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Case Report

Unilateral Trigeminal Mandibular Motor Neuropathy Caused by Tumor in the Foramen Ovale

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Pure trigeminal motor neuropathy is characterized by trigeminal motor weakness without signs of trigeminal sensory or other cranial nerve involvement. We describe a 63-year-old woman with progressive weakness and atrophy of the left masticatory muscles. She had no sensory disturbance. The diagnosis of pure trigeminal motor neuropathy was made on the basis of clinical and electrophysiologic studies. Magnetic resonance imaging of the brain revealed enhancement of the enlarged mandibular branch of the trigeminal nerve coursing through the left foramen ovale. Our observations suggest that pure trigeminal motor neuropathy can be induced by a tumor.
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Trigeminal neuropathy usually presents with sensory disturbance.[1-4] The predominant neurologic finding is hypesthesia in the territory of one or more divisions of the trigeminal nerve. Combined motor and sensory involvement can occasionally be seen in a mandibular branch (V_3) lesion since combined sensory and motor fibers form the fascicular portion prior to exiting at the lower pontine level. Combined sensory and motor V_3 lesions have been reported with extradural tumors, trauma,[5] and viral infection.[4] However, pure trigeminal motor neuropathy is rarely reported.[6-11] We present a patient with unilateral trigeminal motor weakness and muscle atrophy which were not accompanied by trigeminal sensory involvement.

CASE REPORT

A 63-year-old woman noticed progressive left jaw weakness that first appeared 6 months prior to her initial visit. She subsequently noted progressive muscle wasting in the left cheek and temple, but no facial sensory disturbance. She had no preceding febrile illness, head trauma, or stroke. A neurologic examination showed weakness and wasting of the left temporalis and masseter muscles (Fig. 1). Her jaw deviated to the left on mouth opening, and she could not move her jaw to the right side against resistance. Corneal reflex and facial sensation were normal. Electromyography revealed abnormal spontaneous activity (positive sharp waves and fibrillation potentials), chronic neurogenic motor unit potentials, and reduced interference patterns in the left temporalis and

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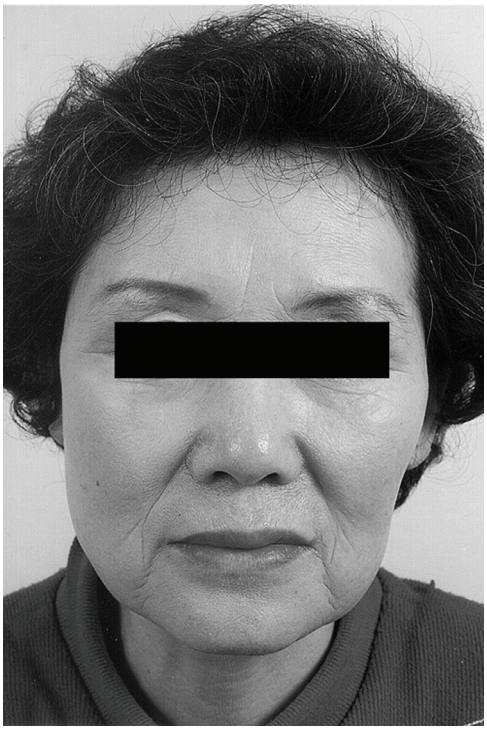

Figure 1. Severe muscle wasting on the left side of the patient's face.

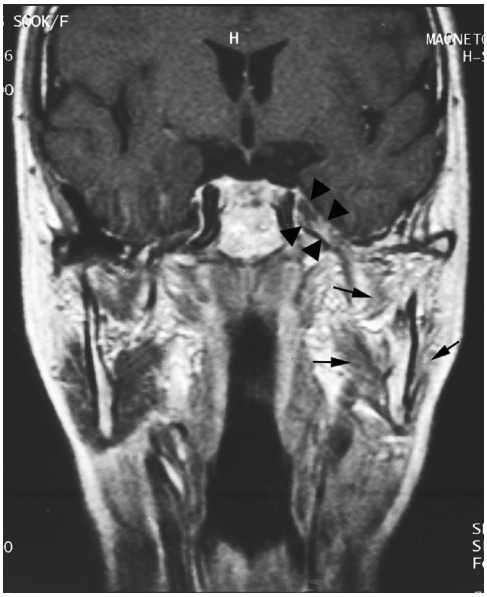

Figure 2. Coronal contrast-enhanced T1-weighted brain MRI. An enhanced mass was evident in the mandibular branch of the trigeminal nerve coursing through the left foramen ovale (arrowheads), with atrophy of the left masseter and pterygoid muscles (arrows).

masseter muscles consistent with mandibular motor neuropathy. Muscles innervated by the facial (VII), spinoaccessory (XI), and hypoglossal (XII) nerves were all normal. Conduction in the left facial nerve, blink reflex, and brainstem auditory evoked potentials were all normal. Coronal magnetic resonance imaging (MRI) views confirmed atrophy of the left temporalis, masseter, and pterygoid muscles. A contrast-enhanced linear mass was evident through the left foramen ovale (Fig. 2). Laboratory results including erythrocyte sedimentation rate, serum glucose, and antinuclear antibodies were normal. The patient refused a biopsy. A focal tumor was diagnosed, and she underwent Gamma Knife radiosurgery with a maximum dose of 30 Gy and a margin dose of 15 Gy. The lesion volume was initially 0.5 cm^3, and had decreased on a follow-up MRI scan performed 12 months later. However, there was no clinical improvement.

