AAEM CASE REPORT #21

A 75-year-old man developed progressive involuntary hemifacial spasm. Electrophysiologic evidence of abnormal cross-transmission between neurons of the facial nerve was demonstrated. Electrodiagnostic studies were used to confirm the diagnosis preoperatively and determine the adequacy of vascular decompression of the facial nerve intraoperatively.
Key words: hemifacial spasm • nerve conduction studies, hemifacial spasm • craniectomy

MUSCLE & NERVE 14:213–218 1991

AAEM CASE REPORT #21: HEMIFACIAL SPASM: PREOPERATIVE DIAGNOSIS AND INTRAOPERATIVE MANAGEMENT

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Hemifacial spasm is a chronic and often progressive disorder characterized by unilateral irregular clonic and tonic contraction of one or more muscles of facial expression. The condition may result from chronic compression of, or injury to, the facial nerve.[21–23] The majority of patients with disabling "idiopathic" hemifacial spasm respond to surgery designed to detect and relieve vascular compression of the facial nerve at its exit from the brainstem.[3,13,16,20,26] Electrodiagnostic studies help distinguish hemifacial spasm from other involuntary movements of the face and may help determine when the facial nerve has been adequately decompressed intraoperatively.

CASE REPORT

Clinical History. A 75-year-old man presented for evaluation of a two-year history of progressive unilateral twitching of the left face. The involuntary facial movements began as mild intermittent brief contractions near the corner of the mouth, gradually progressing to a mixture of brief irregular clonic and more prolonged tonic contractions of all the muscles of facial expression. The movements were worse with emotional stress and persisted during sleep. He denied any sensory disturbance or weakness of the face. There was a long-standing history of partial bilateral hearing loss. Previous treatment with phenytoin and carbamazepine failed to improve the facial movements.

Physical Examination. Abnormalities were limited to frequent irregular clonic contractions and intermittent tonic spasms of the orbicularis oculi and oris, frontalis, buccal, and mentalis muscles. There was no objective weakness of facial muscles or abnormality of facial sensation. Corneal reflexes were normal with no obvious synkinesis noted.

Laboratory Tests. Computerized tomography of the head with contrast showed ectatic dilatation of the upper portion of the left vertebral artery. The intracranial contents were otherwise normal. Transfemoral cerebral angiography confirmed the marked ectasia and tortuousity of the left vertebral artery causing it to swing laterally into the cerebellopontine angle.

ELECTRODIAGNOSTIC EVALUATION

Methods. Facial nerve conduction studies and blink reflexes were performed using standard techniques with percutaneous stimulation and surface recording electrodes. The facial M wave was recorded over the ipsilateral nasalis muscle referenced to an electrode over the contralateral nasalis during stimulation with the cathode just inferior to the stylomastoid foramen. Blink reflexes were

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Accepted for publication April 6, 1990.

CCC 0148–639X/91/030213–06 $04.00

performed with stimulation of the supraorbital nerve recording over the orbicularis oculi muscles bilaterally. Simultaneous recordings were made over the ipsilateral mentalis muscle for the detection of synkinesis. The "lateral spread response" described by Nielsen[21,22] and others[18,19] was recorded preoperatively with surface electrodes over the orbicularis oculi and mentalis muscles during ipsilateral percutaneous stimulation of the mandibular branch of the facial nerve 5 cm proximal to the mentalis recording electrode (cathode proximal, see Fig. 1).

Although not done in this case, the lateral spread response can also be recorded from the mentalis muscle with stimulation of the zygomatic branch of the facial nerve 5 cm proximal to the orbicularis oculi electrode (Fig. 1). The normal range in latency of the lateral spread response with either technique is 7 to 11 ms. A well-defined response can usually be obtained with relatively low stimulus intensities. Supramaximal stimuli may cause the recording to be contaminated by volume-conducted potentials generated by the masseter or facial muscles. Some patients with mild hemifacial spasm will have a lateral spread response with either zygomatic or mandibular stimulation, but not both. Therefore, if a lateral spread response cannot be elicited with one technique, the other should always be performed. Needle electromyography was performed with a standard concentric needle electrode.

