Journal of the Neurological Sciences xxx (2015) xxx–xxx

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Review article

Hemifacial spasm: The past, present and future

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ARTICLE INFO

Article history:
Received 12 February 2015
Received in revised form 14 June 2015
Accepted 15 June 2015
Available online xxxx

Keywords:
Hemifacial spasm
Botulinum toxin therapy
Movement disorder
Review
Differential diagnosis
Pathogenesis
Surgical treatment

ABSTRACT

Hemifacial spasm is characterised by unilateral contractions of the facial muscles. Though considered to be benign by many people, it can lead to functional blindness and a poor quality of life due to social embarrassment for the suffering individual. Botulinum toxin therapy is an excellent noninvasive tool to treat this condition. However, surgical decompression of the aberrant vessel is also an upcoming approach to therapy for this condition.

Contents

1. Introduction

Hemifacial spasm (HFS) is a movement disorder of the seventh cranial nerve which is characterised by either brief or persistent, intermittent twitching of the muscles innervated by the facial nerve. The hallmark of the disease is involuntary clonic and/or tonic contractions of the muscles of facial expression, usually unilaterally, beginning in the periorbital musculature, but later on progressing to involve the perioral, platysma and other muscles of facial expression as well.

Although traditionally perceived as a benign illness, it can lead to increasing embarrassment and social withdrawal for the individual

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http://dx.doi.org/10.1016/j.jns.2015.06.032
0022-510X/

and in severe cases even functional blindness due to involuntary eye closure. Thus, prompt diagnosis and timely therapy for the patient are needed in such cases.

2. Historical perspective

F Schultze, in 1875, probably reported the first case of hemifacial spasm in literature, when he described a 56-year-old man with involuntary movements involving the left side of his face [1]. The post-mortem revealed a giant aneurysm of the left vertebral artery compressing the left facial nerve.

In 1886, Gowers elaborated on this syndrome further and described the classical features of this condition [2]. 6 years later, Édouard Brissaud, made similar observations when he described a 35-year-old lady with clonic contractions of the muscles of right half of her face. He observed that these contractions even though present at rest, worsened at times of stress [3].

The condition received its current terminology, when it was named as 'hémispasme facial' by Babinski in 1905 [4]. Babinski at the same time also described another characteristic feature of this disease, thereafter known as 'the other Babinski sign' i.e. when the orbicularis oculi contracts and the eye closes, the internal part of the frontalis contracts at the same time, and the eyebrow rises during eye occlusion. This typical feature distinguishes hemifacial spasm from blepharospasm in which this sign is absent.

3. Epidemiology

The exact prevalence of the disease is difficult to estimate owing to the large number of under and misdiagnosis of this condition. Broadly two forms of hemifacial spasm exist i.e. primary and secondary. Epidemiological studies usually focus on the primary form. A study by Auger and Whisnant from the United States evaluated data of patients from 1960 through 1984 [1]. They observed that the mean prevalence of the disorder was 11 per 100,000 total population, with a 2:1 female preponderance (prevalence rate among Women being 14.5 per 100,000 and men being 7.4 per 100,000 population) [5]. Another study by Nilsen et al. in 2004 from Oslo, Norway gave a similar prevalence of 9.8 per 100,000 [6]. Few studies suggest a slightly higher prevalence of the disease among some Asian populations compared to Caucasians [7,8] however the reason for this high prevalence in this geographical population is not clear.

Primary hemifacial spasm has a wide age range of presentations, but it typically begins in the fifth to sixth decades of life. 1 to 6% of patients present before the age of 30 years [9], however presentation before the age of 40 years should prompt search for an underlying secondary cause of the condition.

Hemifacial spasm is usually sporadic, and familial cases though reported, are rare [10,11]. Bilateral disease is rare (<1%) and even in bilateral cases, the disease starts unilaterally, and after several months to years, begins to involve the other side. The contractions remain asymmetrical in such patients with the side being involved later, typically having less severe manifestations.

