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CASE REPORT

Hemifacial spasm secondary to vascular loop compression: a rare case report

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Abstract Hemifacial spasm (HFS) is characterised by brief, persistent, involuntary paroxysmal contractions of the facial muscles innervated by the facial nerve. Broadly its aetiology is portrayed as primary and secondary. Primary HFS is a result of vascular compression of the ipsilateral facial nerve at its root exit zone, and secondary HFS can occur after any injury to the facial nerve from the internal auditory canal to the stylomastoid foramen, which may be a result of a cerebellopontine angle tumour, schwannoma, fusiform aneurysm, or demyelinating lesion such as multiple sclerosis. We report a rare case of HFS in a 40-year-old female patient, who presented with a 4-year history of twitching of the left eye and deviation of the mouth towards the left side. An MRI of the brain revealed a vascular anomaly at the root exit zone of the left facial nerve. The present report aims to highlight MRI as a single, non-invasive diagnostic investigation to confirm the diagnosis of HFS.

Keywords Involuntary facial twitching · Tortuous vertebral artery · Vascular compression · Magnetic resonance imaging · Hemifacial spasm

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Introduction

Hemifacial spasm (HFS) has been reported in middle-aged females, particularly those aged 40–50 years [1–3]. At the onset of the disease, involuntary spasms typically start off around the eyelid (90% of cases) and, with time, tend to involve other facial muscles such as the orbicularis oris, zygomatic muscles, frontalis, corrugator, mentalis and platysma. Insomnia has been reported in some cases because of the persistence of involuntary facial twitches during sleep [4]. The involuntary spasms are exaggerated when the patient speaks or is stressed, fatigued or anxious, and are sometimes relieved while at rest [1]. Bilateral involvement is rare, but whenever present, twitches are always asynchronous and asymmetrical [5]. Bilateral and unilateral hearing loss has also been reported in 13% of patients [1]. Aetiologically, HFS can be primary or secondary. Primary HFS occurs as a result of vascular compression of the ipsilateral facial nerve at the root exit zone, and secondary HFS may occur following damage anywhere along the facial nerve from the internal auditory canal to the stylomastoid foramen [6]. Secondary causes include cerebellopontine angle tumour, schwannoma, fusiform aneurysm and demyelinating lesions. The offending vessels causing primary HFS include the anterior–inferior cerebellar artery, the posterior–inferior cerebellar artery and the vertebral artery [7]. Magnetic resonance imaging (MRI) and magnetic resonance angiography of the brain are the preferred investigative modalities in these patients to reveal any vascular compression caused by an artery of the vertebrobasilar system on the ipsilateral facial nerve at the root exit zone [8]. The current case report describes a patient presenting with HFS resulting from a vascular anomaly at the root exit zone.

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Case report

A 40-year-old female presented at the Oral Medicine Department with a 4-year history of episodes of painless twitching on the left side of the face. The patient disclosed that the twitching had increased gradually over the preceding 3 months. The twitching began in the lower left eyelid and extended to involve the lower facial muscles, causing deviation of the angle of the mouth to the left side. Involuntary contractions of the facial musculature were particularly pronounced during speech and were rarely relieved at rest (Fig. 1). The general physical examination and oral examination were unremarkable. The medical history of the patient was significant for mild hypertension, but not for hypercholesterolaemia.

After clinical examination of the patient, a provisional diagnosis of HFS was made. Investigations such as a complete blood haemogram and lipid profile were recommended, but the results of these were not contributory. MRI was recommended because it provides vascular and brain tissue diagnosis in a single non-invasive examination, and is the imaging method of choice in evaluating disorders of the facial nerve along its pathway [9]. A T2 weighted MRI brain sequence and magnetic resonance angiography were carried out to assess the presence of any vascular abnormality at the level of the brainstem. The brain MRI revealed an exceptionally tortuous course of the left vertebral artery impinging on a deep cerebellopontine angle in the area of the root exit zone of the left facial nerve (Figs. 2, 3, 4). Based on the MRI results, a final diagnosis of HFS of the left side of the face secondary to a vascular loop compression was established. Discussion with the patient followed about the therapeutic options available, such as botulinum toxin injection, medicinal treatment, or surgery. Because of monetary issues and the reluctance of the patient to undergo treatment with botulinum toxin, we adopted a conservative medical line of treatment with carbamazepine (100 mg BD) and pregabalin (75 mg OD). Following the initiation of treatment, the patient was asked

