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

Kyung Seok Park, M.D., Jae-Myun Chung, M.D.*, Beom S. Jeon, M.D.,
Seong-Ho Park, M.D., Kwang-Woo Lee, M.D.

Department of Neurology, Seoul National University College of Medicine, Seoul;
Department of Neurology, Inje University College of Medicine*, Kimhae, Korea

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.


Key Words: Trigeminal motor neuropathy, Mandibular nerve, Neoplasm

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...
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 cm³, 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 (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>
<thead>
<tr>
<th>Reference</th>
<th>Sex/age (years) at onset</th>
<th>Masticatory muscle weakness and atrophy</th>
<th>Facial sensory impairment</th>
<th>Suggested cause</th>
<th>Electrophysiologic results</th>
<th>EMG of masticatory muscles</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chia, 1988&lt;sup&gt;6&lt;/sup&gt;</td>
<td>M/30</td>
<td>+</td>
<td>Facial pain on chewing</td>
<td>Viral infection</td>
<td>Normal blink reflex and BAEP</td>
<td>Spontaneous activities, chronic neurogenic MUP</td>
</tr>
<tr>
<td></td>
<td>M/29</td>
<td>+</td>
<td>Cheek soreness</td>
<td>Same as above</td>
<td>Same as above</td>
<td>Same as above</td>
</tr>
<tr>
<td></td>
<td>M/20</td>
<td>+</td>
<td>Cheek pain</td>
<td>Same as above</td>
<td>Same as above</td>
<td>Same as above</td>
</tr>
<tr>
<td></td>
<td>F/22</td>
<td>+</td>
<td>Not described</td>
<td>Unknown</td>
<td>Same as above</td>
<td>Same as above</td>
</tr>
<tr>
<td></td>
<td>M/24</td>
<td>+</td>
<td>-</td>
<td>Unknown</td>
<td>Same as above</td>
<td>Same as above</td>
</tr>
<tr>
<td>Moon et al., 1993&lt;sup&gt;7&lt;/sup&gt;</td>
<td>M/58</td>
<td>+</td>
<td>Cheek pain</td>
<td>Viral infection</td>
<td>Normal blink reflex</td>
<td>Same as above</td>
</tr>
<tr>
<td>Beydoun, 1993&lt;sup&gt;8&lt;/sup&gt;</td>
<td>M/29</td>
<td>+</td>
<td>-</td>
<td>NF2</td>
<td>Normal blink reflex; prolonged latency or absence of waves I and V in BAEP</td>
<td>Spontaneous activities, decreased number of motor units</td>
</tr>
<tr>
<td>Takamatsu et al., 1993&lt;sup&gt;11&lt;/sup&gt;</td>
<td>M/38</td>
<td>+</td>
<td>-</td>
<td>Multiple sclerosis</td>
<td>Normal blink reflex, BAEP, and trigeminal sensory evoked potential</td>
<td>Neurogenic pattern</td>
</tr>
<tr>
<td>Ko and Chan, 1995&lt;sup&gt;10&lt;/sup&gt;</td>
<td>F/70</td>
<td>+</td>
<td>-</td>
<td>Head trauma</td>
<td>Normal blink reflex and BAEP</td>
<td>Spontaneous activities, scanty or no MUP</td>
</tr>
<tr>
<td>Kang et al., 2000&lt;sup&gt;9&lt;/sup&gt;</td>
<td>M/38</td>
<td>+</td>
<td>-</td>
<td>Autoimmune reaction to a viral infection</td>
<td>Normal blink reflex, BAEP, and trigeminal sensory evoked potential; ipsilateral trivial response in masseter reflex</td>
<td>Spontaneous activities, chronic neurogenic MUP</td>
</tr>
<tr>
<td>Present case</td>
<td>F/64</td>
<td>+</td>
<td>-</td>
<td>Tumor</td>
<td>Normal blink reflex and BAEP</td>
<td>Spontaneous activities, chronic neurogenic MUP</td>
</tr>
</tbody>
</table>

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 had preceding symptoms of upper respiratory infection. A similar case attributed to an autoimmune response has been reported recently. In our case, the contrast-enhanced enlarged mandibular branch of the trigeminal nerve exiting the foramen ovale was strongly suggestive of a tumor. Beydoun 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 perineural tumor) spread arising from the face or nasal tissue 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.

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.

REFERENCES