4. Pathophysiology

The root exit/entry zone of a nerve is the junction between the central and peripheral nerve segments of a cranial nerve. In this area there is a transition of the cells responsible for myelination of a cranial nerve .i.e. from the central oligodendroglial cells to the peripheral Schwann cells. Also, the cranial nerves in this zone lack an epineurium, being protected by an arachnoid membrane only. This special segment of the nerve thus is highly susceptible to injury [12].

The most common cause for hemifacial spasm reported in literature is an ectatic or aberrant blood vessel, which compresses the facial nerve at this root entry/exit zone leading to local demyelination [13]. Several theories have been put forward to explain how this compression of the facial nerve at its root exit/entry zone leads to hemifacial spasm.

One of them — the nerve origin hypothesis or the peripheral theory postulates that there is ephaptic transmission of impulses between neighbouring neurons (i.e. coupling of adjacent nerve fibres due to local exchange of ions or local electric fields) leading to excessive or abnormal firing. Myelination is a natural inhibitor of ephaptic transmission and the demyelination due to local compression thus leads to hemifacial spasm [11].

The other — the nuclear origin hypothesis or the central theory states that hemifacial spasm results from the hyperexcitability of the facial motor nucleus due to irritative feedback from peripheral lesions of the nerve [14].

5. Etiology

As previously described, hemifacial spasm can either be primary or secondary. Primary HFS results from compression of the seventh nerve at the root exit zone in the posterior cranial fossa by an aberrant or ectatic vessel, most commonly the superior cerebellar, anterior inferior cerebellar or vertebral artery [15].

The list for causes of secondary hemifacial spasm is a long one and includes [16]:

1. Cerebellopontine angle tumours — acoustic neuroma, meningioma
2. Epidermoid, arachnoid cyst, lipoma
3. Arteriovenous malformations — fistulas, venous angiomas and arterial aneurysms
4. Brainstem lesions — stroke, trauma, demyelinating disorders
5. Infections — otitis media, tubercular meningitis
6. Structural abnormalities of the posterior cranial fossa — Paget's disease, Chiari malformation
7. Parotid tumours
8. Bell's palsy

6. Clinical features

Hemifacial spasm classically begins unilaterally in the upper face (around the eyes) — most commonly in the orbicularis oculi muscle (90%) [17], brief repetitive contractions of which lead to sudden, involuntary eye closure. This is typically associated with elevation of the eyebrows — the 'other Babinski sign'. Patients usually complain of a flurry of twitches eventually leading to a sustained spasm.

This presentation is more commonly seen in the primary type of hemifacial spasm, whereas simultaneous involvement of the upper and lower face is more typical for secondary cases [18]. Naraghi et al. believe that this observation has a basis in the anatomy of the facial nerve and nucleus [19]. In the facial nerve, the fibres innervating the upper part of the face are present dorsally, and the anterior inferior cerebellar artery, which is the most common culprit vessel responsible for the syndrome, is most commonly located in relation to the dorsal aspect of the nerve in a majority of patients.

After beginning in the upper half of the face, the contractions gradually spread over time to involve the lower half of face as well i.e. the perioral muscles and eventually the platysma — leading to their irregular clonic or tonic contractions. Unlike most movement disorders, contractions of hemifacial spasm persist during sleep which may add to the morbidity of the condition by predisposing the affected individual to disturbed sleep and insomnia. Other rare findings in patients with hemifacial spasm may include paroxysmal clicking sounds in the ear due to involvement of the stapedius muscle, unilateral or bilateral hearing loss and subtle facial nerve palsy [16]. As classically noted by Brissaud, the symptoms increase during times of stress, reading, speaking and sometimes eating while they abate with relaxation techniques and occasionally touching some parts of the face (sensory tricks).

Please cite this article as: N. Chaudhry, et al., Hemifacial spasm: The past, present and future, J Neurol Sci (2015), http://dx.doi.org/10.1016/j.jns.2015.06.032