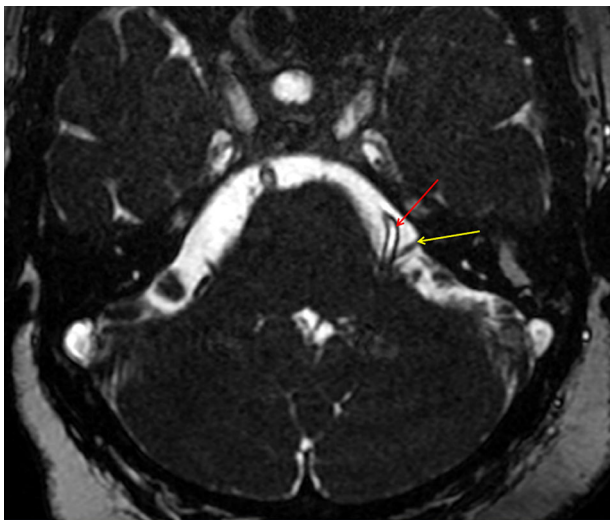

Fig. 2 Axial magnetic resonance image (3D FIESTA sequence) showing the left vertebral artery (*red arrow*) taking a tortuous course and impinging on the deep cerebellopontine angle in the area of root exit zone of the left facial nerve (*yellow arrow*)

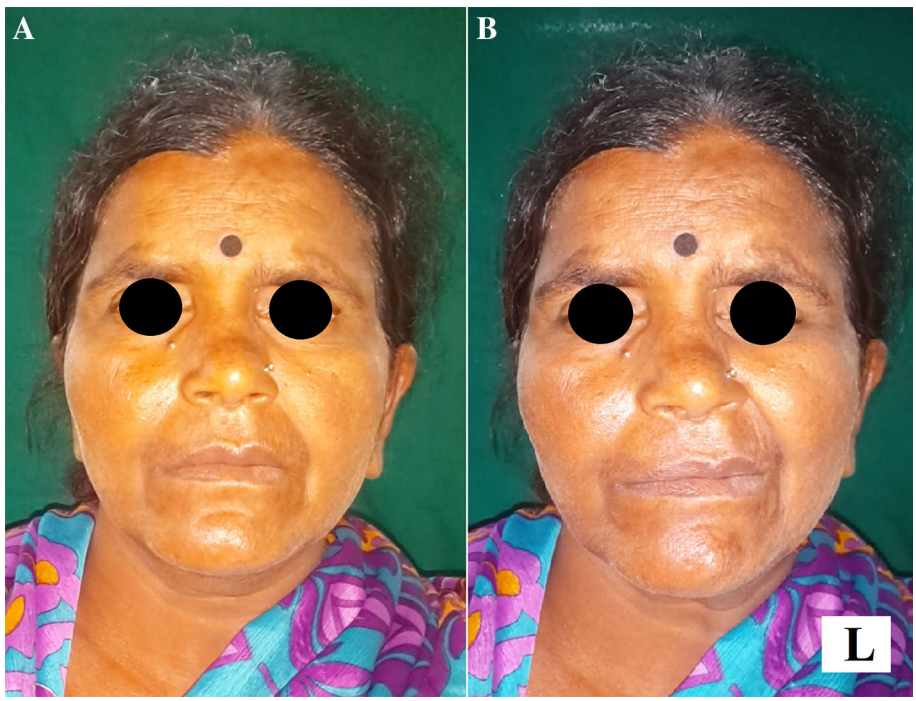

Fig. 1 Profile of the patient without clinical symptoms (**a**), and with clinical symptoms (**b**)

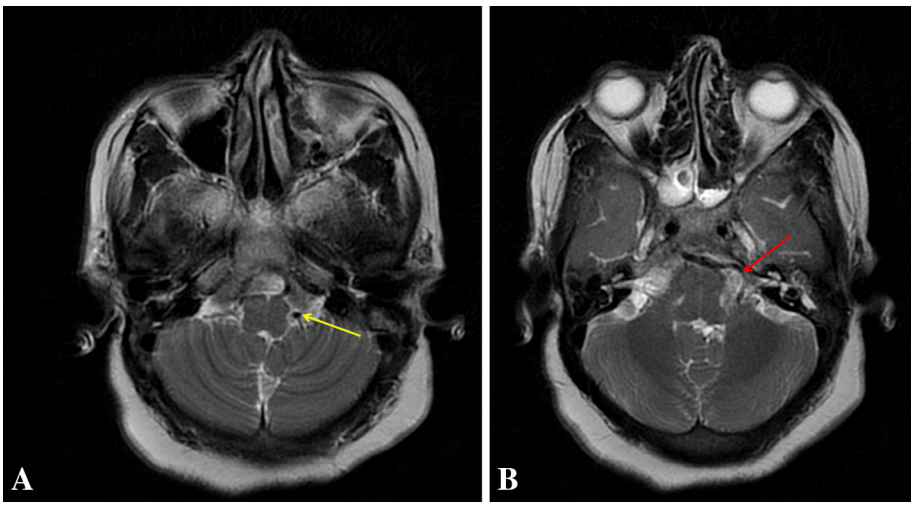

Fig. 3 Axial T2W magnetic resonance images taken at the cerebellopontine angle, showing the left facial nerve (*yellow arrow*) (**a**), and showing the flow void of the tortuous left vertebral artery (*red arrow*) (**b**) grossly deviated from its usual course, causing significant compression of the left facial nerve at its root exit zone from the brainstem. There is no evidence of other pathological changes such as dolichoectasia

