AANEM Case Report # 26
Seventh Cranial Neuropathy

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American Association of Neuromuscular &
Electrodiagnostic Medicine

AAEM CASE REPORT #26: SEVENTH CRANIAL NEUROPATHY

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CME STUDY GUIDE

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CERTIFYING ORGANIZATION

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EDUCATIONAL OBJECTIVES

The purpose of this case report is to help the electromyographer understand and utilize electrodiagnostic techniques in developing a diagnosis and prognosis in disorders of the seventh cranial nerve. The techniques of facial motor nerve conduction study, blink reflex and nerve excitability will be reviewed.

INSTRUCTIONS

1. The reader should carefully and thoroughly study this case report. If further clarification is needed, the references should be consulted. Do not neglect illustrative material.

2. Read the CME questions at the end of the case report. Choose the correct answer to each question and record it on the CME Registration form on the last page. Retain a copy of your answers for your records.

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6. Review those parts of the article dealing with the question(s) you answered incorrectly, and read the supplemental materials on this aspect of the subject listed in the references.

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The seventh cranial nerve is affected by a number of different pathologic entities, and is often the initial sign of neurologic disease. The resulting facial weakness is always a major concern to patients and provokes early medical consultation as to the cause, treatment, and prognosis. A seventh cranial neuropathy can be distinguished from a "central" facial palsy by the lack of ipsilateral involvement of the frontalis muscle in the latter, due to bilateral supranuclear innervation of the frontalis. The seventh cranial neuropathy is commonly referred to as Bell's palsy, after Sir Charles Bell who described facial palsy in 1829.[2] Hyperacusis from involvement of the stapedial branch, abnormal taste from involvement of the chorda tympani branch, and abnormalities of lacrimation and salivation may be other findings in facial neuropathy. After exiting the pons, the seventh cranial nerve traverses the cerebellopontine angle and enters the internal auditory meatus, then courses along the facial canal before taking a 90-degree turn near the middle ear and exiting the skull at the stylomastoid foramen. It passes through the parotid gland during which, and shortly after, it divides into individual muscle branches. This long course exposes the facial nerve to any number of pathologic entities but, along with its superficial extracranial location, makes it one of the few cranial nerves directly testable by electrodiagnostic techniques.

CASE REPORT

Clinical History. A 25-year-old man was admitted to the hospital with bilateral facial palsies, the left occurring 1 week prior to admission and the right the morning before admission. Four weeks prior to admission, he had developed a right earache, right scalp pain, lymphadenopathy, and a fever to 104°F after scuba diving off the coast of Rhode Island. He was told he had adult mumps and was treated with oral erythromycin. One week prior to admission, he noticed tingling and numbness of his left face, followed by paralysis of the entire left face. Another physician gave him a tapering dose of corticosteroids over 1 week with improvement in his right scalp pain but with no improvement in his left facial paralysis. The morning before admission, he awoke with paralysis of his entire right face. He denied any diplopia, visual abnormalities, dysarthria, or sensorimotor disturbances other than his face. For the past several days, he had noticed that all his food tasted salty and he had difficulty localizing sounds but without a decrease in hearing acuity. He reported a rash on the back of his neck 2 months before and low-grade fevers (to 101°F) at night for the preceding few weeks. He and his wife lived in a wooded area and had found ticks on themselves. They also were in the habit of letting their two dogs sleep in their bed.

Physical Examination. He was afebrile and without meningism. No skin rash was visible. His cranial nerve exam was normal with the exception of bilateral facial paralysis involving all muscles of facial innervation. He could not close either eye. His facial sensory exam was normal. His hearing was intact and no hyperacusis was documented to bedside exam. On taste testing, he had diminished ability to denote sweet on either side. Sensory and motor exams of trunk and limbs were normal.

