

Unilateral Pure Trigeminal Motor Neuropathy

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PRESENTATION OF CASE

A 17-year-old male reported to us in the Department of Medicine, Maharishi Markandeshwar Institute of Medical Sciences & Research, Mullana, Ambala, Haryana, India, in July 2019 with a complaint of left sided facial asymmetry since one and half years which was insidious in onset. The patient was himself unaware of the facial asymmetry and it was initially noticed by his mother. Gradually, the asymmetry of face became more prominent and was noticed by the patient and other relatives as well. There was no history of pain, numbness, difficulty in chewing or mouth opening, swallowing of food. He had no preceding febrile illness or local trauma. On local examination, there was no tenderness on left side of face. There was visible asymmetry of face due to atrophy of left masseter muscle [figure 1]. No sensory deficit was elicited during neurological examination. The diagnosis of unilateral pure trigeminal motor neuropathy was made clinically. Confirmation of the neuropathy was done using Electromyography study which showed some fibrillation potentials and positive sharp waves in the muscles innervated by trigeminal nerve (masseter), however nerve conduction study was normal. In our patient, the MRI Brain (done at other centre) didn't show any intracranial abnormality and was inconclusive to diagnose atrophy and fat deposition in masseter muscle. Demonstration of fat deposition and atrophy of masseter muscle on MRI Face would have been diagnostic in this case but it could not be obtained as consent for repeat MRI was not given by the patient. No nerve/muscle biopsy was undertaken in the present case. The diagnosis was primarily on the basis of clinical examination and collaborative EMG findings. To rule out these aetiologies, radiological evaluation of the patient including MRI Brain is indicated.

There is no definite etiological factor associated with pure motor trigeminal neuropathy, although there are reports of its association with following underlying disorders:

1. Infectious (Viral).
2. Autoimmune (RA, Sjogren Syndrome).
3. Vascular (Infarction).
4. Neoplastic (Meningioma).
5. Multiple Sclerosis.
6. Trauma.

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DISCUSSION OF MANAGEMENT

Since the patient didn't have any complaints apart from the asymmetry of face, he was counselled regarding his neurological condition and need of regular follow-up.

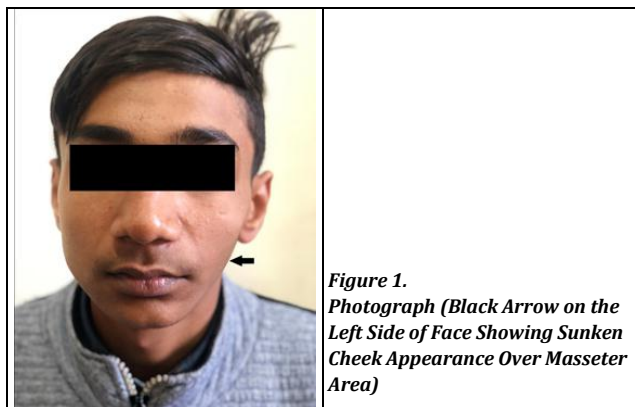


Figure 1. Photograph (Black Arrow on the Left Side of Face Showing Sunken Cheek Appearance Over Masseter Area)

DISCUSSION

Pure trigeminal nerve motor neuropathy is a clinical rarity with no estimated incidence or prevalence in literature. Although the condition was first described by Chia LG et al in 1988,^[1] but to the best of our knowledge, very few cases have been reported in literature till date. UTMN is characterized by mandibular branch motor weakness without any signs of trigeminal sensory or other cranial nerve involvement. The underlying causes reported include infections, cerebrovascular accident, neoplastic or traumatic events.^[2] The clinical manifestation varies with the chronicity of the process, like facial asymmetry, pain, weakness on chewing. Opening deviation is one of the important symptoms of pure trigeminal motor neuropathy.^[3] In some patients, there may be initial symptoms like mastication problems and feeling of weakness in the jaw muscles, most of patients were reported after visible asymmetry of the face was recognized by the patient or some relative. In the present case also, the facial asymmetry was first recognized by the mother of the patient. The trigeminal nerve has four brainstem nuclei: the main sensory nucleus, the spinal nucleus, the motor nucleus, and the mesencephalic nucleus, which is involved in proprioception. The motor nucleus of the trigeminal nerve is located medial to the principal sensory nucleus in the rostral pons. The large sensory and smaller motor roots exit via the lateral pons as a common trunk and the motor root remains inferior to the sensory root through the course in the preganglionic segment

and the trigeminal ganglion. The trigeminal ganglion lies in Meckel's cave, and distal to the ganglion, the trigeminal nerve trifurcates into its three principal branches, the ophthalmic (V1), maxillary (V2), and mandibular nerves (V3).^[4] Any lesion which involves the fifth cranial nerve along its protracted course from its nucleus in the brainstem to its distal facial ramifications to the masticatory muscles may result in the features of trigeminal motor involvement. Selective involvement of the motor fibres may be possibly caused by inflammatory, infectious, and post viral autoimmune processes.^[5]

Diagnosis of pure motor trigeminal neuropathy involves clinical neurological examination, radiological evaluation including MRI (which may help in differential diagnosis) and Electromyographic evaluation. There is no effective treatment for this condition. The management may include surgical intervention in cases in whom definite underlying pathological cause like a tumour is located on radiological evaluation. In other patients where no aetiology is confirmed, the treatment of pure motor trigeminal neuropathy includes reassurance and physiotherapy, although some attempts have been made to surgically correct the disability.^[3]

FINAL DIAGNOSIS

The final diagnosis of pure motor trigeminal neuropathy was made clinically. There was no sensory involvement. Investigations didn't reveal any underlying secondary aetiology for the disorder.

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