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Case Report

Nonspastic hemifacial spasm confirmed by abnormal muscle responses

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Abstract

Background: Hemifacial spasm is usually diagnosed by inspection which mainly identifies involuntary movements of orbicularis oculi. Assessing abnormal muscle responses (AMR) is another diagnostic method.

Case Description: We report a case of left hemifacial spasm without detectable involuntary facial movements. The patient was a 48-year-old man with a long history of subjective left facial twitching. On magnetic resonance imaging (MRI), the left VIIth cranial nerve was compressed by the left anterior inferior cerebellar artery (AICA), which was in turn compressed by the left vertebral artery. We initially treated him with botulinum toxin. We were able to record AMR, and hemifacial spasm occurred after AMR stimulation, although no spasm was detectable by inspection. Subsequently, we performed microvascular decompression with transposition of the AICA that compressed the VIIth cranial nerve. His hemifacial spasm resolved by 5 weeks after surgery and was not induced by AMR stimulation.

Conclusion: Hemifacial spasm can sometimes be diagnosed by detecting AMR rather than by visual inspection. We propose that such hemifacial spasm should be termed nonspastic hemifacial spasm.

Key Words: Abnormal muscle response, hemifacial spasm, nonspastic hemifacial spasm

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INTRODUCTION

Hemifacial spasm (HFS) is defined as involuntary, irregular, clonic or tonic contraction of muscles innervated by the VIIth cranial nerve. It usually commences as twitching of the lower eyelid, followed by involvement of other periorbital, facial, and perioral muscles, as well as platysma, often leading to social embarrassment and/or interference with vision due to involuntary eye movements.[7] HFS is sometimes confused with other

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facial movement disorders, such as blepharospasm, facial tic, myokymia, and aberrant regeneration with synkinesis after Bell's palsy.[7] Here, we report a case of HFS with no abnormal movements detectable visually, although the patient felt twitching of the left face.

CASE REPORT

A 48-year-old man complained of a twitching feeling on the left side of the face for more than 3 years. He had been treated with botulinum toxin (BTX), although there were no clear symptoms of HFS. He was then referred to our outpatient clinic for diagnosis and further treatment. He complained of twitching that mainly affected the infraorbital region of the left face, however, HFS was not apparent by inspection. Magnetic resonance imaging (MRI) with fast imaging employing steady-state acquisition (FIESTA) showed that the left anterior inferior cerebellar artery (AICA) compressed the root exit zone (REZ) of the left VIIth nerve [Figure 1a]. At his request, we repeated treatment with BTX five times at the region where he had a twitching sensation because he noted disappearance of the twitching sensation after treatment. We decided to record abnormal muscle responses (AMR) for differential diagnosis of HFS. AMR were recorded from the orbitalis oculi and mentalis muscles using surface electrodes (located 1 cm apart) placed on cleaned and degreased skin over the muscles, with the stimulus intensity adjusted to that of the frontalis muscle. With stimulation at 6.5 mA, we obtained clear AMR at 15 ms, and HFS was induced which lasted for approximately 1 minute after stimulation [Figure 2a].

The patient eventually agreed to surgical treatment due to severe discomfort. Microvascular decompression (MVD) was performed via left retrosigmoid lateral suboccipital craniotomy with intraoperative monitoring of AMR. Electromyograms (EMG) were recorded directly from the muscles with needle electrodes under intravenous general anesthesia using propofol and fentanyl. The AMR detected at 20 ms [Figure 2b] disappeared just after transposition of the AICA to relieve firm compression of the VIIth nerve REZ [Figure 2c]. Twitching completely subsided by 4 weeks after MVD, and AMR were not recorded at 5 weeks postoperatively. Decompression of the REZ after transposition of the AICA was confirmed by MRI FIESTA [Figure 1b]. He was satisfied by the complete improvement of symptoms without adverse effects of MVD.

