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Focal facial spasm

Article abstract—An unusual case of focal facial spasm modified by factors affecting the peripheral facial nerve was investigated in a 32-year-old woman with involuntary contractions at the left mouth and nasal area. Voluntary facial movements were normal. The involuntary spasms ceased with digital pressure over the facial nerve in the left stylomastoid area. A difference between voluntary and these involuntary facial movements occurred both with local anesthetic blockade and with crushing of the facial nerve. Blink reflexes demonstrated unilateral left synkinesis, and facial EMG showed clonic discharges and individual motor units that discharged rapidly (200 Hz). Treatment with diphenylhydantoin, carbamazepine, and prednisone was ineffective. Neurolysis of the peripheral facial nerve resulted in temporary relief, whereas biofeedback controlled the spasms. Focal facial spasms may represent a disorder of the facial nucleus influenced by both peripheral and central mechanisms.

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Involuntary facial movements differ in clinical expression and etiologies. Lesions of the facial nerve or nucleus may cause hemifacial spasm, facial synkinesis, or myokymia. These three disorders are identified by clinical and electrophysiologic criteria.

In *hemifacial spasm*, tonic or tonic-clonic facial muscle contractions may be associated with posterior fossa lesions,¹⁻¹⁰ including scarring, stretching, local pressure, ischemia, arachnoiditis, or irritation of the extra-axial portion of the seventh cranial. However, in most cases of hemifacial spasm, no intracranial abnormality is defined.¹¹ Peripheral nerve conduction velocities and blink reflex latencies are normal, but pathologic synkinesis and abnormal EMG recruitment patterns are seen during the spasm.¹² In *facial myokymia*, continuous quivering of the facial muscles is typically associated with intramedullary lesions such as multiple sclerosis^{13,14} or brainstem tumors,¹⁵ but is also seen in polyradiculopathies.^{16,17} Typical "myokymic" discharges^{16,18} are present on EMG analysis, and blink reflex latencies may be prolonged.¹⁶ *Involuntary synkinesis* is attributed to aberrant regeneration after facial nerve injury,^{19,20} with combinations of slowing of motor nerve conduction velocities, synkinesis,²¹ abnormal EMG recruitment patterns, and polyphasic motor units.²²

Facial movement disorders can also arise from disorders of the basal ganglia (as in tardive dyskinesia or Huntington's chorea) or cerebral cortex, as in focal motor epileptic discharges. In these conditions electrophysiologic analysis of peripheral nerve, short latency brainstem reflexes, and facial muscles are normal, although the late components of the blink reflex may be abnormal.²³

We recently studied a patient whose disorder was clinically and electrophysiologically similar to that of hemifacial spasm but with several unique characteristics.

