The prognosis depends on treatment selection and response. Anticonvulsants, such as carbamazepine, may be tried with variable efficacy (level U) and limited by side effects, such as sleepiness, headaches, dizziness and ataxia.[2] The response to anticonvulsants in certain cases of HFS may suggest another pathophysiological mechanism.

Microvascular decompression (MVD) has been shown to be effective in 84-98% of primary HFS patients (level C);[4,7] with delayed response in 25.4%,[8] sometimes taking up to three years for resolution of symptoms.[9] Recurrence may develop in 10.3% of patients within two years.[7] Causes of failed MVD may be related to missing the offending vessel, not detaching all offending vessels, or

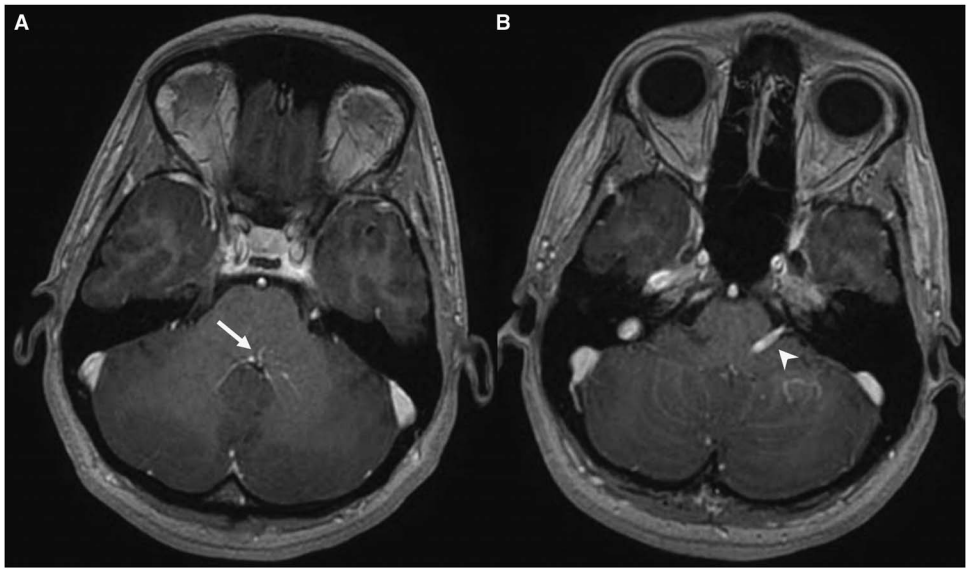

Figure 1: *Two selected images from axial post-contrast 3D T1 MRI sequence acquisition show a tuft of tributary veins, or "caput medusae" (A, arrow), consistent with a developmental venous anomaly[14], in the left posterior pons near the fourth ventricle leading to the larger draining abnormal vein (B, arrowhead) which is seen apposed to the left 7th cranial nerve in its root exit zone in the cerebellopontine angle cistern. There was no infarction, hemorrhage, mass or other relevant finding on the study.*

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Table 1: Reported cases in the literature of HFS associated with DVA.

Reference	Age/ Sex	Onset of HFS (years)	Lesion	Treatment
Mahran et al.[16]	58 F	60	Venous angioma located in the IAC, causing compression on right CN VII and VIII.	Vascular decompression (right retrosigmoid craniectomy)
Chen et al.[15]	53 F	51	Left cerebellar hemisphere venous angioma with distal portion of parenchymal segment compressing CN VII	Vascular decompression (left retrosigmoid craniectomy)
Arita et al.[17]	38 F	21	Left pontine cavernous angioma with venous angioma exiting ventrally to CN VII in prepontine cistern (non-compressive).	Botulinum toxin (unsatisfactory); left retrosigmoid craniotomy, but no evidence of compression.
Chiaramonte et al.[13]	59 M	59	Left cerebellar DVA with oblique course to the sigmoidal sinus; compression of Left CN VII at REZ.	CBZ 600 mg/d
Our Case	32 F	5	Left pontine DVA with oblique course and compression of left CN VII at REZ.	Botulinum toxin was preferred by the patient over surgery or anticonvulsants.

CBZ: carbamazepine; DVA: developmental venous anomaly; F: female; HFS: hemifacial spasm; M: male; CN: cranial nerve; REZ: root exit zone; IAC: internal auditory canal.

related to a difficult surgical approach.[10] Intraoperative electromyography monitoring over the mentalis, looking for the reduction or elimination of the abnormal muscle response may help increase the success of MVD.[7] Although repeat MVD may be required with good reported success,[8] this puts patients again at risk of surgical complications, including hearing loss, facial palsy, ataxia, cerebrospinal fluid leakage, diplopia, headache, wound infection, and vertigo.[11,12] Due to these surgical risks, delayed response, and occasional difficulty in identifying a culprit vessel, botulinum toxin may be used as first line for HFS. Microvascular decompression may be performed after failed treatment with botulinum toxin.[11,12]

A recent study of 15 patients (mean 52.8 years) suggests that patients with HFS associated with venous compression show a higher incidence of neck (platysma muscle) involvement and tonic contraction (tonus) of the facial muscles,[5] as seen in our patient. Infratentorial veins may be the primary or secondary offending vessel in HFS. Female sex and venous involvement have been associated with recurrence of HFS after MVD.[5] This may be related to location or severity of compression.