DISCUSSION

Pure trigeminal motor neuropathy is characterized by mandibular branch motor weakness without any signs of trigeminal sensory or other cranial nerve involvement. We are aware of only ten patients with isolated trigeminal motor neuropathy that have been reported since the first description in five patients by Chia[6] (Table 1).

The motor portion of the trigeminal nerve has its nucleus in the pontine tegmentum. The fascicular portion becomes incorporated into the mandibular branch of the trigeminal nerve in Meckel's cave. The mandibular branch containing both sensory and motor fibers exits the skull base through the foramen ovale, and supplies the masseter, temporalis, pterygoid, mylohyoid, and tensor veli palatini muscles and the anterior belly of the digastric muscle. A lesion anywhere along its course from the pons to distal peripheral nerve-innervating muscles can produce symptoms and signs of trigeminal motor involvement. Since both sensory and motor fibers form the mandibular branch, a lesion at this level should result in sensory and motor deficits.

Selective vulnerability is commonly seen in inflammatory, infectious, and postviral autoimmune processes.

Table 1. Summary of reported cases of unilateral trigeminal pure motor neuropathy

Reference	Sex/age (years) at onset	Masticatory muscle weakness and atrophy	Facial sensory impairment	Suggested cause	Electrophysiologic results	EMG of masticatory muscles
Chia, 1988[6]	M/30	+	Facial pain on chewing	Viral infection	Normal blink reflex and BAEP	Spontaneous activities, chronic neurogenic MUP
	M/29	+	Cheek soreness	Same as above	Same as above	Same as above
	M/20	+	Cheek pain	Same as above	Same as above	Same as above
	F/22	+	Not described	Unknown	Same as above	Same as above
	M/24	+	-	Unknown	Same as above	Same as above
Moon et al., 1993[7]	M/58	+	Cheek pain	Viral infection	Normal blink reflex	Same as above
Beydoun, 1993[8]	M/29	+	-	NF2	Normal blink reflex; prolonged latency or absence of waves I and V in BAEP	Spontaneous activities, decreased number of motor units
Takamatsu et al., 1993[11]	M/38	+	-	Multiple sclerosis	Normal blink reflex, BAEP, and trigeminal sensory evoked potential	Neurogenic pattern
Ko and Chan, 1995[10]	F/70	+	-	Head trauma	Normal blink reflex and BAEP	Spontaneous activities, scanty or no MUP
Kang et al., 2000[9]	M/38	+	-	Autoimmune reaction to a viral infection	Normal blink reflex, BAEP, and trigeminal sensory evoked potential; ipsilateral trivial response in masseter reflex	Spontaneous activities, chronic neurogenic MUP
Present case	F/64	+	-	Tumor	Normal blink reflex and BAEP	Spontaneous activities, chronic neurogenic MUP

BAEP; brainstem auditory evoked potentials, EMG; electromyography, MUP; motor unit potential, NF2; neurofibromatosis type 2, +; present, -, absent

A predominance of motor involvement is frequent in Guillain-Barre syndrome, whereas sensory involvement predominates in varicella-zoster reactivation. Three of the five cases originally reported by Chia[6] had preceding symptoms of upper respiratory infection. A similar case attributed to an autoimmune response has been reported recently.[9] In our case, the contrast-enhanced enlarged mandibular branch of the trigeminal nerve exiting the foramen ovale was strongly suggestive of a tumor. Beydoun[8] presented a case with unilateral trigeminal motor neuropathy as a presenting feature of neurofibromatosis type 2, in which brain MRI revealed a lesion in the vicinity of the foramen ovale. Beydoun suggested that the lesion most likely represented a schwannoma involving the mandibular motor fiber, which is similar to the lesion seen on MRI in our patient. However, our patient had no clinical feature of neurofibromatosis type 2. Meningioma (or perineurial tumor) spread arising from the face or nasal tissue[12] may also occur, but is less likely given similar MRI findings. Facial sensory disturbance has been emphasized in most reported cases of tumor, but motor weakness may also occur.[13]

Although a histologic confirmation was lacking in our case, the lesion appeared to be a schwannoma involving the mandibular motor branch of the trigeminal nerve based on the prolonged clinical course and the MRI appearance. Our observations suggest that isolated motor weakness in the distribution of the trigeminal nerve may occur from a nerve sheath tumor even in the absence of a sensory deficit.

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