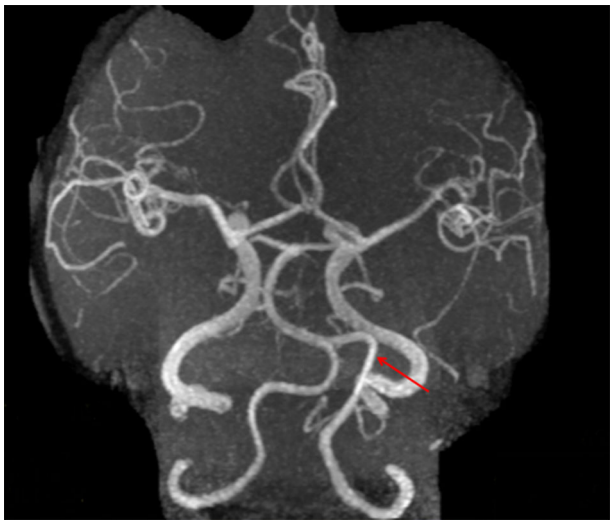

Fig. 4 Coronal magnetic resonance angiography image showing tortuous looping of the left vertebral artery (*red arrow*)

to regularly register any occurrence of the involuntary twitches. After 1 month, the patient's entries revealed 80% resolution of the involuntary spasms. The treatment regimen was continued, with the patient on regular follow-up.

Discussion

Hemifacial spasm is a peripherally induced neuromuscular movement disorder typically delineated by brief or persistent involuntary tonic–clonic contractions of the muscles innervated by the facial nerve. The earliest description of HFS was provided by Gowers in 1884 [10]. The prevalence is highest in subjects aged 40–79 years [4] and in the Asian population [11], with a male to female ratio of 1:2 [12]. The average age-adjusted annual incidence of HFS is 0.78/100,000 (0.81/100,000 in women and 0.74/100,000 in men) [13]. HFS is often unilateral, but occasional bilateral cases have been reported with a prevalence of 0.6–5% [14].

Initially HFS occurs around the eye (the orbicularis oculi in 90% of cases) [1], with involuntary closure of the eyelid on elevation of the eyebrow, and progressively spreads to the cheek and perioral musculature, as observed in our case. In severe cases, the platysma is also affected. Spasms are accentuated during stress, fatigue and vocal activities, are sometimes relieved at rest, and persist even during sleep. However, insomnia, stress and anxiety were obviated in our case after assessment with the Insomnia Severity Index and the Depression Anxiety Stress Score.

The differential diagnoses considered included blepharospasm, oromandibular dystonia, facial nerve tic and hemimasticatory spasm. In contrast with the unilateral spasms of HFS, blepharospasm presents with bilateral, symmetrical, synchronous contractions of the eyelids [12]. Oromandibular dystonia presents with muscle contractions that primarily affect the lower part of the face, mouth, maxilla, tongue and pharynx. Facial nerve tics manifest as non-rhythmic movements that alternate between the right and left side of the face [12] and, in contrast with HFS, these tics are voluntarily reproducible and transiently suppressible. Hemimasticatory spasm elicits unilateral painful contractions of the muscles of mastication [12].

Clinically, HFS can be diagnosed by a physical examination manoeuvre known as the "brow lift sign", which is positive when a patient lifts an eyebrow with ipsilateral eye closure, denoting the synchronised activity of the frontalis and orbicularis muscles characterising HFS [14], as exhibited in our case. Motor testing of the facial nerve revealed no abnormality. Sensory testing and gustometry were not included because the patient had no complaints regarding altered taste sensation.

Computed tomography (CT) and MRI are used to diagnose and differentiate primary HFS from secondary HFS [14]. However, CT was not recommended because of its limited usefulness and inferior resolution in evaluating the posterior fossa and the first portion of the facial nerve, and poor exhibition of vascular interactions with the parenchyma. MRI, however, clearly delineates the path of the facial nerve from the root exit zone to the internal auditory canal and its relationship to the vertebrobasilar system [15].

Therapeutic options for HFS include the use of anticonvulsants such as carbamazepine and clonazepam; various benzodiazepines; GABAergic drugs such as pregabalin, gabapentin and baclofen; subcutaneous injection of botulinum neurotoxin; and microvascular decompression surgery. Microvascular decompression is a definite, effective and safe treatment with a success rate of >90% [16].

Correct diagnosis remains the key to successful management of HFS. MRI and magnetic resonance angiography of the brain have been proven to be non-invasive diagnostic investigations that provide essential details about the vertebral basilar system and the vascular parenchymal relationship.

Compliance with ethical standards

Conflict of interest Pratibha S. Sharma, Atul P. Sattur, Preetam B. Patil, Kirty R. Nandimath, Kruthika S. Guttal and Krishna Burde declare that they have no conflict of interest.

Ethical standards All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008(5). Informed consent was obtained from all patients for being included in the study.

Research involving human participants and/or animals This article does not contain any studies with animal subjects performed by any of the authors.

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