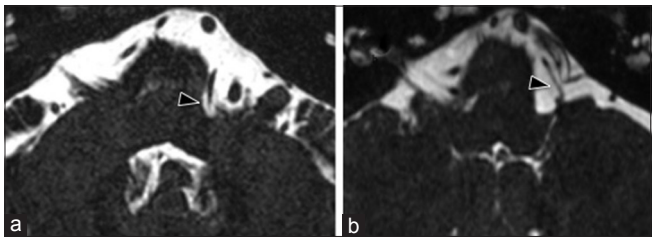

Figure 1: Root exit zone compression by the anterior inferior cerebellar artery (AICA, arrowhead) was seen on axial MRI (FIESTA) (a) and was released by transposition of the AICA (b)

DISCUSSION

HFS is defined as involuntary, irregular, clonic or tonic contraction of muscles innervated by the VIIth cranial nerve. Symptoms usually start with twitching of the lower eyelid, which is followed by involvement of other periorbital, facial, perioral, and cervical muscles, often leading to social embarrassment, and interference with vision by involuntary eye movements.[7] HFS can be mistaken for other facial movement disorders, including blepharospasm, facial tic, myokymia, and aberrant regeneration with synkinesis after Bell's palsy.[7] Our patient with HFS did not have evident facial muscle involvement, but the condition manifested as a twitching feeling in the left side of the face. If we had used a needle electrode for examination, a spontaneous EMG might have been obtained, but we did not because this is an invasive examination, and it was considered difficult to detect the spontaneous EMG with such weak twitching.

Two hypotheses for the electrophysiological mechanism of facial spasm have been proposed: (1) compression of the facial nerve by a blood vessel injures the myelin sheath and facilitates ephaptic transmission between nerve fibers to increase spontaneous activity; or (2) spasm is induced by hyperexcitability of the facial motor nucleus itself.[3-6] Our observation that stimulation of the facial nerve caused AMR and typical HFS in this patient may support the latter hypothesis.

We propose two possible mechanisms as the cause of atypical HFS. The facial muscles may receive very weak stimulation by current leaking from nerve fibers with demyelination. If nerve fibers undergo demyelination, the action potential is reduced because the intermodal density of sodium channels is too low for action potential propagation in demyelinated axons.[1] Therefore, the current increases when resistance is decreased to maintain the action potential and the current could easily and

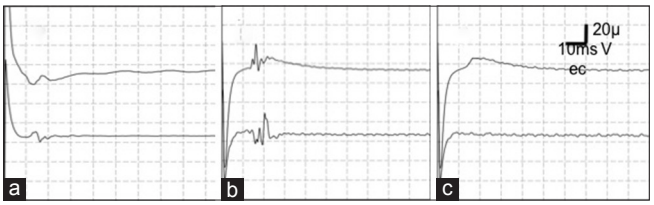

Figure 2: Abnormal muscle responses (AMR) were recorded from the orbicularis oculi muscle (upper) and mentalis muscle (lower) at the outpatient clinic (a) and during surgery (b) with 6.5 mA stimulation. AMR disappeared completely after microvascular decompression (c). AMR are not M-responses at 2.9 ms but F-responses at 10–15 ms after stimulation of the nerve to the ipsilateral frontalis muscle

rapidly leak from such damaged nerve fibers. Alternatively, demyelinated axons may generate central and peripheral currents in response to vascular compression, resulting in an excitatory evoked potential from the central current. This potential is an efferent current, and is attenuated through interference with the central current. The next factor to consider is muscle characteristics. In the limb muscles, myofibers receive mononeuronal innervation, however, the myofibers of facial muscles receive polyneuronal innervation.[2] Movements of facial muscles are relatively slow and weak compared with those of limb muscles. The facial muscles contain two types of muscle fibers (type I and II), which are slow and fast twitch fibers, respectively.[7] The orbicularis oculi muscle is largely formed of fast twitch fibers (90% type II fibers), whereas the mentalis and orbicularis oris muscles have a fast: slow twitch fiber ratio of approximately 2:1.[2] Differences due to the various combinations of type I and II fibers in the facial muscles may influence the symptoms of HFS.

When we encounter a patient who complains of facial twitching but cannot detect typical HFS by visual inspection, we should remember that the present case might be rare, but such cases can occur. Even if HFS cannot be detected, if MRI apparently shows vascular compression of the facial nerve, we need to check whether AMR and HFS are induced by stimulation. If AMR and HFS are induced, the patient should be treated with BTX or should have surgery if the symptoms are severe enough to be problematic for daily life.

CONCLUSION

In conclusion, we reported a patient who had with HFS without obvious involuntary facial movements. The diagnosis was based on electrophysiological studies and MRI, and symptoms were cured by MVD. We propose that such atypical HFS should be known as "nonspastic hemifacial spasm."

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Conflicts of interest
There are no conflicts of interest.

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