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Facial Myokymia in the Guillain-Barré Syndrome

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• Facial myokymia, a unique involuntary movement of facial muscles, is described in a patient with Guillain-Barré syndrome. Although this involuntary movement is most often described with intramedullary lesions of the brain stem, it may also appear with lesions external to the neuraxis. We review the literature and propose a more widespread distribution of potential lesions along the facial nerve pathway, which may produce facial myokymia.

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Facial myokymia is a continuous, involuntary, undulating movement of the facial muscles. It has been described most prominently in association with intramedullary brain stem lesions such as multiple sclerosis,¹ pontine gliomas, and other posterior fossa tumors,²⁻⁵ and occasionally with poliomyelitis affecting the facial nerve nucleus⁴ and brain stem vascular diseases.^{4,6} Several cases of the phenomenon have been described with extramedullary brain stem tumors as acoustic schwannoma,² but in these instances, the tumors were of sufficient size to compress the brain stem. Thus, facial myokymia has been considered a clinical sign of an intramedullary brain stem lesion producing hyperexcitability (or perhaps loss of inhibitory control) in the nucleus of the facial nerve. The patient described in this report had postinfectious polyradiculopathy (the Guillain-Barré syndrome) with prominent facial myo-

kymia, indicating that these movements can also occur with lesions of the facial nerve extrinsic to the brain stem.

REPORT OF A CASE

A 30-year-old woman in previously good health noted the onset of a "flu-like" illness in April 1976 manifested by fever, chills, generalized fatigue, and myalgias. After five days, these symptoms subsided, but were then followed in about three days by numbness and tingling in the distal aspects of both hands and feet. One week later, she noted an involuntary twitching and "worm-like" movement of the lower half of the right side of her face. In the next few days, these movements spread to involve the entire right side of the face and, to some extent, the lower portion of the left side of the face.

On admission to the hospital in May of 1976, the examination revealed continuous flickering movements of the entire right side of the face extending from the platysma muscle to the frontalis. Similar movements of the buccal musculature of the left side of the face were also evident. There was no facial weakness or change in facial sensation. Taste and hearing were likewise intact. The patient had a mild degree of weakness in the lower extremities and in the distal musculature of the upper extremities, but fasciculations and myokymia were not present. All muscle stretch reflexes were absent, except for a trace response from the right biceps. There was mild symmetrical distal sensory impairment to all modalities in the extremities. The remainder of the examination was within normal limits.

The involuntary facial movements diminished, but did not totally disappear during sleep. Analyses of the urine for porphobilinogen and heavy metals showed no abnormalities. The cerebrospinal fluid obtained from a lumbar puncture was of normal pressure, but yellow-tinged. It

contained one lymphocyte, no red blood cells, a protein level of 340 mg/100 ml, and a glucose level of 60 mg/100 ml. An electromyogram (EMG) was not performed. A clinical diagnosis of Guillain-Barré syndrome was made. Over the next two months, the patient's condition gradually improved. By June of 1976, there remained only a trace of undulating facial movement in the right lower part of the face, slight distal weakness in all four extremities, absent muscle stretch reflexes, and mild distal sensory loss to all modalities. One month later, involuntary facial movements could not be detected. The reflexes were present in the upper limbs, but still absent in lower limbs. The patient's strength was now intact, but the mild distal sensory deficits persisted.

COMMENT

This case illustrates an infrequently observed clinical association of facial myokymia with the Guillain-Barré syndrome.⁶ Facial myokymia has been well described in association with multiple sclerosis and intramedullary tumors of the pons, and rarely with extramedullary tumors that compress the brain stem.¹⁻⁴ The facial myokymia in all of these cases has been presumed to result from abnormalities within the brain stem affecting the nucleus of the facial nerve. Tenser and Corbett³ thought this hypothesis was reasonable, since the myokymia was eliminated in a case of brain stem glioma by an anesthetic block of the 7th cranial nerve at the stylomastoid foramen.

Facial myokymia is to be distinguished from other involuntary facial movements such as hemifacial spasm,⁷ facial contractures,⁸ facial synkinesias accompanying regeneration of the 7th cranial nerve, facial tics, tardive

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dyskinesias, rhythmic facial contractions occurring with palatal myoclonus, seizures from focal cortical lesions, and fasciculations of facial muscles with motor neuron disease. These distinctions can be made both clinically and electrophysiologically. The characteristic EMG pattern of facial myokymia is one of spontaneous, rhythmic discharges of motor units appearing in singles, doubles, or groups, with relatively stable frequency (between 0.8 and 30 cps). There may be two types: continuous, in which rhythmic single or double discharges of one or a few motor units occur at a relatively high frequency, or discontinuous, in which rhythmic discharges of several units in groups occur at a relatively lower frequency.⁶

In the patient presented in this report, facial myokymia was the result of a disease process causing inflammation and demyelination of the facial nerve root⁹ extrinsic to the brain stem. It is possible that there were accompanying retrograde changes in the cells of the motor nucleus to provide depolarization and subsequent spontaneous discharges, but one might have expected some degree of facial weakness if this were true.

Alternately, the peripheral nerve root lesion itself may have resulted in the myokymia, in a manner similar to the instances of myokymia known to occur with peripheral nerve lesions.¹⁰ Wallis et al¹¹ characterized myokymia of peripheral nerve origin as being multiple, continuous fasciculations giving the appearance of undulation rather than isolated muscle twitches. They believed that the myokymia represented spontaneous isolated discharges of individual motor units caused by the peripheral nerve lesion.

The persistence of myokymia, even after spinal anesthesia, supports the idea that spontaneous activity originated from the peripheral nerves themselves, without participation of their cell bodies. Williamson and Brooke¹² agreed with the peripheral nerve origin of myokymia, and referred to the work of Denny-Brown and Foley,¹³ who showed that repeated bursts of action potentials characteristic of myokymia could occur in normal individuals following the return of blood flow to a nerve subjected to ischemia for longer than 15 minutes.

In an unusual case described by Welch et al,¹⁴ both facial and limb muscle myokymia developed in a young woman. Though the authors do not specifically address themselves to the mechanisms of the facial movements, they present evidence of segmental demyelination from a sural nerve biopsy to substantiate that a peripheral nerve lesion was associated with the myokymia. They postulated the existence of increased excitability of the peripheral nerve as the mechanism for the myokymia, since spinal anesthesia did not eliminate the abnormal movement in their patient. It seems plausible to consider that the facial nerve was also affected by the segmental demyelination.

Demyelination is a striking finding in patients with the Guillain-Barré syndrome, and the finding of facial myokymia may reflect the increased spontaneous discharges of the facial muscles as a consequence of this lesion. It may even be that the myokymia seen in patients with multiple sclerosis or intramedullary brain stem lesions can be caused by the same peripheral nerve mechanisms, since the 7th cranial nerve has a lengthy course through the brain

stem and secondary changes in 7th-nerve fiber could result from the brain stem lesion itself. However, it is more likely that facial myokymia is a neurological sign of increased excitability in the facial motor system, beginning with the supranuclear pathways to the facial nucleus, the neurons of the facial nucleus, the axons both within and extrinsic to the brain stem, and the neuromuscular junction at the facial muscles. Lesions anywhere along this pathway can result in the characteristic involuntary movements described as myokymia.

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