Recordings of spontaneous EMG activity in facial muscles, the lateral spread response, and ipsilateral brainstem auditory evoked potentials were performed during suboccipital craniectomy and vascular decompression of the facial nerve. Pairs of fine nichrome wire electrodes were placed in the orbicularis oculi and oris muscles. Spontaneous EMG activity was monitored continuously at standard gains, sweep speeds, and filter settings for needle electromyography. The same intramuscular electrodes in the orbicularis oculi muscle were used to record the lateral spread response. The mandibular branch of the facial nerve was stimulated with 2 platinum needle electrodes placed after induction of anesthesia. Neuromuscular blocking agents were excluded from the anesthetic regimen.

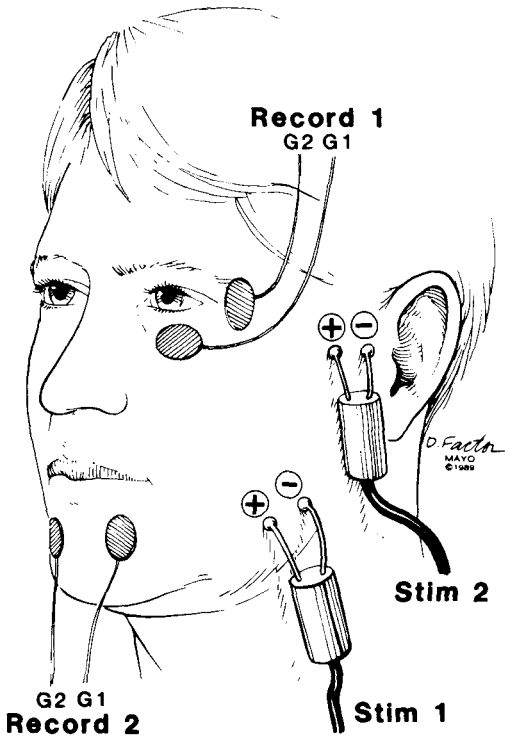

FIGURE 1. Technique for recording the lateral spread response from the orbicularis oculi muscle (Record 1) with stimulation of the mandibular branch of the facial nerve (Stim 1) or from the mentalis muscle (Record 2) with stimulation of the zygomatic branch of the facial nerve (Stim 2). The mandibular branch is stimulated with the cathode 5 cm proximal to the mentalis electrode. The zygomatic branch is stimulated with the cathode 5 cm proximal to the orbicularis electrode (G1) along the zygomatic arch.

PREOPERATIVE STUDIES

Nerve Conduction Studies. The results of facial nerve conduction studies and blink reflexes are illustrated in Table 1. Facial nerve conduction studies were normal. The only abnormality of the blink reflex was a synkinetic response of the left mentalis muscle with ipsilateral stimulation of the supraorbital nerve (Fig. 2). A lateral spread response with an approximate latency of 10 ms was recorded on the left (Fig. 3). No synkinesis or lateral spread response could be recorded on the right.

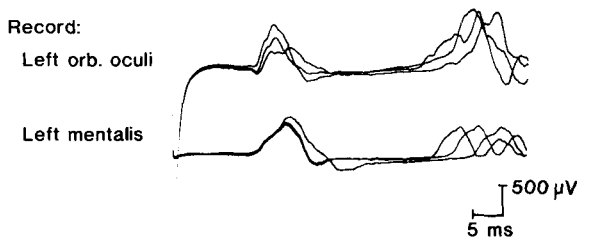

FIGURE 2. Blink reflex demonstrating synkinesis of the orbicularis oculi and mentalis muscles to stimulation of the left-supraorbital nerve.

Table 1. Preoperative nerve conduction studies.