There are very few cases of HFS due to DVA reported in the literature (Table 1). Developmental venous anomalies, also known as venous angiomas, are congenital benign anatomic variants which account for over 60% of cerebral vascular malformations; with very low risk for bleeding or for causing symptoms, unless associated with a cavernous malformation.[13] Typically, neurosurgical manipulation of DVAs is generally avoided, due to their relatively benign nature and vital role in drainage of normal brain tissue, fragility and risk of venous infarction from manipulation.[14,15] There is minimal literature on MVD for DVAs associated with HFS, with resolution of HFS symptoms in two[15,16] of the three reported cases; without long-term post-operative followup.[15-17] Botulinum toxin is thus a reasonable option for symptomatic treatment of HFS associated with isolated DVA, as selected for our patient.

Of the commercially available formulations of botulinum toxin in Canada, evidence supports a level B recommendation for onabotulinumtoxinA, and a level U recommendation for incobotulinumtoxinA for the symptomatic treatment of HFS.[18] The latency of improvement after botulinum toxin injections in HFS is between 2-14 days, with mean duration of improvement of 15.7 weeks, and 85% improvement in spasms, as reported by patients.[19] Side effects are transient and most commonly include mild ptosis (mean: 24%) and facial weakness (mean: 27%).[19]

Our patient is the youngest reported case of HFS associated with isolated pontine DVA with symptoms presenting in childhood. Because the complication rate and recurrence risk is high for MVD of veins associated with HFS, botulinum toxin may be a reasonable treatment to offer such patients.

Disclosures

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Statement of Authorship

P Rizek – Study concept, design, writing of the first draft and revision of the manuscript. N Kumar – Study concept and design. M Sharma – Selection of imaging and interpretation. M Jog – Study supervision, revision and critique of the manuscript.

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Supplementary Material

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References

1. Lu AY, Yeung JT, Gerrard JL, Michaelides EM, Sekula RF, Bulsara KR. Hemifacial spasm and neurovascular compression. Sci World J. 2014;2014:349319.
2. Chaudhry N, Srivastava A, Joshi L. Hemifacial spasm: The past, present and future. J Neurol Sci. 2015;356:27-31.
3. Glocker FX, Krauss JK, Deuschl G, Seeger W, Lucking CH. Hemifacial spasm due to posterior fossa tumors: the impact of tumor location on electrophysiological findings. Clin Neurol Neurosurg. 1998;10:104-11.
4. Campos-Benitez M, Kaufmann AM. Neurovascular compression findings in hemifacial spasm. J Neurosurg. 2008;109:416-20.
5. Wang X, Thirumala P, Shah A. The role of vein in microvascular decompression for hemifacial spasm: a clinical analysis of 15 cases. Neurol Res. 2013:389-94.
6. Jannetta PJ. Hemifacial spasm caused by a venule: case report. Neurosurgery. 1984;14(1):89-92.
7. Zhu J, Li S-T, Zhong J, et al. Role of arterioles in management of microvascular decompression in patients with hemifacial spasm. J Clin Neurosci. 2012;19:375-9.
8. Xia L, Zhong J, Zhu J, Dou N-N, Liu M-X, Li S-T. Delayed Relief of Hemifacial Spasm After Microvascular Decompression. J Craniofac Surg. 2015;26:408-10.
9. Jo KW, Kong D-S, Park K. Microvascular decompression for hemifacial spasm: long-term outcome and prognostic factors, with emphasis on delayed cure. Neurosurg Rev. 2013;36:297-301; discussion 301-2.
10. Zhong J, Li ST, Zhu J, et al. A clinical analysis on microvascular decompression surgery in a series of 3000 cases. Clin Neurol Neurosurg. 2012;114:846-51.
11. Miller LE, Miller VM. Safety and effectiveness of microvascular decompression for treatment of hemifacial spasm: a systematic review. Br J Neurosurg. 2012;26:438-44.
12. Wang X, Thirumala PD, Shah A, et al. Effect of previous botulinum neurotoxin treatment on microvascular decompression for hemifacial spasm. Neurosurg Focus. 2013;34:E3.
13. Chiaramonte R, Bonfiglio M, Amore AD, Chiaramonte I. Developmental Venous Anomaly Responsible for Hemifacial Spasm. Neuroradiol J. 2013:201-7.
14. Ruíz DSM, Yilmaz H, Gailloud P. Cerebral developmental venous anomalies: current concepts. Ann Neurol. 2009;66(3): 271-83.
15. Chen HJ, Lee TC, Lui CC. Hemifacial spasm caused by a venous angioma. Case report. J Neurosurg. 1996;85:716-7.
16. Mahran A, Samii M, Penkert G, Ostertag H. Vascular lesions of the internal auditory canal. Skull Base Surg. 1991;1: 78-84.
17. Arita H, Kishima H, Hosomi K, et al. Hemifacial spasm caused by intra-axial brainstem cavernous angioma with venous angiomas. Br J Neurosurg. 2012;26:281-3.
18. Hallett M, Albanese A, Dressler D, et al. Evidence-based review and assessment of botulinum neurotoxin for the treatment of movement disorders. Toxicon. 2013;67:94-114.
19. Sorgun MH, Yilmaz R, Akin YA, Mercan FN, Akbostanci MC. Botulinum toxin injections for the treatment of hemifacial spasm over 16 years. J Clin Neurosci. 2015;22:1319-25.