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Original article

Diagnosis of primary hemifacial spasm

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ABSTRACT

The diagnosis of primary hemifacial spasm (pHFS), due to a benign compression of the facial motor nerve by a vessel, within or close to its root exit zone, is often made with delay. Misdiagnosis includes psychogenic spasm, tics, facial myokymia or blepharospasm, but in fact post-facial palsy synkinesis (post-paralytic HFS) is the closest clinical condition, because it is limited to the territory of the facial nerve of a single hemiface. The differential diagnosis between these two entities, whose pathophysiological mechanisms are very different, can be made by electroneuromyographic (ENMG) examination. In addition, magnetic resonance imaging (MRI) is essential to show the offending vessel at the origin of pHFS and the absence of other causes of nerve compression. However, the diagnosis cannot be made on MRI basis alone, since a neurovascular conflict can be present in clinically asymptomatic subjects. This article reviews the clinical, MRI, and ENMG features in favour of a pHFS diagnosis as well as the various differential diagnoses of this involuntary facial movement disorder.

1. Clinical diagnosis

Hemifacial spasm (pHFS) is attributed to a benign compression of the facial motor nerve by a vessel, within or close to its root exit zone (REZ) from the brainstem, as evidenced by the successful outcome of surgical microvascular decompression (MVD) [1–4]. The clinical picture is characterized by involuntary tonic and clonic muscle contractions involving one hemiface [5–7]. The spasms can be very brief and localized or more generally occur in bursts, contracting the whole hemiface in a more tonic way, resulting to a disfiguring grimace in severe cases. Diagnosis may be difficult when pHFS is not very active; in outpatient, triggering maneuvers can be helpful in eliciting repetitive facial contractions to differentiate them from normal contractions.

In most patients (up to 80–90% of cases [6,8]), pHFS is initially limited to the zygomatic territory with only periocular muscle contractions (eyelid closure), but progressively spreads thereafter to the territories of the other facial nerve branches, i.e. temporal (frontalis muscle), buccal (orbicularis oris muscle), marginal mandibular (triangularis or mentalis muscle), and even cervical (platysma muscle). At this time, pHFS appears as involuntary, uncontrollable, synkinetic contractions of the hemiface.

Triggering factors of pHFS onset are usually obscure, even if a variety of circumstances can be identified and reported by some patients [6]. At the onset of symptoms, patients are often between 40 and 50 years of age [6,9,10], but a higher average age at onset has been reported in several series (about 52–55 years of age) [8,11–13]. There is a definite female predominance: women-to-men ratio ranges between 1.8 and 2 [8,9,12], but can even reach 3 [13]. Although a few epidemiologic data are available [14], it seems that pHFS is more common in some Asian populations than in Caucasians. In addition, occasional familial cases of pHFS have been described [15–17] with a maximum incidence of 2–3% in large series [8,11,12].

Left-sided pHFS, as the first case reported in the literature by Shültze in 1875, are slightly more frequent than right-sided pHFS [8,12]. Rare cases of bilateral pHFS were also described, the two sides being affected non-synchronously, but with a certain delay [6,18,19].

At least one-third of the patients reported aggravating factors, such as anxiety, stress, fatigue, sleep deprivation, reading, light exposure, chewing, or particular positions of the head [8,10]. The symptoms may persist during sleep [6], but are usually alleviated by relaxation.

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Rare patients may develop ipsilateral facial hypertrophy but never any motor weakness [6]. There are also no objective sensory signs, although pHFS may produce some discomfort or even be considered painful in the most severe cases [20]. However, in extremely rare cases, pHFS coexists with trigeminal neuralgia [6,12], suggesting that both facial and trigeminal nerves are compromised by neurovascular conflicts. This particular clinical syndrome was called "tic convulsif" [21].

On the sensorial grounds, pHFS can be accompanied by tinnitus. Tinnitus may be under the form of "clicking" or "ticking" sound, occurring synchronously with the facial contractions and resulting from tensor tympani or stapedius muscle contraction [22]. Tinnitus associated with hemifacial spasm can be also caused by a neurovascular conflict reaching the cochlear nerve. In this eventuality, a detailed otologic examination including recording of brainstem auditory evoked potentials should help in establishing a differential diagnosis. In addition, pHFS may interfere with vision due to eyelid closure, causing difficulty with reading and driving, and the involuntary orofacial movements may even provoke dysarthria [6].

Although not life threatening, pHFS causes social embarrassment and a major impact on the quality of life [23,24]. Psychiatric comorbidities are not rare. In most cases, the pHFS and the discomfort it causes worsens over time and, unfortunately, it very rarely disappears spontaneously (less than 10% of the cases) [25].