Nerve	Record	Amplitudes (mV)		Distal latency (ms)
Right facial, motor	Nasalis	1.6		2.5
Left facial, motor	Nasalis	1.9		3.0
		Ipsilateral (ms)		Contralateral (ms)
		R1	R2	R2
Right trigeminal, blink	Orbicularis oculi	11.3	27	31
Left trigeminal, blink	Orbicularis oculi	11.3	29	30

Needle Examination. The left and right orbicularis oculi and oris muscles and the left masseter and mentalis muscles were examined. Abnormalities were confined to irregular brief high-frequency bursts of motor unit potentials recorded in the left orbicularis oculi and oris muscles (Fig. 4). No fibrillation potentials or abnormalities of motor unit potential amplitude, duration, recruitment, or morphology were noted.

Interpretation. The electrodiagnostic studies showed synkinesis of the left blink reflex, a lateral spread response from one branch of the facial nerve to another, and high-frequency motor unit potential discharges in facial muscles. In the context of involuntary facial movements, this constellation of findings is diagnostic of hemifacial spasm.

CLINICAL COURSE

The patient underwet a left suboccipital craniectomy with continuous intraoperative monitoring of spontaneous EMG activity in facial muscles and brainstem auditory evoked potentials. The lateral spread response of the left orbicularis oculi muscle to stimulation of the mandibular branch of the left facial nerve was recorded at 10-minute intervals before and continuously during vascular decompression of the facial nerve. At surgery, the posterior inferior cerebellar artery was clearly compressing the inferior aspect of the facial nerve at its exit from the brainstem. When the vessel was removed from the nerve, the lateral spread response disappeared only to reappear when the vessel was allowed to fall back in contact with the nerve (Fig. 5). The vessel was then permanently displaced from the nerve with Gelfoam and muscle. The lateral spread response could not be detected during the remainder of the procedure.

Manipulation of the facial nerve intraoperatively produced high-frequency "neurotonic" discharges in facial muscles which provided the surgeon immediate warning of potential iatrogenic injury to the facial nerve (Fig. 6). Brainstem auditory evoked potentials remained stable during the procedure indicating that the integrity of the cochlear nerve was being preserved. All involuntary facial movements were absent postoperatively with no facial weakness. The patient has remained symptom-free over 3 months of follow-up.

DISCUSSION

Hemifacial spasm was initially described by Gowers in 1884.[10] It typically begins in the fifth or sixth decade as mild unilateral irregular involuntary twitching of the orbicularis oculi. With time, there is progressive involvement of other facial muscles and brief clonic twitching may be mixed with or replaced by sustained tonic contractions of several muscles. Mild facial weakness may occur

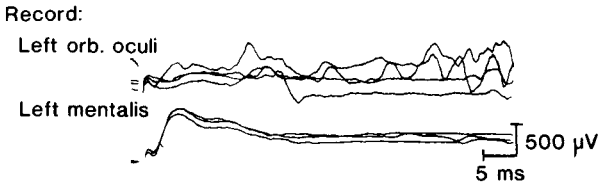

FIGURE 3. Lateral spread response of left orbicularis oculi muscles to stimulation of ipsilateral mandibular branch VII of facial nerve.

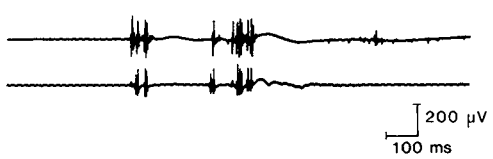

FIGURE 4. Needle electromyography of orbicularis oculi (top) and oris muscles (lower) demonstrating high-frequency discharges of motor unit potentials concurrent with visible contraction of facial muscles.

