Pathophysiology of hemifacial spasm

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Summary

The results of studies in patients undergoing microvascular decompression to relieve hemifacial spasm have provided considerable evidence that the symptoms and signs (spasm and synkinesis) of this disorder are results of changes in the function of the facial motoneuron. Results of animal experiments show that similar signs may develop after chronic electrical stimulation of the facial nerve or after the facial nerve has been injured slightly (by placing a chromic suture around it) and a blood vessel brought into close contact with the nerve. Injury alone or a close contact with a blood vessel alone does not cause the development of such signs.

There is considerable evidence that vascular compression of cranial nerves (V, VII, VIII, IX, and X) occurs frequently without causing any noticeable symptoms or signs of hemifacial spasm. This supports the hypothesis that vascular compression alone does not cause any noticeable symptoms but that another factor is also necessary. It is not known what that other factor may be. It could be a slight injury of the respective nerve or a predisposition in the respective nucleus or both. Since vascular compression seems to be necessary (although not sufficient) to cause symptoms, microvascular decompression is an effective treatment, since the other factor(s) do not seem to cause any noticeable symptoms or signs.

Key words: Animal models, microvascular decompression, facial nucleus, hyperactivity.

Introduction

The pathophysiology of hemifacial spasm (HFS) is not completely understood, despite the fact that extensive experience over the past 20–30 years has shown that HFS can be cured permanently by microvascular decompression (MVD) of the facial nerve where it exits the brainstem [1–8] (and for a recent review see Möller [9]). Reports from several institutions where MVD to relieve HFS is routinely performed show similar results, namely a success rate for cure of HFS between 85 and 95% [9]. In these cases there is no reported evidence of a natural recurrence of HFS, as there is for other disorders that are caused by vascular compression of a cranial nerve root, such as trigeminal neuralgia [10, 11].

Two hypotheses have been proposed to explain the characteristic signs – spasm and synkinesis – of hemifacial spasm [12]. One hypothesis postulates that the symptoms and signs of HFS can be explained by the development of a crosstalk between individual nerve fibers of the facial nerve at the location of the vascular compression [13]. This hypothesis postulates that such ephaptic transmission between denuded motor axons causes the synkinesis as well as the spasm [14, 15]. It is assumed that aberrant neural activity is generated at the location of the cross-compression, which is believed to generate the spasm in patients with HFS.

The other hypothesis, usually associated with work by Ferguson [16], claims that the signs and symptoms of HFS are a result of hyperactivity of the facial motoneuron. It is assumed that this hyperactivity causes both the spasm and the synkinesis, and that it develops as a result of the vascular compression (or irritation) of the root of the facial nerve.

Throughout the past decade considerable research efforts have been devoted to unraveling the pathophysiology of HFS. Studies have been performed in awake patients with HFS [15, 17–21] as well as in patients undergoing MVD operations to relieve HFS [22–30]. More recently, experiments in animals have shown that it is possible to create signs similar to those in patients with HFS by causing an artificial irritation of the facial nerve [31–35]. Thus the abnormal muscle response, synkinesis, and involuntary contractions of facial muscles that are the signs of HFS may develop in animals as a result of a combination of vascular irritation and slight injury to the facial nerve [34, 35] or by chronic electrical stimulation of the facial nerve [31–33].

The results of studies in awake human and in patients who were undergoing MVD operations to re-
lieve HFS as well as the results of experiments in animals have indicated that the pathophysiology of HFS is more complex than was assumed by the original hypotheses. On the basis of such studies, the development of HFS, which usually occurs rather slowly, is assumed by some investigators [22, 28, 36, 37] to depend on neural plasticity, and it has been suggested that a mechanism similar to the kindling phenomenon [38, 39] is involved in the development of HFS [22]. It has also been suggested that the trigeminal system may be involved in the development of HFS [26, 29].

The abnormal muscle contraction has been extensively studied in human as well as in animals, and it has played an important role in studies on the pathophysiology of HFS [15, 18, 22, 26–28]. Thus, the fact that patients with HFS have several measurable signs that are related to HFS makes it possible to perform quantitative studies on the pathological signs of this disorder. Other disorders that can be cured by vascular decompression, such as trigeminal neuralgia (TN), disabling positional vertigo (DPV), and tinnitus lack such signs. The results of studies of HFS may be applicable to other disorders that are caused by vascular compression of a cranial nerve.

**Symptoms and signs of hemifacial spasm**

Involuntary contractions of smaller or larger portions of facial muscles, synkinesis, and the abnormal muscle response are typical signs of HFS [12]. The synkinesis is often demonstrated objectively by recording the blink reflex, but it is particularly the abnormal muscle response that has been used in studies of the pathophysiology of HFS.

The abnormal muscle response

When a branch of the facial nerve in patients with HFS is stimulated electrically, not only does the muscles that are innervated by that branch contract but also do muscles innervated by other branches [15, 22, 40, 41]. The abnormal muscle response can be elicited by electrical stimulation of the temporal branch of the facial nerve and recorded from the mentalis muscle, or it can be elicited from the marginal mandibular branch and recorded from the orbicularis oculi muscles (Fig. 1A). When elicited in either way the abnormal muscle contraction consists of an initial response with a latency of about 10 ms, thus much longer than that of the direct muscle response (M-response) from the muscles that are innervated by the branch of the facial nerve that is being stimulated electrically. This initial component of the abnormal muscle response is followed by a burst of EMG activity, the duration of which typically varies from time to time (Fig. 1B). The abnormal muscle response (also

![Diagram of abnormal muscle response](image-url)