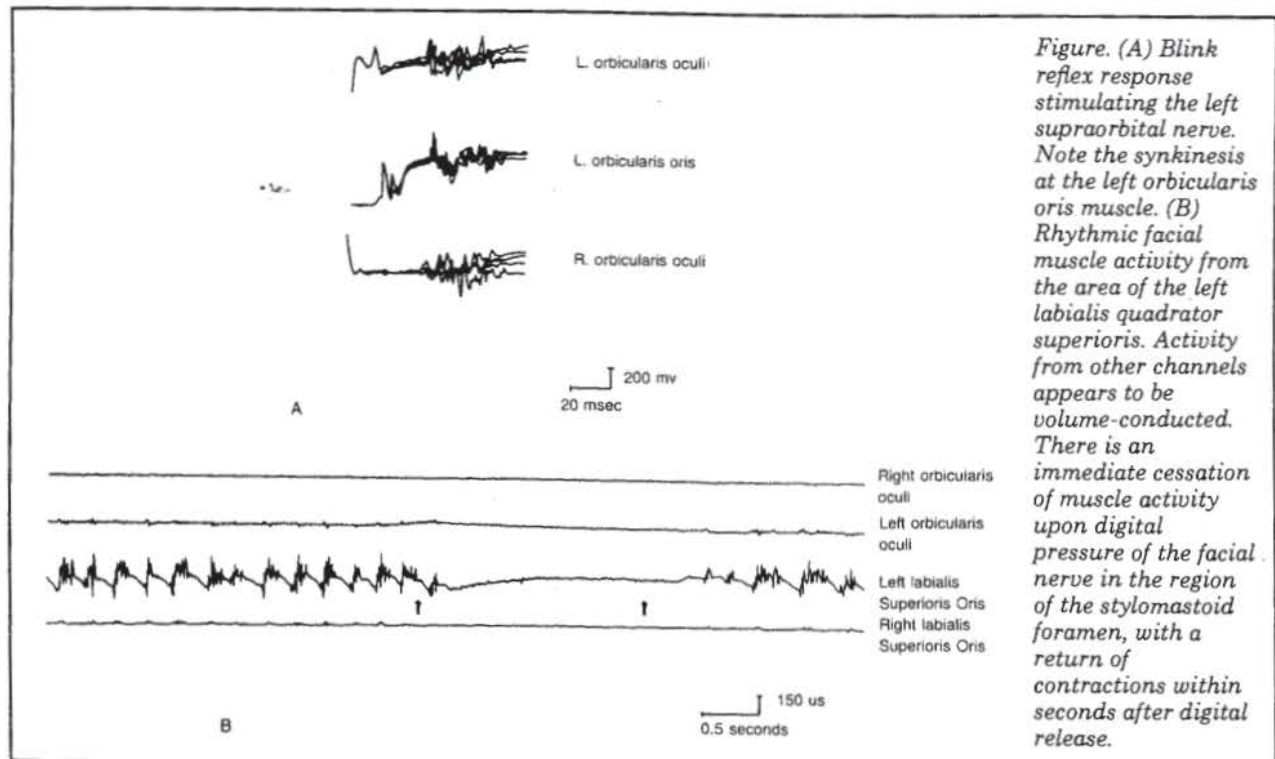
Case report. This thirty-four-year-old woman was well

until she noted the gradual onset of "numbness and tightness" of the skin in the left maxillary area. Within 3 weeks, she noted rapid involuntary muscular twitches in the left nasal area for a few seconds at a time. In the next few weeks the spasms lasted longer and spread toward the mouth. The contractions were irregular, rapid, and painless. Within 3 months, the spasms were present most of her waking hours, but disappeared with sleep. Grimacing or forced smiling immediately arrested the spontaneous contractions. Digital pressure at the left stylomastoid area immediately stopped the contractions, and the movements did not recur as long as pressure was maintained (she used this technique to stop the spasms in embarrassing situations). The spasms returned within 5 seconds when she removed the finger. Emotionally stressful situations increased the frequency of the spasms.

General physical examination was normal. There was a subjective, irregular, circular area of decreased sensation in the left maxillary area measuring approximately 2 cm in diameter. This area gradually diminished in diameter and eventually disappeared. Continuous contractions were observed in the left nasal and mouth areas. When slight digital pressure was applied (by patient or examiner) near the left stylomastoid foramen, the involuntary contractions ceased, but the patient could maintain a full range of voluntary motor facial movements. Forced smiling and grimaces also temporarily arrested the movements. The remainder of the neurologic examination was normal.

Routine laboratory tests were normal, including EEG, ABR, CSF, tomograms of the petrous temporal bone, CAT of the posterior fossa, and cerebral angiograms with selected views to exclude compression of the facial nerve by a small posterior fossa vessel.

Facial motor latencies and blink reflexes were performed with a standard technique.¹⁶ The motor latencies were 3.7 msec on the left side and 3.8 msec on the right (normal <4.0 msec), and the motor amplitudes (2.0 mV) were symmetric and normal. With the blink reflex, the orbicularis oculi early response (R1) on the left and right sides was 10.2 and 10.3 msec (normal <12.0). Ipsilateral and contralateral late orbicularis oculi (R2) responses were also normal (33 and 35 msec when stimulating the left supraorbital nerve and 34 and 36 msec when stimulating the right supraorbital nerve). Symmetric decremental responses of R2 components were seen with repetitive



stimuli. A synkinetic response of quadratus labii superioris was obtained with left-sided stimulation (figure, A), whereas no such activity was observed with right-sided stimulation.

