The other Babinski sign: a brief review of hemifacial spasm

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Joseph Babinski is famous for his 1896 description of the Babinski sign, an abnormal plantar reflex associated with dysfunction of the pyramidal system. He is less well-known for his “other” Babinski sign: paradoxical raising of the eyebrow associated with closure of the eye, a feature typical of hemifacial spasm.

As Babinski noted in 1905, hemifacial spasm (HFS) is a disorder of painless, involuntary facial twitches confined to the muscles supplied by the facial nerve. While benign, it causes significant morbidity due to excessive closure of one eye, and the social effects of an abnormal appearance. Unfortunately, facial twitches are often attributed to stress or anxiety, leading to a delay in diagnosis. A vigilant search for organic causes is essential when these symptoms arise, as highly efficacious treatment options for HFS are available and can significantly improve quality of life.

HFS occurs more commonly in females (2:1) and has an overall prevalence of approximately 10/100,000. The orbicularis oculi is the site of onset in the large majority of cases. Over months to years, the disease spreads to other muscles. Spasms are described as clonic or tonic, are aggravated by stress, fatigue and anxiety, and persist during sleep. Disease is usually unilateral, and spasms are synchronous in all affected ipsilateral muscles. Bilateral disease occurs in a small minority of cases, and spasms are never synchronous bilaterally.

In most cases, hemifacial spasm is due to arterial compression of the root exit zone (REZ) of the facial nerve. This is similar to the proposed mechanism of trigeminal neuralgia, in which the trigeminal nerve is compressed in the REZ, causing demyelination and recurrent episodes of intense pain. In much the same way, compression of the facial nerve causes spontaneous irregular discharges, leading to involuntary muscle contractions. As well, non-synaptic neural transmission (ephespsis) between branches of CN VII may occur, causing synkinesis.

Most commonly, the aberrant artery is the posterior-inferior cerebellar artery (PICA), the antero-inferior cerebellar artery (AICA), or the vertebo-basilar artery (VB). In one series, these vessels were responsible 61%, 56% and 27% of the time, respectively. The same study found that multiple vessels were responsible 40% of the time. However, up to 25% of the population has vascular loops compressing CN VII, suggesting that this alone is insufficient to produce HFS. Vascular loop decompression surgery via the posterior cranial fossa is very effective in providing relief from spasms. For some patients, relief is almost immediate, while for others it can take up to a year following surgery to see the full effects. This delayed cure suggests that HFS is not only due to mechanical compression. Demyelination of the nerve from the brainstem and hyper-excitability of the facial nerve nucleus due to irritation from neurovascular compression may play a role. The slow reversal of these plastic changes explains the delay.

Several other diagnoses can present in a similar fashion, including benign essential blepharospasm (BEB), focal motor seizures, and craniofacial tremor. These are differentiated largely based on clinical presentation, so a careful history and physical exam are essential. For example, BEB often presents with spasms of the orbicularis oculi and adjacent muscles, which may be difficult to distinguish from HFS. However, BEB is often associated with eye irritation, vague ocular pain and photophobia. If the facial spasms are associated with body tremor, or a family history, craniofacial tremor is more likely. Focal motor seizures are implicated when post-ictal weakness and a greater involvement of the lower face are present. If the clinical presentation is ambiguous, electrophysiologic studies and imaging can aid in the diagnosis.

The characteristic sign of HFS on electrophysiologic studies is a response in distant facial nerve branches following a stimulus to a single branch. This can be seen, for example, by spread of the blink reflex on stimulation of the supraorbital nerve to muscles other than orbicularis oculi. This is referred to as lateral spread response (LSR) and is attributed to spread of excitation at the site of ephapsis.

Rarely, HFS is caused by tumors, arteriovenous malformations, and venous compression. Because the risk of tumor is low, imaging may not be cost effective when the diagnosis is clear. However, a careful neurologic exam for focal signs is essential, and imaging is suggested if these are present or if conservative management fails. Advanced MRI techniques, such as constructive interference in steady state, are effective at identifying neurovascular contact. Imaging may also assist in surgical planning.

While spontaneous remission of symptoms is possible, it occurs in less than 10% of patients, and significant effects on quality of life usually demand intervention. Several oral medications have been studied, including carbamazepine, anticholinergics, baclofen, clonazepam and haloperidol. However, sample sizes have been small and it is difficult to confidently interpret the results. The effects of oral medications are usually transient, and unacceptable side effects often occur. For many patients, this is not a viable treatment option.