2. Investigations

The diagnosis and assessment of pHFS only require two types of tests: magnetic resonance imaging (MRI) to visualize the neurovascular conflict and electroneuromyography (ENMG) to characterize the type of nerve excitability disorder.

2.1. Magnetic resonance imaging (MRI)

The development of new MRI and angiography techniques has improved the detection of neurovascular conflict, especially at the REZ level, in the context of pHFS [26–28]. The standard reference for the detection of vascular compression is the combination of high resolution 3D-T2-weighted sequences (Constructive Interference Steady State (CISS) and Fast Imaging Employing Steady State Acquisition (FIESTA) thin section MR images) with 3D-time-of-flight (TOF) angiography in case of arterial compression or 3D-T1-weighted gadolinium enhanced sequences in case of venous conflict, including multiplanar fusions and reconstructions (Fig. 1).

The neurovascular conflict can be observed in nearly all cases when present [29]. The offending vessel is usually an arterial loop from the antero-inferior, or postero-inferior cerebellar artery or the vertebral artery (sometimes dolichoectatic) and more rarely a vein (less than 5% of cases). Multiple compression sites, involving two or more vessels, are present in up to 40% of cases. In addition, in a substantial number of cases, the conflict is not observed directly on the facial nerve, even at the REZ, but appears to be at a distance on the lateral face of the brainstem, leading to a false negative diagnosis of conflict.

Finally, in very few cases, HFS is not due to benign vascular conflict but rather results from a tumour in the cerebellopontine angle or a pontine glioma [30], an aneurysm, an arteriovenous malformation [31], or other causes of nerve compression which do not lead to facial palsy but only facial nerve hyperexcitability [6]. Therefore, MRI investigation is compulsory in the context of pHFS to eliminate a specific cause of compression other than benign vascular one and is also required as a preoperative assessment prior to MVD surgery. However, it is important to emphasize that the fact of not seeing a conflict does not exclude its existence and that, on the other hand, a conflict can be present in totally asymptomatic patients.

2.2. Electroneuromyography (ENMG)

In our experience, the overall pattern of ENMG changes is pathognomonic of pHFS, although the abnormalities observed in some tests are similar between pHFS and other forms of HFS [32].

First, in needle electromyography (EMG), facial muscles usually show some spontaneous activity at rest, even in the absence of concomitant spasm, as a feature of nerve hyperexcitability. In contrast, voluntary contraction activity is strictly normal, without any neurogenic sign of muscle denervation. One important point is to study needle EMG in an upper facial muscle (e.g., frontalis muscle) during focal voluntary contraction of a lower facial muscle (e.g., mentalis muscle) and vice-versa for the study of synkinesis. Even if the involuntary spasms are synkinetic, it must be emphasized that usually no definite synkinesis can be confirmed by needle EMG between upper and lower facial muscles in patients with pHFS when triggered by mild focal voluntary contraction. In fact, the literature is very confusing regarding this point, since synkinesis is often described in the clinical picture of pHFS. However, it is generally acknowledged that synkinesis is more commonly observed after Bell's palsy or other types of facial nerve lesion than in pHFS.

Second, in a nerve conduction study to electrical stimulation, it is crucial to perform selective distal stimulation of the temporal branch and then in a second stage of the marginal mandibular branch of the facial nerve involved in the pHFS and to record muscle responses concomitantly in one muscle of each facial nerve branch

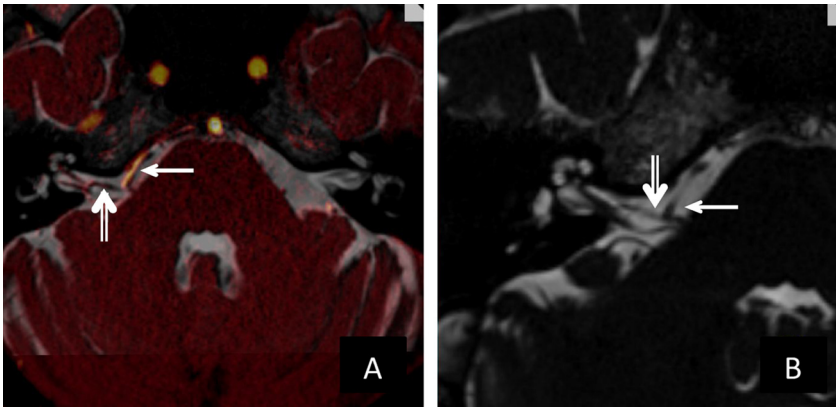

Fig. 1. Right-sided primary hemifacial spasm caused by loop of an antero-inferior cerebellar artery (AICA) (arrow) at level of the pontocerebellar trigone arachnoidal cistern (double arrow), as shown in A (axial reformatted images obtained by fusion of 3D-T2-weighted CISS) and TOF angiography sequences) and B (axial 3D-T2-weighted CISS sequence).

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