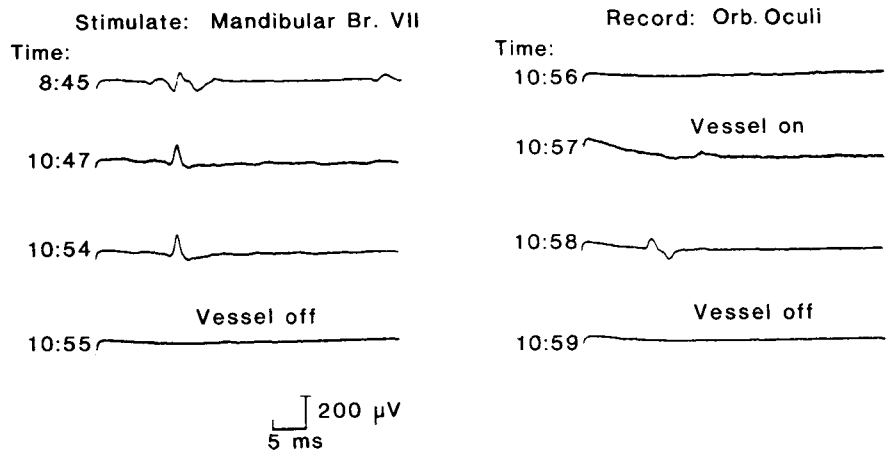

FIGURE 5. Intraoperative recording of the lateral spread response during microvascular decompression of the facial nerve. Stimulation of the mandibular branch of the facial nerve produced a CMAP in ipsilateral orbicularis oculi muscle. The response disappeared with adequate vascular decompression of the facial nerve.

but no other neurologic deficits are usually found. Voluntary movements may be associated with synkinetic contraction of muscles innervated by different branches of the facial nerve. The involuntary movements are typically worsened by emotional stress and persist during sleep.

Hemifacial spasm has been reported in patients with intra-axial brainstem lesions,[25] tumors of the posterior fossa[28] and temporal bone,[7] parotid tumors,[8] basilar meningitis,[30] arteriovenous malformations,[17] and Paget's disease.[6] The common denominator in all reported cases is a lesion that causes chronic compression of, or partial injury to, the facial nerve. In most patients, there is no obvious compressive lesion identified on imaging studies. Evidence accumulated from a variety of sources suggests that the majority of idiopathic cases are caused by vascular compression of the facial nerve within the cerebellopontine angle.

Pathologic studies in cases with hemifacial spasm and apparent vascular compression of the facial nerve at its exit from the brainstem have shown focal demyelination and narrowing of axons at the site of compression.[14,15,29] These alterations may create changes in the neurophysiology of conduction that lead to ectopic generation of action potentials.[24,27] These ectopic impulses may cause hemifacial spasm by ephaptic transmission to other axons at the site of compression[21] or by centripetal transmission of action potentials leading to hyperexcitability of the facial nucleus.[19] A variety of arguments in support of both theories have been proposed.[18,24] Although the experimental data available can be explained by either theory, the demonstration of slowed conduction in the pre-ephaptic segment,[21] and the frequency of the spontaneous discharges recorded in hemifacial spasm lend support to the ephaptic transmission theory.

FIGURE 6. Neurotonic discharges recorded from intramuscular electrodes in the orbicularis oculi muscle produced by inadvertent mechanical stimulation of the facial nerve during vascular decompression.

In 1984, Nielsen described an electrophysiologic manifestation of abnormal communication between neurons of the facial nerve in patients with hemifacial spasm. Stimulation of one branch of the facial nerve consistently produced a compound muscle action potential (CMAP) in a muscle innervated by a different branch of the facial nerve. This "lateral spread response" could only be recorded on the side of the spasm and disappeared postoperatively in patients successfully treated by vascular decompression of the facial nerve.[23]

In addition, Auger[2] and others[22] described the synkinetic spread of the blink reflex from one branch of the facial nerve to another in hemifacial spasm. The synkinesis in most patients with hemifacial spasm was found to be different from that observed in patients with Bell's palsy.[2] In hemifacial spasm, the synkinetic response was frequently

inconstant in its presence and latency, whereas in Bell's palsy, it was consistently elicited with each stimulus and occurred at a more constant latency. Therefore, the variable nature of the synkinetic response in hemifacial spasm, as well as its disappearance following vascular decompression, help distinguish it from synkinesis associated with aberrent reinnervation of the facial nerve. Needle examination of muscles involved with the hemifacial spasm typically shows irregular bursts of motor unit potentials with intraburst firing frequencies of 80 to 150 Hz. The motor unit amplitude potential, duration, and morphology are usually normal in patients with idiopathic hemifacial spasm.