In contrast, many physicians consider botulism toxin (BTX) to be the treatment of choice. While few double-blind randomized studies evaluating BTX for HFS exist, the available evidence is encouraging. Park et al described an open-label study of 101 HFS patients, 98.6% of which had excellent results after injection of botulism-A. The mean duration of effect was 16.5 weeks, with a range of 11-40 weeks. In a review of the HFS-BTX literature, Jost et al found a total of 37 open case-control studies (N=2295) in...
which good to excellent results were reported in 76-100% of patients.\textsuperscript{11} Adverse effects include facial droop, ptosis, diplopia, lid edema and ecchymosis,\textsuperscript{12} with facial weakness being the most common.\textsuperscript{1}

Micrvascular decompression (MVD) of the facial nerve is the only truly curative option. It involves freeing the nerve from the compressing vessel(s) at the cerebellopontine angle. The offending vessel is gently mobilized away from the facial nerve and a small, rectangular piece of Teflon sponge is then interposed between the REZ on one side and the aberrant vessel(s) on the other.\textsuperscript{8}

While MVD has a high success rate (> 90% in some series), the recurrence rate remains up to 20%.\textsuperscript{2} Furthermore, the potential risks are significant, including facial nerve dysfunction, lower cranial nerve dysfunction, intracranial infections, cerebrospinal fluid leaks, and temporary or permanent damage to CN VIII.\textsuperscript{2} According to some reports, 7-26% of patients will suffer a degree of temporary or permanent hearing loss.\textsuperscript{2} A large study of 668 patients by Park \textit{et al} found that 2.2% of patients experienced permanent hearing loss, with 0.7% of these suffering total deafness.\textsuperscript{14}

According to several studies, monitoring of brainstem auditory evoked potentials (BAEP) reduces the risk of hearing loss complications.\textsuperscript{8} BAEP monitoring allows surgeons to watch for the increased latency of peak V and decreased amplitude of peak I, which are the early warning signs of excessive stretching of the cochlear nerve and impairment of the cochlear vascular supply, respectively.\textsuperscript{8} This allows dangerous maneuvers to be stopped or corrected, and appears to significantly reduce the risk of postoperative hearing loss.\textsuperscript{8}

Intraoperative monitoring of lateral spread response (LSR) is more controversial. The goal is to see disappearance of the LSR when the aberrant vessel is moved away from the facial nerve.\textsuperscript{4} This can augment surgical decision-making, and some authors suggest that it leads to improved outcomes.\textsuperscript{15} In 1987, Moller and Jannetta advocated routine intraoperative monitoring of LSR, contending that this would ensure adequate decompression. If LSR was observed at the end of the procedure, they reasoned, symptoms were likely to persist.\textsuperscript{8} Therefore, if the LSR does not disappear following decompression of the most obvious site, a search for a second site can be undertaken.\textsuperscript{7} In a study by Mooij \textit{et al}, a second compression site was found in the majority of these cases.\textsuperscript{16} Several other authors advocate for LSR monitoring during MVD, including Sekula \textit{et al}, who found that the probability of cure was 4.2 times greater if LSR was abolished during surgery than if it persisted.\textsuperscript{15}

Conversely, a number of studies have questioned the value of LSR monitoring. Joo \textit{et al} examined 32 patients who had continuing LSR after MVD. Of these, 21 did not display HFS at 1-week follow-up. The authors argued that demyelination of the REZ may lead to a delay in normalization of electrophysiologic muscle responses, even when decompression is effective.\textsuperscript{15} Similarly, Sindou examined outcomes in the longer term and found good results even in patients with persistent LSR at the end of MVD.\textsuperscript{7} One third of patients assessed became spasm free only after a significant interval (4 to 12 months). Since delayed effects of MVD are common, he argued, final assessment of outcome should not be made until 1 year post-operative.\textsuperscript{7} Neves \textit{et al} noted that, in addition to delayed success, HFS can also recur after an MVD that was successful in terms of LSR disappearance. They found that intraoperative LSR changes did not correlate with HFS relief on the first post-operative day.\textsuperscript{7} However, they did find a correlation between absence of LSR at the end of surgery and the long-term efficacy of the MVD.\textsuperscript{7} Whether elimination of LSR at the time of surgery predicts a good clinical outcome remains unclear.\textsuperscript{4}

Involuntary facial movements are often attributed to stress and disregarded. An awareness of HFS and its presentation is essential, as HFS can result in severe social and functional disability\textsuperscript{4} and highly efficacious treatment options exist. However, the benign nature of this disorder requires that these treatment options be carefully weighed. No treatment is without risk, but MVD in particular is an invasive solution that carries a risk of significant disability. A careful risk-benefit analysis must be undertaken in order to balance the potential of symptom relief with the costs of therapy.

References

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