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Letter to the Editor

More than hemifacial spasm? A case of unilateral facial spasms with systematic review of red flags

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ABSTRACT

Unilateral facial spasms (UFS) are frequently caused by hemifacial spasm (HFS), a disorder that usually results from vascular loop compression at the root exit zone of the facial nerve. However, UFS can also be a manifestation of other conditions, including brainstem tumours or demyelination, post-Bell's synkinesis, lesions of the facial nerve in the Faloppio canal and the parotid gland, dystonia, epilepsy, psychogenic conditions, tics and hemimasticatory spasm. In this report, we present a case of UFS, not due to HFS, highlighting clinical red flags for an alternative diagnosis. In addition, a systematic review was conducted to provide a comprehensive summary of UFS differential diagnoses with a list of red flags to assist neurologists in the evaluation of patients with UFS.

Unilateral facial spasms (UFS) represent a common clinical sign where, in most cases, a diagnosis of hemifacial spasm (HFS), a disorder caused by a mechanical irritation of the facial nerve at its brainstem exit by an aberrant arteriole, is usually considered [1]. However, other causes of UFS are not that uncommon, accounting for one out of five patients, with reported aetiologies ranging from post-paralytic facial synkinesis (Post-Bell's synkinesis), facial injury, brainstem tumours to demyelination [2,3]. Certain disorders may also mimic HFS, including psychogenic movements, tics, dystonia, myoclonus and hemimasticatory spasm. In this article, we present the case of a patient with UFS, highlighting potential red flags that suggest an alternative diagnosis to HFS. A systematic review was also conducted to identify a comprehensive list of red flags for different causes of UFS.

A 21-year-old woman presented with a one-year history of left eyelid twitching, which, over the next three months, rapidly progressed to involve all her left facial muscles. Although she did not suffer any headache, weakness, or ataxia, careful examination revealed tonic, rather than clonic, spasms of her left upper and lower facial muscles in association with vermicular-like movements of the left orbicularis oculi and oris muscles (Supplementary video clip 1). Also, her left nasolabial groove was prominent, in association with a narrow palpebral fissure, whereas the Babinski-2 sign was absent. A subtly reduced pinprick sensation was demonstrated in the left trigeminal nerve distribution, together with a slight reduction of left corneal reflex. The rest of the examination was unremarkable. The brain MRI scan showed a 2*3 cm ill-defined hyperintensity lesion involving the left dorsal pons and the superior and middle cerebellar peduncles (Fig. 1A). Concentric needle electromyography of the left facial muscles demonstrated duplets and multiplets of myokymic discharges with a characteristic audio signature typical of 'Marching soldiers in the snow', whereas motor unit action potentials and recruitment patterns were normal (Fig. 1B; Supplementary video clip 2). Stereotactic lesional biopsy confirmed a pathological diagnosis of low-grade glioma. Based on her possible low-risk for progression as identified by the age and the tumour size (< 4 cm), the patient opted for observation where there was no clinical or MRI progression at a 1-year follow-up visit [4]. As her tumour was deemed to be unresectable, it is difficult to determine the overall survival rate. However, in those with favourable prognosis (< 40 years of age and gross tumour resection), the overall survival rates at 2 and 5 years were 99 and 93%, respectively [4].

The clinical presentation of this patient highlights several red flags that, are suggestive of an alternative diagnosis, not HFS, as the cause of her UFS, including rapid progression, sustained tonic rather than clonic contractions of facial muscles, myokymic discharges, and simultaneous onset of the upper and lower facial spasms with trigeminal sensory neuropathy. In order to identify a comprehensive list of red flags of UFS, a full literature search was conducted according to the Preferred of Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. These were categorised into movement characteristics, abnormal movement distribution, clinical course, coexisting features, and investigations (Table 1). Full details of the literature search, including search terms and methodology are provided as Supplementary data 1.

