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Video Abstracts

Bilateral Facial Spasm Following Guillain–Barré Syndrome

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Abstract

Background: We report a patient who developed lower facial muscle spasm at rest and bilateral facial synkinesis several months after treatment of Guillain–Barré syndrome (GBS); this finding, to our knowledge, is hitherto unreported.

Phenomenology Shown: Bilateral synkinesis, facial muscles spasm at rest, bilateral postparalytic facial syndrome.

Educational Value: Aberrant regeneration of nerve fibers post GBS, resulting in facial muscles spasm at rest, bilateral synkinesis.

Keywords: Facial spasm, Guillain–Barré syndrome, aberrant regeneration, synkinesis

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Ethics Statement: All patients that appear on video have provided written informed consent; authorization for the videotaping and for publication of the videotape was provided.

A 45-year-old female was diagnosed with Guillain–Barré Syndrome (GBS) 4 months previously. She had subacute distal weakness, dysarthria, and dysphagia. She demonstrated facial diplegia, distal weakness, and absent deep tendon reflexes. Cerebrospinal fluid analysis showed elevated protein of 528 mg/dL (normal 15–45 mg/dL). The remainder of the results were normal (red blood cells, 0 cells/mm3; white blood cells, 6 cells/mm3; glucose, 73 mg/dL) or negative (Lyme's titer, angiotensin-converting enzyme). Magnetic resonance imaging of the spine showed nerve root enhancement and electromyography (EMG)/nerve conduction studies showed generalized sensorimotor predominantly demyelinating neuropathy. Intravenous administration of immunoglobulin for 3 days improved the motor symptoms.

Four months later, she complained of extreme tightness in the lower facial muscles. She was also noted to have synkinesis (volitional contraction of one muscle accompanied by synchronous contraction of other muscles) and the lower lip was turning in when she opened her mouth. EMG demonstrated spontaneous motor unit discharges in the lower facial muscles with no evidence of myokymia (Video 1). With this EMG finding and the complaint of tightness at rest, we labeled this facial spasm. She showed synkinesis of ipsilateral facial muscles on both sides. Treatment with botulinum toxin improved her facial spasms.

In cases of Bell's palsy, Wallerian degeneration in nerve fibers leads to a variable degree of aberrant regeneration. This may lead to post-paralytic facial syndrome (PFS), featuring synkinesis, myokymia, and involuntary mass contractions of muscles on the affected side.[1] Synkinesis is also noted in primary hemifacial spasm (HFS). EMG helps to differentiate HFS from PFS. Abnormal synkinetic EMG activity recorded in the orbicularis oris with blinking is not always present in the patient with HFS but is invariably seen in PFS. Spontaneous high-frequency, synchronized, repetitive firing of action potentials is seen in HFS. Myokymic discharges (low-frequency, small action potentials firing irregularly in facial muscles) are characteristic EMG findings in PFS.[1] Our patient, showed no myokymic discharges, but demonstrated constant ongoing motor unit activity.

Facial spasm in the present case may have resulted from spontaneous activity generated in the facial nucleus being amplified at trigger zones at the site of lesion and causing spontaneous contraction of muscles innervated by the nerve.[2] In PFS, Cossu and colleagues[3] recorded small amplitude polyphasic motor unit action potential (MUAPs) that were heard to fire at a frequency variable between 1 and 20 Hz, even if the patient was requested to relax. We hypothesize that the somatotopic organization of facial motonucleus resulted in selective lower facial muscle spasm at rest in addition to synkinesis, presumably due to aberrant regeneration bilaterally.

Video 1. **Bilateral Facial Spasm. Segment 1.** Involuntary contraction of bilateral lower facial muscles when she is blinking her eyes (synkinesis), which are not simultaneous and are asynchronous on both the sides (there are a few very brief instances of contractions limited to one side of the face, indicating that contractions on each side of the face are independent). She also complains of extreme tightness in lower facial muscles, when she is at rest. **Segment 2.** Electromyography (EMG) ongoing spontaneous motor unit discharges even when she is at rest, and with blinking increased EMG activity is noted in the mentalis muscles (EMG needle is in mentalis muscle).

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