

Hemifacial spasm resulting from vertebral artery dolichoectasia

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ABSTRACT

A 32-year-old man presented with left hemifacial spasm. Neurophysiological findings revealed an absent ipsilateral R1 on blink reflex. An MRI showed a dolichoectatic left vertebral artery impinging on the root exit zone of the left facial nerve. Botulinum toxin injections relieved the manifestations of hemifacial spasm. This case demonstrates that MRI/MRA is an essential part of the work-up for hemifacial spasm, and shows that in accordance with the literature, vertebral dolichoectasia is an uncommon cause of hemifacial spasm.

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Hemifacial spasm (HFS) is one of the most common movement disorders in young and middle-aged patients, and increases with age.¹ It presents as involuntary clonic contractions or twitching of the ipsilateral facial muscles innervated by the seventh cranial nerve. Advanced neuroimaging techniques reveal that compression of the facial nerve, as it emerges from the brainstem (root exit zone), by normal or aberrant vascular structures is by far the most common cause, accounting for 30% of cases.² Occasionally, the compression may be the result of a tumor (1%), bony abnormality (1%), multiple sclerosis or nerve injury (0.5%).³ Last but not least, idiopathic causes account for the remainder. Although the exact pathophysiological mechanism needs to be unraveled, 2 physiopathological hypotheses may explain HFS: in the emphatic theory, hyperactivity and antidromic impulses are generated by the offending vessel probably leading through demyelination of the nerve trunk to an abnormal excitation of motoneurons in the facial nerve nucleus; alternatively, in the nuclear theory, hyperactivity of the facial nerve is due to an unusual and involuntary activity of the facial nerve nucleus

itself, induced by the compressing vascular structure.⁴ Carbamazepine, baclofen, clonazepam, gabapentin and, recently, levetiracetam, are effective in the treatment of the spasms.^{5,6} However, repeated Botox (Botulinum toxin type A) injections (often ad infinitum) of the affected muscles every few months, and neurovascular decompression are now established procedures to treat HFS.⁵ Dolichoectasia (also called mega dolichoectasia, fusiform aneurysm or tortuous arteries) is an uncommon but well-recognized vascular anomaly, and is defined as a dilatation and elongation of the artery (vascular loop) as a result of pathophysiological factors such as hypertension and arteriosclerosis. Dolichoectasia of intracranial arteries most commonly involves the basilar artery, the supraclinoid segment of the internal carotid artery, middle, anterior and posterior cerebral arteries.⁷ This report shows the MRI/MRA features associated with HFS as an unusual complication of vertebral dolichoectasia.

Case Report. A 32-year-old right-handed man presented with a 6-month history of painless

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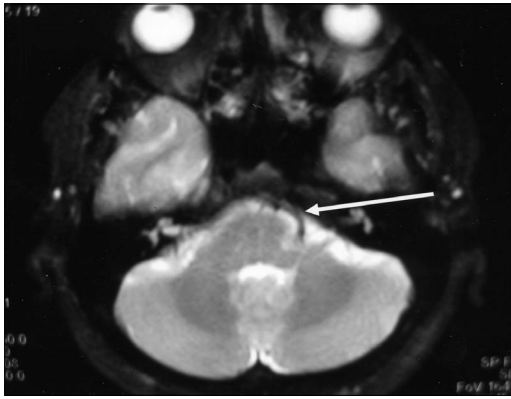


Figure 1 - Tortuous left vertebral artery (arrow) impinging on the root exit zone of the left facial nerve.