The differential diagnosis of involuntary facial movements includes myokymia, synkinesis following facial neuropathy, essential blepharospasm, facial tic and partial seizures. The electrodiagnostic evaluation can provide information essential to the differential diagnosis of hemifacial spasm. The presence of characteristic high-frequency discharges on needle examination, in association with the lateral spread response and variable synkinesis of the blink reflex, represent the electrophysiologic hallmark of hemifacial spasm.

The appropriate treatment of hemifacial spasm depends on the severity of the spasm and the general medical condition of the patient. Mild cases may improve with carbamezapine[1] or other medications designed to reduce neuronal excitability.[12,31] Local injection of botulinum toxin is particularly successful in cases with spasm of the orbicularis oculi muscle.[9] Side effects are usually minimal although ptosis and diplopia have been reported.[5] The major disadvantage is the need for repeated injections every 3 to 4 months. Suboccipital craniectomy with microvascular decompression of the facial nerve has become the surgical treatment of choice for patients with moderate to severe spasm. The results of several large clinical series suggest that spasm is eliminted completely in 80% to 85% of patients with 90% obtaining at least partial relief.[3,20,26]

Moller and Jannetta recorded the lateral spread response intraoperatively in a large group of patients undergoing vascular decompression for hemifacial spasm.[20] In many patients, they observed a striking relationship between the physical contact of the offending vessel with the facial nerve, and the ability to record the lateral spread response. Frequently, this response disappeared within seconds of removing the vessel only to reappear immediately when the vessel was allowed to recontact the nerve. Hemifacial spasm was completely eliminated in 95% of patients whose lateral spread response was eliminted intraoperatively. In contrast, 4 of 7 patients who had no change in the lateral spread response intraoperatively had persistent spasm severe enough to require reoperation. Therefore, it appears that monitoring the lateral spread response intraoperatively can provide valuable information regarding the completeness of the vascular decompression.

The reason for the immediate elimination of the lateral spread response with vascular decompression is unknown. With pathologic studies showing focal demyelination at the site of compression, one might expect a delay in the resolution of the lateral spread response and clinical spasm that correlated with healing of the pathologic lesion. It is clear from intraoperative monitoring during posterior fossa surgery, however, that the facial and other cranial nerves are very sensitive to mechanical stimulation, and that the motor unit potential discharges observed have firing patterns very similar to those in hemifacial spasm.[11] Mechanical irritation of the facial nerve by a pulsating vessel may provide the stimulus necessary to produce hemifacial spasm when superimposed on a predisposing demyelinating lesion.

Although vascular decompression is an effective treatment for hemifacial spasm, complications have been reported in up to 35% of patients.[3] Most complications are mild and transient; however, permanent hearing loss and facial weakness have a reported incidence of 15% and 4%, respectively.[3] Intraoperative monitoring of auditory and facial nerve function may help reduce the frequency of these complications. Auditory function can be monitored with brainstem auditory evoked potentials, electrocochleography, or recording of nerve action potentials directly from the eighth cranial nerve during surgery. The facial nerve is best monitored by recording spontaneous electromyographic activity with intramuscular wires in several facial muscles. Mechanical manipulation of the facial nerve during surgery produces characteristic high-frequency bursts or trains of motor unit potentials. These neurotonic discharges provide an immediate signal to the surgeon warning of potential injury to the facial nerve. These techniques have been shown to help reduce the incidence of facial nerve injury in patients undergoing resection of acoustic neuromas.[11]

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