A Typical Presentation for a Rarely Diagnosed Disease, Facial Onset Sensory and Motor Neuronopathy: A Case Report

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Case

A 66-year-old male with a history of Factor V Leiden mutation, degenerative disc disease, and hypertension presented for left-sided facial numbness gradually extending from nares to orbit over 5-months. Associated symptoms included slurred speech and left upper extremity (LUE) weakness. Over 3-years, symptoms progressively included bilateral facial weakness and numbness, worsening dysarthria, dysphagia, jaw drop, and left lower extremity weakness.

Diagnostics

MRI Brain: Nonspecific medial left temporal lobe T2/Flair Hyperintensities.
MRI C-Spine: Multilevel discogenic and hypertrophic changes, spinal stenosis without myelopathy, neuroforaminal narrowing.
VFSS: Mild-moderate pharyngeal dysphagia.
Negative paraneoplastic, ganglioside, myasthenia gravis autoantibody panels.
LP: elevated CSF protein, 103.

Electrodiagnostics

Reported findings include widespread axonal loss, more commonly in cervical than lumbar myotomes, and diminished SNAPs. Reduced or absent corneal and blink reflexes are early findings due to trigeminal nerve involvement.
Our findings on 2 EMGs 3 months apart were most consistent with diffuse, sensorimotor, axonal more than demyelinating polyradiculoneuropathy, with substantial LUE axon-loss on needle EMG.

Treatment

Oral steroids for possible inflammatory peripheral polyneuropathy possibly plateaued symptoms; however, symptoms ultimately worsened. At this time, there is no known effective treatment.

Motor NCS

Left arm: Borderline slow distal latencies and conduction velocities with normal amplitudes. Bilateral tibial: small amplitudes. Facial Motor: normal distal latency and amplitude

Sensory NCS

Left arm: slightly slow peak latencies with borderline small normal amplitudes.
Right and Left Sural: Absent

Needle EMG

Cranial nerve innervated muscles were normal.
Fibrillations in 7 of 9 tested muscles in left arm, also bilateral gastrocnemius, abductor hallucis, left posterior tibialis, and thoracic paraspinals at T4.
No fasciculations were noted.

Blink Reflex

Present and normal with exception of a possible absent right contralateral R2 response.

Discussion

First described by Vucic et al. (2006), the slowly progressive syndrome, Facial Onset Sensory and Motor Neuronopathy (FOSMN), has few documented cases. Onset is typically later in life. Symptoms typically consist of paresthesias in a trigeminal distribution, followed by facial and bulbar weakness, and UE weakness. FOSMN may be under diagnosed and share similar pathophysiologic features with amyotrophic lateral sclerosis and frontotemporal dementia.

Conclusions

Consider FOSMN in patients presenting with: asymmetric facial sensory symptoms with associated features of bulbar greater than appendicular, lower motor neuron degeneration. Pathogenesis remains uncertain, although FOSMN likely represents a neurodegenerative proteinopathy.

References: Q Zheng, et al., (2016) Facial onset sensory and motor neuronopathy − Springer
Fluchere, et al., (2011). Clinical features and follow-up of four new cases of facial-onset sensory and motor neuronopathy - Muscle & nerve.