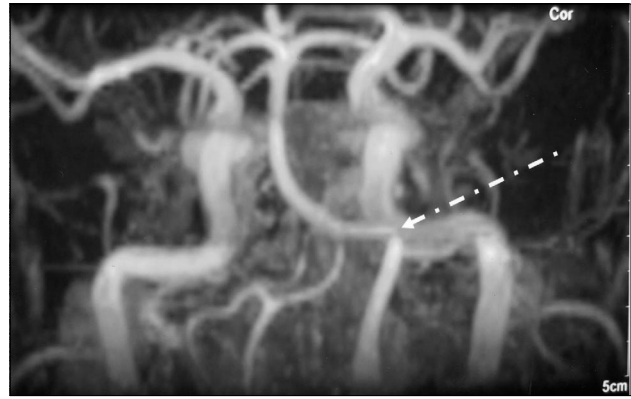


Figure 2 - Reveals the tortuous pattern of the left vertebral artery with its extreme bending towards the left caudal pons area where it compresses the root of the seventh cranial nerve (arrow).

intermittent twitching on the left side of the eyes and upper cheek, exacerbated by eye blinking and chewing. He had no significant past medical or family history. Neurological examination revealed ipsilateral, involuntary, paroxysmal contraction of the facial musculature, particularly provoked during activity (speech, blinking and so forth), consistent with hemifacial spasm. The ophthalmologic evaluation was normal and hearing was intact. Corneal and facial sensations were bilaterally normal. There was no facial weakness. No long-tract signs were observed and there was no cerebellar or akinetic-rigid syndrome. Biochemical analysis of blood, including serum ceruloplasmin was normal. High-resolution MRI/MRA of the caudal pons with focus at the cerebellopontine angle revealed the presence of a dolichoectatic left vertebral artery impinging on the root exit zone of the left facial nerve (**Figures 1 & 2**). Electromyography revealed synchronous bursts of repetitive high-frequency motor unit discharges in the left orbicularis oculi and oris muscles, and the blink reflex studies showed absent R1 on the affected side. The latter indicates ipsilateral facial nerve lesion. He was successfully treated with Botox infiltrations but refused neurosurgical intervention.

Discussion. An MRI or MRA is the optimal imaging techniques to visualize facial nerve compression in HFS, with a sensitivity of approximately 88%.⁸ Its advantages over cerebral angiography and computed tomography are its non-invasiveness and the possibility of demonstrating the vascular loop directly compressing the facial nerve. Microsurgical vascular decompression is the curative treatment of choice, and is effective in 70-95% of patients.⁵ Despite this, recurrences (10-20%), treatment failure (3-5%), and major

neurosurgical complications (including hearing loss [2.3%], permanent facial paresis [1.4%], cerebellar hematoma and brainstem infarction) may occur.⁹ As reported in the literature, we observed improvement of HFS in our patient, after botox infiltration.¹⁰ Our patient presented with a dolichoectasia of the ipsilateral vertebral artery which caused compression of the facial nerve at the root exit zone. Large studies correlating the angiographic manifestations and intraoperative findings revealed that the anterior inferior cerebellar and posterior inferior cerebellar artery predominantly caused the nerve-vessel conflict.^{9,11} The main trunk of the basilar or vertebral artery itself, however, has rarely been described as the causative factor. The vertebral artery was the offending vessel in less than 13% of cases. In addition, all these reports are related to anatomical variations and do not refer to dolichoectasia. Although several cases of HFS have been reported, few of them are actually caused by vertebral artery dolichoectasia, and displayed the characteristic neuroimaging abnormalities of HFS caused by compression of the facial nerve due to this dolichoectasia.¹²⁻¹⁴ In a series of 40 patients diagnosed with HFS at our institution, none of them had neuroimaging evidence of dolichoectasia. In contrast to vertebral artery dolichoectasia, basilar artery dolichoectasia demonstrated by MRI/MRA appears to be relatively more common.¹⁵ Besides its potential for compressive and mass effect on the brainstem, the importance of diagnosing dolichoectasia is related to its relative high risk of thrombosis and even hemorrhage.⁷

In conclusion, our case demonstrates that MRI/MRA is an essential part of the work-up for HFS, and shows that confronted with the literature, vertebral dolichoectasia is an uncommon cause of HFS.

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