Facial-Onset Sensory Motor Neuronopathy Syndrome is not Always Facial Onset

Weijun Zhang\(^a\)
Qin Huang\(^a\)
Zhengzheng Xuan\(^a\)
Shibiao Wu\(^b\)
Yuanqi Zhao\(^b\)
Haoyou Xu\(^b\)

\(^a\)Departments of Neurophysiology and
\(^b\)Neurology, The Second Affiliated Hospital of Guangzhou University of Chinese Medicine, Guangzhou, China

Dear Editor,

Facial-onset sensory motor neuronopathy (FOSMN) is a rare disorder characterized initially by trigeminal sensory involvement followed by sensory propagation and motor neuronopathy.\(^1\) The literature indicates that FOSMN typically commences with facial symptoms. Here we report a case in which FOSMN did not involve a facial onset.

A 53-year-old male developed numbness in the left palm in December 2015, and 2 years later it had spread to the shoulder. He presented with weakness when grasping with the left hand, and the arm muscles were slightly atrophied. A nerve conduction study revealed sensory axonal damage to the left median and ulnar nerves. The symptoms did not improve significantly after he received neurotrophic treatment at his local hospital. The atrophy of the left upper limb and left shoulder muscles became aggravated in August 2018, and he was diagnosed with cervical spondylosis by magnetic resonance imaging (MRI) of the cervical spine at his local hospital. He underwent surgery, but this did not improve the symptoms. Ten months later he was unable to lift his left upper limb, and the left lower limb was numb. He gradually developed dysphagia, dysarthria, limitation of the right upper limb lifting grip, and obvious numbness on the left side of the face. He had no obvious abnormality in a mutational assay for the SOD1 gene. He was admitted to our hospital on November 25, 2019.

A neurological examination showed decreased superficial sensation on the left side of the face. Fasciculation was observed on the chin muscle. The muscle strength of the left masticatory muscle was slightly decreased. His articulation was not clear, there was occasional coughing when drinking water, the pharyngeal reflex was weakened, and the left side of the tongue was atrophied with fasciculation. There was wasting of the left brachioradialis, supraspinatus, infraspinatus, biceps, triceps, deltoid, and forearm muscle groups. The muscle strengths on the Medical Research Council Scale were 1/5, 3/5, 4/5, and 5/5 in the left upper limb, right upper, left lower, and right lower limbs, respectively. Both superficial and deep sensations were reduced in the left upper and lower limbs, and the tendon reflex was absent in the left upper limb.

The findings of general laboratory measurements including lipids and the KD/SBMA gene were negative except for creatine kinase (241 U/L) and prolactin (698.4 mIU/L). The findings for autoantibodies including the ganglioside antibody spectrum, the antinuclear antibody spectrum, and the systemic-vasculitis-associated antibody were negative except for the antinuclear antibody (titer=1:100). The findings of cerebrospinal fluid analyses for glucose, oligoclonal bands, and cytology were unremarkable except for the white blood count \((12 \times 10^6/L)\) and protein level \((604 \text{ mg/L})\). Breast ultrasound and brain MRI produced unremarkable findings.

The blink reflex showed delayed or absent responses bilaterally. The main findings of a nerve conduction study were reduced amplitudes of the motor and sensory nerve action potentials in the bilateral median and left ulnar nerves. Needle electromyography showed wide-
spread neurogenic changes.

The findings of the electrodiagnostic studies and the negative genetic analysis excluded amyotrophic lateral sclerosis and Kennedy disease. Tangier disease, which is a disorder of lipid metabolism, was excluded based on the normal lipid levels. The unremarkable MRI findings excluded syringomyelia and syringobulbia. After excluding other disorders that are likely to present similar findings, the clinical and electrodiagnostic findings were considered to be most consistent with FOSMN. The patient received neurotrophic treatment and hormone therapy, but his condition continued to progress. At the last follow-up, his muscle strengths were approximately 0/5, 2/5, 3/5, and 4/5 in the left upper, right upper, left lower, and right lower limbs, respectively, and he could not walk up stairs without assistance.

FOSMN is a rare, slowly progressive neurodegenerative disease whose presenting symptoms include paresthesia and numbness in the trigeminal nerve distribution, and it slowly progresses to affect the scalp, neck, upper trunk, and upper extremities. FOSMN is characterized by the facial onset of sensory abnormalities and the subsequent development of motor deficits. It is particularly noteworthy that our patient did not report initial facial symptoms. We speculate that either the patient ignored the facial symptoms at the beginning or that the initial symptom was not facial numbness. We consider that the latter explanation is more likely based on our review of the clinic and in-hospital records of the patients after he developed numbness in the left palm, and given that professional examinations of the neurological system revealed no positive facial signs. We reconfirmed with both the patient and his relatives that no facial symptoms were initially present. Therefore, this case seems to be a variant of FOSMN, and indicates that facial onset should not be used as a diagnostic criterion for FOSMN. Instead, FOSMN should be considered whenever there are symptoms of medulla oblongata, muscle weakness, atrophy, and fascicular combined with facial sensory disorders.

This case is a special case of FOSMN that complicates its symptomatology. It is necessary to follow up and collect more relevant cases in order to improve the understanding of FOSMN.

Author Contributions
Conceptualization: Haoyou Xu. Methodology: Shibiao Wu. Supervision: Yuanqi Zhao. Validation: Haoyou Xu. Writing—original draft: Weijun Zhang. Writing—review & editing: Qin Huang, Zhengzheng Xuan.

ORCID iDs
Weijun Zhang https://orcid.org/0000-0003-4161-8088
Qin Huang https://orcid.org/0000-0003-1964-1572
Zhengzheng Xuan https://orcid.org/0000-0003-2908-2524
Shibiao Wu https://orcid.org/0000-0002-0444-6814
Yuanqi Zhao https://orcid.org/0000-0003-1254-6732
Haoyou Xu https://orcid.org/0000-0003-1497-022X

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

Acknowledgements
None.

REFERENCES
1. Vucic S, Tian D, Chong PS, Cudkowicz ME, Hedley-Whyte ET, Cros D. Facial onset sensory and motor neuronopathy (FOSMN syndrome): a novel syndrome in neurology. Brain 2006;129:3384-3390.