Laboratory Tests. Serum electrolytes and glucose were normal. Complete blood ceil count was normal as was a chest x-ray. Cerebrospinal fluid (CSF) analysis on the day of admission revealed a total protein of 168 mg/dL, a glucose of 76 mg/dL, and 108 white blood cells of which 98% were lymphocytes. CSF cytology was unremarkable and CSF VDRL was nonreactive. Mumps serology was normal in CSF and serum, and serum human immunodeficiency virus (HIV) serology was negative. His serum Lyme titers were positive with IgG 1:256 and IgM 1:64. Convalescent Lyme titers done 3 weeks later were IgG 1:1024 and IgM 1:128.

ELECTRODIAGNOSTIC EXAMINATION

Methods. A TECA TD-20 electromyograph was used for all electrodiagnostic studies. Nerve conduction studies (NCS) utilized conventional surface recording electrodes and percutaneous supramaximal stimulation via a constant voltage stimulator. Facial NCSs were done by placing the active recording electrode over the lower portion of the orbicularis oculi (o. oculi) muscle with the reference electrode on the bridge of the nose. The ground electrode was placed on the chin. Stimulation of the facial nerve was done at the stylomastoid process just inferior to the pinna of the ear. Latency was measured to the initial deflection from baseline, up or down. Amplitude of the compound muscle action potentials (CMAPs) was measured from baseline to negative peak. Recording parameters were a gain of 1–2 mV/div, a timebase of 2 ms/div and filter settings of 2-Hz low-frequency filter and 10-kHz high-frequency filter.

The blink reflex was done placing the recording electrode over the o. oculi and stimulating the supraorbital nerve in the supraorbital notch. Simultaneous recording of responses from the left and right o. oculi was done to unilateral stimulation on the right and on the left. The shortest latency of eight responses was measured for the ipsilateral R_1 and bilateral R_2 responses. Filter settings were 2 Hz for the low-frequency filter and 10 kHz for the high-frequency filter. A gain of 200–500 μV/div and a timebase of 10 ms/div was used.

Needle electromyography (EMG) was done using a Dantec concentric needle electrode, gauge 26 × 1¾″, with a recording surface of 0.07 mm^2. Fibrillations and positive waves were graded as follows: 0, none; 1+, persistent single trains in at least 2 areas; 2+, moderate numbers of fibrillations in more than 2 areas; 3+, many fibrillations in most areas; 4+, fibrillations filling the baseline in all areas.

Nerve Conduction Studies. NCSs were initially performed the day after admission, 2 days after onset of right and 8 days after onset of left facial paralysis (Table 1). The right facial motor nerve study was within normal limits. The direct facial motor and blink reflex studies revealed no response at all on the left, and a delayed right R_1 and R_2 to right and left supraorbital nerve stimulation.

NCSs were repeated 6 weeks later (Table 2). On the right, the facial motor latency was shorter and the blink reflex was normal. There was an improved left facial motor study with normal latency and a small CMAP, but no change in left blink reflex studies.

Needle Electromyography. Needle electromyography was initially performed the day after admission. On the right, facial muscles had decreased numbers of recruited motor unit potentials (MUPs) with several showing an increased discharge frequency (up to 40 Hz). MUP morphology was normal and no abnormal spontaneous activity was found. Muscles of the left face had no voluntary MUPs and no abnormal spontaneous activity (fibrillations and positive waves).

EMG was repeated 6 weeks later but only on muscles of the left face. No voluntary MUPs were

Table 1. Nerve conduction studies done day after admission.

Nerve	Nerve conduction study	Left	Right	Normal
Facial	Motor, latency (ms)	NR	3.4	<4.1 ms
	Motor, amplitude (mV)	NR	1.1	<50% diff.*
Blink	R_1 latency, ipsilateral stim.	NR	13.2	<13 ms
Reflex	R_2 latency, ipsilateral stim.	NR	42.1	<40 ms
	R_2 latency, contralateral stim.	NR	40.8	<41 ms

A 50% difference is not applicable when both sides are affected.

Table 2. Nerve conduction studies done 6 weeks later.