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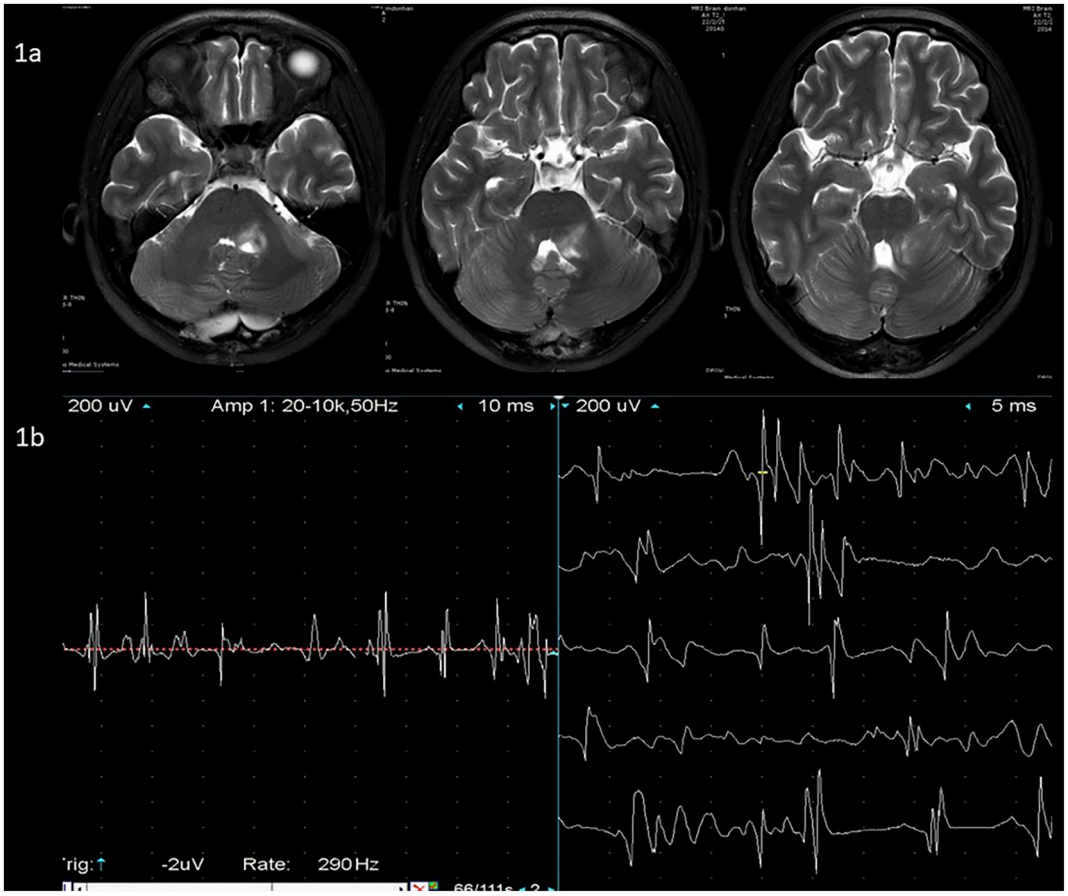

Fig. 1. A: The brain MRI scan (T2-weight images) showed a 2*3 cm ill-defined hyperintensity lesion involving the left dorsal pons, left superior and middle cerebellar peduncles, and anterior aspect of left cerebellum; B: Concentric needle electromyography of left orbicularis oris muscle demonstrated brief bursts of doublets and triplets firing at irregular intervals ranging from 50 to 150 Hz.

From this systematic review, we identified several features that are associated with other causes of UFS, not limited to HFS, and frequently indicative of brainstem pathologies. We have chosen to classify UFS according to the aetiology of the HFS and other pathologies as we find the terms 'atypical HFS', 'HFS mimickers', or 'symptomatic HFS' not clear enough. For example, to name it 'symptomatic HFS' seems misleading as all conditions associated with UFS are clearly symptomatic from the patients' perspective. In our case, the presence of sustained contracture or tonic spasms of paretic unilateral facial muscles, with prominent nasolabial groove and narrow palpebral fissure, is suggestive of intrinsic pontine lesions involving corticofacial fibers [5]. While considered to be rare, it is suggestive of demyelination or low-grade glioma [5,6]. Indeed, the prevalence of pontine glioma presenting with UFS is unknown but thought to be very uncommon where nine case reports documented this rare association with at least four cases reported in children [5,7,8]. Also, facial myokymia should be specifically looked for when spasms are sustained as its presence further reinforces the possibility of a lesion within the facial nucleus or surrounding pontine area [5,8]. However, clinical manifestations of myokymia can be subtle and its appearance can be temporarily, therefore a high level

Table 1
Red flags for alternative causes of unilateral facial spasms, not hemifacial spasm.