With concentric needle electromyography there was no spontaneous activity in facial muscles. Normal motor units were present on voluntary activity, and the recruitment pattern was normal. During involuntary spasms, groups of motor units fired mainly as clonic bursts. The individual motor units were normal in amplitude, shape, and duration, and rapid discharges of up to 200 Hz frequency were observed during the clonic bursts. The latency intervals between clonic bursts of motor unit activity were up to 400 msec, and the spontaneous rhythmic clonic EMG activity typically ceased on compression of the styloid area (figure, B).

Course. Clinical trials of diphenylhydantoin, carbamazepine, and diazepam did not alter the spasms. Oral prednisone (60 mg/day) seemed to reduce the frequency, but spontaneous activity returned to pretreatment levels in a few days.

Infiltration of the facial nerve at the stylomastoid area with 2.0 cc of 1% xylocaine resulted in the gradual loss of voluntary motor facial muscle activity, followed by loss of involuntary spasms. The completeness of the block was not verified by electrical stimulation. By clinical observation, the involuntary spasms returned approximately 15 minutes after the injection, while there was still paralysis of voluntary motion.

Surgical exploration of the facial nerve trunk and the major branches revealed no gross abnormalities. A left partial parotidectomy was performed, and no scars, tumors, or adhesions were found over the left

seventh cranial nerve from the stylomastoid foramen to the peripheral branches. The distal branches that innervated the maxillary and nasal area were identified by direct nerve stimulation, and then they were crushed; complete motor nerve block was documented by a loss of recordable motor response to direct facial nerve stimulation. Postoperatively, the spasms were absent, and voluntary facial strength was normal except for the lower face. Three weeks later, the spasms reappeared, and within 6 weeks the activity was just as it had been originally. The facial paralysis slowly recovered only after the spasms returned.

After two biofeedback sessions, the patient could suppress the spasms by "concentrating" and anticipating the movement. At times of emotional stress, the spasms occasionally reappeared. The patient returned to work and has had total control of the spasms for the past 2 years. There have been no other changes in her neurologic status.

Discussion. The term "focal facial spasm" is an appropriate description of the facial movements described here. The movements were involuntary, localized to a restricted region near one nasolabial fold, and were characterized by intermittent irregular contractions. The movements were sensitive to a number of factors affecting the peripheral facial nerve that distinguished between voluntary and involuntary facial movements. First, digital pressure applied by either the patient or the examiner over the seventh cranial nerve at the stylomastoid foramen was accompanied by abrupt cessation of the move-

ments, without interfering with voluntary contractions. Second, paralysis of voluntary facial movements occurred with infiltration of a local anesthetic into the region of the facial nerve before the spasms ceased, and the spasms then reappeared before voluntary facial movements were possible. Finally, crushing the facial nerve resulted in the loss of both voluntary and involuntary movements, but the focal spasms returned several weeks before voluntary motion.

The spasms are similar in some respects to hemifacial spasms, but with some distinguishing features. First, in hemifacial spasm the abnormal muscle contractions may be precipitated by voluntary facial movements such as chewing, smiling, laughing, or blinking,^{24,25} but in our patient the spasms ceased during voluntary activity. Second, in hemifacial spasm contractions usually begin around the eye rather than near the mouth, as occurred in our patient. Third, abnormal sensations are not a feature of hemifacial spasm,^{26,27} whereas our patient had transient paresthesia in the maxillary area. Fourth, in patients with hemifacial spasm, injection of alcohol into the seventh nerve at the stylomastoid area is accompanied by paralysis,^{28,29} with voluntary motor functions usually returning before the spasms. This sequence was reversed in our patient.