Nerve	Nerve conduction study	Left	Right	Normal
Facial	Motor, latency (ms)	3.2	2.8	<4.1 ms
	Motor, amplitude (mV)	0.4	1.3	<50% diff.*
Blink	R_1 latency, ipsilateral stim.	NR	11.2	<13 ms
	R_2 latency, ipsilateral stim.	NR	34.0	<40 ms
	R_2 latency, contralateral stim.	NR	32.4	<41 ms

*A 50% difference is not applicable when both sides are affected.

seen in any muscle and 1–2+ fibrillations and positive waves were seen in all.

Interpretation. The electrodiagnostic studies done the day after hospital admission demonstrated bilateral facial neuropathies, left more severe than right. The left facial neuropathy was axonopathic based on conduction studies, the absence of denervation potentials being secondary to the acuity of the lesion. The presence of motor evoked responses and voluntary MUPs on the right indicated a likely neurapraxic lesion with good prognosis for recovery, though the acuity of the lesion prevented a definite interpretation.

The second study confirmed our interpretation of the first, i.e., of an axonotmetic lesion of the left seventh cranial nerve and a neurapraxic lesion of the right.

CLINICAL COURSE

The patient was given the presumptive diagnosis of Lyme disease with bilateral seventh cranial neuritis and meningitis. He was empirically begun on a course of intravenous Ceftriaxone in the hospital and was sent home to finish a 21-day course. When seen 2 weeks later, he had complete resolution of his right-facial palsy with no change of the left-facial paralysis. When seen 1 month and again 4 months later, he had no improvement in the paralysis of left-facial muscles.

DISCUSSION

Lyme disease is the eponym given to the disease caused by infection with the spirochete *Borrelia burgdorferi*. In Europe the disease is given the eponym of Bannworth syndrome.[3] The infective agent is transmitted to humans by the bite of infected deer ticks (Ixodes dammini),[3] which are endemic to southern Rhode Island and several islands off the coast of Rhode Island, among other places. Analogous to another spirochetal infection, syphilis, Lyme disease has been said to occur in three stages of clinical manifestations.[21] Most workers, however, now prefer to consider Lyme disease as acute localized (fever, rash [erythema chronicum migrans], lymphadenopathy, and meningismus); acute disseminated, which often occurs 4–12 weeks after infection (cardiac, acute arthritis, and neurologic manifestations such as meningoencephalitis and cranial and peripheral neuropathies); and chronic disseminated (chronic arthritis, radiculoneuropathy, and encephalitis).[12] Unilateral or bilateral facial palsies are the most frequent neuropathic manifestation, occurring in 50% of patients.[21] The course of our patient was quite typical with the only event unusual being the failure of the left facial palsy to resolve, as residual symptoms of Lyme-associated facial neuritis are uncommon.[6]

In endemic areas, the presence of facial palsy, with or without meningitis, with a history of tick exposure is enough to empirically initiate treatment. Serologic confirmation of the diagnosis is possible and often desirable but is hampered by significant methodologic difficulties, the major problem being false positive results.[3] Our patient had confirmatory positive serology for Lyme disease, and a fourfold rise in titers over 3 weeks.

Electrodiagnostic studies in patients with facial palsy are useful for determining the extent of any associated peripheral and cranial nerve involvement, estimating the nature and severity of the injury, and developing a prognosis. The electrodiagnostic methods most commonly used to study the facial nerve, as illustrated in this case report, are direct facial motor NCSs, electromyography of facial innervated muscles, and the blink reflex. In addition, the nerve excitability test, which was not done in this patient, will be discussed later.