Red flag category	Characteristic/feature	Underlying disorder
Movement characteristics	Tonic, rather than clonic, unilateral facial spasms (lasting > 3 s, spastic paretic hemifacial contracture)	Brainstem lesions (usually pontine glioma or demyelination)
	Undulating, vermicular-like movements under the skin (Myokymia)	Facial myokymia (Lesions could be central or peripheral). In patients with unilateral facial spasms, it is usually indicative of pontine glioma or demyelination.)
	Repetitive, rhythmic, slow (1–4 Hz) movements (Myorhythmia)	Brainstem, thalamus, diencephalic lesions (reported in stroke, anti-*N*-methyl-D-aspartate encephalitis, steroid encephalopathy associated with autoimmune disorders, Whipple's disease, and multiple sclerosis)
	Unwanted muscle contractions (e.g. chin and neck contraction) during voluntary facial movements (e.g. smile, eye closure)	Facial synkinesis (due to aberrant facial nerve regeneration)
	Absence of the other Babinski's or Babinski-2 sign	Various causes of unilateral facial spasms, not HFS)
		Blepharospasm
	Stereotyped facial contractions with/without eye deviations	Epilepsia partialis continua, hemifacial seizures
	Inconsistent and incongruous, tonic contraction, bilateral asynchronous hemifacial movements, ipsilateral downward deviation of mouth's angle	Psychogenic facial spasm
	Forceful eyelid contraction, bilateral but can be asymmetric	Blepharospasm
	Sudden, repetitive, non-rhythmic movements, frequently associated with urge to move	Motor tics
	Paroxysmal contraction of one or more of jaw closing muscles	Hemimasticatory spasm
Movement distribution	Bilateral hemifacial spasms	Psychogenic facial spasm, rarely occur in HFS (with a latency of 33 months for the contralateral side to be affected).
	Isolated lower facial spasms Oromandibular dystonia	Psychogenic facial spasm
	Homuncular spreading of tonic-clonic spasms	Epilepsia partialis continua
	Involvement of other body parts	Myokymia
		Myorhythmia
	Temporomandibular joint	Bruxism
Clinical course	Acute onset	Cortical seizures Psychogenic facial spasm
	Rapid progression	Reported cases in demyelination, tumours (e.g. pontine glioma, schwannoma, meningioma, parotid gland tumour, pilocytic astrocytoma, cyst), and vascular lesions (e.g. cavernous angioma, aneurysm, arteriovenous malformation, hemangioma)

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Table 1 (*continued*)

Red flag category	Characteristic/feature	Underlying disorder
Coexisted features	Cerebellar signs and cranial neuropathies	Brainstem lesions
		Cerebellopontine angle lesions
	Somatisation	Psychogenic facial spasm
	Obsessive-compulsive disorder Attention-deficit hyperactivity disorder	Tics or Tourette's syndrome
	Marked facial palsy	Brainstem lesions Facial synkinesis
	Paroxysmal eye deviation with or without nystagmus	Seizures
	Parotid swelling	Parotid tumour
Investigations	MRI showing focal brainstem and/or cerebellar lesions	Various causes of unilateral facial spasms, not HFS
	EMG showing rhythmic bursts of single motor unit potential of doublets, triplets or multiplets at 5–150 Hz	Myokymia
	EEG showing epileptiform discharges	Cortical seizures (Epileptiform discharges may not be evident in subcortical and hemifacial seizures.)

HFS: Hemifacial spasm; EEG: Electroencephalopathy; EMG: Electromyography.

of clinical acumen is needed to identify the characteristic undulating rippling muscles, like tiny snakes wriggling beneath the skin, though they can be confirmed by electromyography demonstrating doublets, triplets, or multiplets firing regularly or irregularly, in rapid succession, at rates of 5–150 Hz [9]. Although not entirely specific, the presence of the 'Babinski-2' sign or 'the Other' Babinski's sign (a paroxysmal synkinesis when orbicularis oculi contracts and the eye closes, the internal part of the frontalis contracts at the same time, the eyebrow rises during occlusion) is supportive of HFS as the most likely diagnosis, followed by post-paralytic facial synkinesis and rarely Brissaud-Sicard syndrome, but excluding blepharospasm [10–12]. As the 'Babinski-2' sign was reported to be present in 34–68% of patients in HFS, neurologists should not rely on this clinical sign alone in the differential diagnosis of UFS [2,12,13].

Table 1 summarises the red flags, expanding the aetiological spectrum of UFS to include other disorders, not limited to HFS, multiple sclerosis and pontine glioma, but also ranging from post-facial palsy synkinesis, hemifacial seizures, brainstem vascular lesions, lesions of the facial nerve in the Faloppi canal and the parotid gland, oromandibular dystonia, blepharospasm, facial tics, hemimasticatory spasm to psychogenic facial spasm [2,6,14–16]. Considering the full spectrum of UFS and looking for red flags not seen in HFS helps avoid a misdiagnosis of HFS, and subsequent initiation of inadequate or incorrect, and often, as in our patient, causal therapy.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jns.2019.116532.

Declaration of Competing Interest

None.

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