A number of observations have confirmed that manipulation of the facial nerve intracranially can alter hemifacial spasms. Phenol injections may stop the spasms with only temporary paralysis.³⁰ Direct manipulation or touching the intracranial portion of the facial nerve results in the temporary cessation of the spasm,⁶ but to our knowledge transient cessation of motor activity has not been reported with superficial pressure on the peripheral nerve, as occurred in our patient. More prolonged improvement of the spasms has also been reported after crushing of the facial nerve at the stylomastoid area,^{25,31} partial lysis of the facial nerve trunk,^{32,33} or sectioning of terminal motor branches of the facial nerve.²⁴ These findings do not necessarily imply that peripheral nerve mechanisms generate the spasm, but that the abnormal movements can be modified by manipulation of peripheral nerve at different sites.

In our case, sensitivity of the involuntary movements to peripheral nerve mechanisms was best exemplified by the beneficial effects of focal pressure. We tried this maneuver on two patients with classical hemifacial spasm and one patient with facial myokymia but without effect.

In 1907, Ramsey Hunt postulated that a focally irritative afferent impulse from the facial nerve could generate spontaneous facial movements.³⁴ Experimentally injured peripheral nerves can produce a local self-generating abnormality, which may be associated with an artificial axon syndrome or ephaptic transmission,^{35,36} but these are typically limited in distribution.^{37,38} Spontaneously generated activity after nerve injury also seems to be short-lived³⁹ and may not explain persistent hemifacial

spasm or focal facial spasm. Although local nerve injury could have initiated the focal facial spasm, it would appear more likely that the persistent spontaneous contractions in our patient were generated centrally in the facial nucleus.

Nerve injuries are associated with secondary chromatolytic changes in the cells of origin,^{38,39} and synaptic contacts on those cell bodies are altered.⁴⁰⁻⁴² These mechanisms might affect neural discharges to account in our patient for both the abnormal electromyographic firing patterns and the differential susceptibility of voluntary and involuntary movements to either anesthetic blockade or recovery from crushing of the nerve. It is also evident that excitability of facial motor neurons involved in focal facial spasm can be influenced by descending pathways, since the focal movements ceased after biofeedback in our patient.

Treatment of cryptogenic facial movement disorders is unsatisfactory. Anticonvulsant medications and sedatives are ineffective. Surgical decompression of the facial nerve in the posterior fossa is sometimes effective, but in the majority of patients no specific lesions are defined other than focal demyelination as associated with aberrant blood vessels. Furthermore, it is not known how a compressive lesion of the facial nerve causes facial spasm. Our patient responded to biofeedback therapy, which may be an efficacious noninvasive treatment modality for selected patients with facial movement disorders.

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Dominant chondrodysplasia punctata with neurologic symptoms

Article abstract—There are few descriptions of major neurologic dysfunctions in either the recessive or the dominant form of chondrodysplasia punctata. In the dominant trait, often called Conradi-Hünemann disease, a normal life expectancy with normal neurologic development is the usual course for those who survive the first few weeks of life. We studied an affected infant with a severe spinal cord abnormality that was present at birth and has not been reported in either recessive or dominant chondrodysplasia punctata.

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Richard G. Curless, MD

Hereditary chondrodysplasia punctata includes macrocephaly with a flat facies and depressed nasal bridge, ichthyosis, and clinical or radiologic evidence of rhizomelia (proximal shortening of the limbs).¹ Additional radiographic features include deformities of the vertebral bodies and punctate calcifications of the distal long bones, vertebrae, and pubis. The dominant, or Conradi-Hünemann,

form is generally much milder than the recessive, or "rhizomelic" form, which usually leads to death before age 2 from repeated infections or respiratory failure.^{2,3} Occasional late survivors of the recessive form have severe mental retardation and spasticity. We studied an infant with the clinical characteristics of Conradi-Hünemann syndrome and congenital flaccid paraparesis.