The generally good recovery of patients with Bell's palsy (86% have a complete recovery)[22] means that most patients referred to the EMG laboratory because of a facial paresis are sent for a prognosis. The prognosis is directly related to whether the facial nerve lesion is demyelinative or axonopathic. Electrodiagnostic studies are quite useful for making that determination, but are limited because Wallerian degeneration takes 5–8 days after axonal injury and NCSs will be of little prognostic value before that time.[10]

Amplitude of the direct motor nerve evoked response has been used for prognosis[20] and is probably the best available method.[8] Again, one must wait until 5–7 days after onset to get reliable results. When the CMAP amplitude is less than 10% of that on the healthy side, maximum recov-

ery will be delayed 6–12 months and function will be moderately or severely limited. If the amplitude is 10–30% of the healthy side, recovery may take 2–8 months with mild to moderate residua. If the CMAP amplitude is >30% of normal, full complete recovery can be expected at 2 months after onset.[20]

Latency of the direct facial motor nerve stimulation has been studied as a prognostic indicator,[18,23] but is clearly not as useful as amplitude measurement. When done 5–7 days after onset, three types of evoked responses are found: (1) normal, which virtually assures patients of a complete recovery without aberrant recovery; (2) prolonged latency compared with the opposite side, with frequent good recovery but some chance of synkinesis; and (3) no response, with high incidence of synkinesis and some patients with no recovery.

Needle electromyography can potentially detect abnormalities in facial-innervated muscles within hours of onset, but these changes (e.g., decreased interference pattern, increased MUP firing rate) are of little prognostic value as they do not help differentiate demyelinating from axonal lesions. The presence of even a few voluntary MUPs in a patient with complete clinical paralysis, however, indicates the nerve remains in continuity and is consistent with a better prognosis than those patients with none.[1] Fibrillations and positive waves indicate the presence of axonal degeneration and increase, but do not confirm the chance of a prolonged and incomplete recovery, yet are unlikely to be seen earlier than 1–2 weeks after onset and may not be seen for up to 3 weeks. EMG can also be helpful in evaluating the degree and pace of reinnervation as volitional MUPs reappear at a time when movement of facial muscles is not yet visible.[1] Unstable polyphasic MUPs imply ongoing reinnervation and suggest further clinical functional recovery is to come.

The blink reflex is the electric correlate of the bedside corneal reflex. In its usual format,[4] the ophthalmic division of the trigeminal nerve is stimulated electrically at the supraorbital foramen. The impulse is propagated back to the trigeminal nucleus, ipsilaterally excites the facial nucleus via an oligosynaptic reflex, and traverses a polysynaptic pathway leading to bilateral facial nucleus excitation. The resulting facial nerve potentials are recorded as motor waveforms from the orbicularis oculi. Variants of this technique can test other divisions of the trigeminal and facial nerves by stimulating the infraorbital or mental nerves and recording over the nasalis or mentalis muscles.[4]

The evoked responses to such stimulation are an ipsilateral early response, called the R_1, and a bilateral late response, called the R_2. The test requires sequential stimulation of both sides while recording simultaneously from the right and the left sides. The blink reflex differs from direct facial nerve study in that it examines the trigeminal nerve and the pons in addition to the facial nerve, and assesses proximal segments of the facial nerve inaccessible to the direct stimulation technique. A delayed or absent R_1 could be from either trigeminal or facial neuropathy but coupled with an abnormal R_2 response on the paretic side, regardless of the side of stimulation, indicates slowing in the ipsilateral facial nerve.[15] An abnormal blink reflex showing this latter pattern consistent with facial neuropathy has been found in such entities as Bell's palsy,[16] inflammatory demyelinating neuropathy,[13] hereditary neuropathy,[13] diabetes,[13] multiple sclerosis,[14] and acoustic neuroma.[9] Kimura et al.,[16] studied 144 patients with Bell's palsy and all had either absent or delayed R_1 responses on the paretic side during the first week of symptoms.

The blink reflex as a prognostic method has not been particularly helpful in that it offers little beyond direct facial nerve studies and is limited by the same time constraints. One hundred of 127 patients[15,16] with Bell's palsy with a preserved direct facial motor response had return of an absent R_1 or R_2 response, paralleling a good clinical recovery and indicating a demyelinative lesion. The 27 patients with markedly diminished direct facial motor response had no return of R_1 or R_2 responses on reflex testing, had a poorer recovery and frequent synkinesis,[17] indicative of axonal degeneration.

Nerve stimulation to evoke muscle twitch and test nerve excitability has been advocated as an early method for determining prognosis[5] but is unlikely to reliably assist the electromyographer beyond the above, more quantitative, tests.[8] Nerve excitability is tested by stimulating the facial nerve with increasing electric intensity looking for visible signs of muscle contraction.[4] The least current intensity necessary to evoke a visible contraction is the minimal excitability value. This current intensity is dependent on skin resistance, skin temperature, and the anatomic localization of the nerve. A constant current stimulator is required, with comparison to the unaffected side.[15] This test is not useful in bilateral facial neuropathies. Moreover, nerve excitability can remain normal up to 4 days after complete sectioning of the facial nerve before being lost secondary to Wallerian degeneration.[10]

Some investigators have found it possible to assess prognosis as early as 72 hours after onset using nerve excitability[7] though others have concluded that the nerve never becomes inexcitable before the fourth day.[11] The normal difference between sides is <2 mA. When the side-to-side difference is <5 mA, nerve excitability does not reliably predict outcome.[11] Patients with a large increase in excitability threshold (>10 mA) have a poor prognosis for recovery, and those with an intermediate increase (5–10 mA) have a reasonable prognosis for recovery if the threshold improves or normalizes by 1 week after onset, but a poor prognosis otherwise.[7]

At this time, there is no electrodiagnostic technique which reliably predicts prognosis in Bell's palsy within the first 24–48 hours after onset. CMAP amplitude comparing side-to-side difference at day 5–7 after onset appears to be the most reliable parameter for ultimate prognosis.[8]

The time course of recovery from facial palsy is dependent on the type and degree of pathophysiologic lesion. A neurapraxic, or purely demyelinative, lesion will recover within days to weeks as the Schwann cell remyelinates the affected segment. More severe lesions resulting in axonal damage and distal Wallerian degeneration will take longer to recover function as nerve regeneration will be required. The regeneration may well be incomplete, resulting in residual weakness, or aberrant, resulting in facial synkinesis.[17] Massey and Sanders[19] used single fiber EMG to study a traumatic facial neuropathy from 15 days after injury to full clinical recovery. They showed that jitter, a measure of neuromuscular transmission, initially deteriorated, consonant with axonal degeneration and denervation as seen on EMG. The fiber density, an electrophysiologic correlate of reinnervation, was increased a few weeks later, consistent with beginning reinnervation. Both measures reached maximum increase 5 weeks after onset, at the time of initial return of facial motor function. Both measures were still maximally abnormal by 10 weeks, when functional recovery was complete. Thereafter, fiber density and jitter returned toward normal, suggesting "the original axons had resumed function and reestablished innervation to the muscle, after which point collateral sprouting at least partially disappeared resulting in remodeling of the motor units."[19]

The major sequelae of facial palsy are persistent paresis or paralysis, corneal injury secondary to incomplete eyelid closure, facial muscle contracture, and synkinesis. Synkinesis is the phenomenon whereby the intended activation of one muscle will result in contraction of other muscles as well. This cocontraction may be due to either aberrant nerve fiber regeneration or to ephaptic transmission. Facial synkinesis can involve motor nerve fibers as well as fibers to the lacrimal and salivatory glands, resulting in the so-called "crocodile tears" phenomenon, in which gustatory salivation causes tearing as well. Facial synkinesis is seen in the majority of patients with Bell's palsy and evidence of axonopathy.[17] Facial motor synkinesis can be assessed by expanding the set-up for the blink reflex to also include recording electrodes over the o. oris or other facial muscles.[17] In a normal patient, supraorbital nerve stimulation results in a response recorded only from the o. oculi. In patients with aberrant regeneration, a response will be found in both the o. oculi and the o. oris.[17]

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