



Clinical Reasoning: A 57-year-old man with jaw spasms

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SECTION 1

A 57-year-old man presented with spasms of his left jaw. Two years prior, he had developed left-sided facial numbness followed by development of left-sided shock-like pain and then involuntary and repetitive movements of the jaw-closing muscles. Jaw muscle contractions were episodic, interfered with chewing and talking, and led to frequent tongue biting. Individual spasms varied from seconds to minutes and he reported that function in between episodes was normal, although his wife felt he spoke with reduced mouth opening. He sometimes awoke with a bloody tongue, suggesting that contractions occurred during waking and sleep. He had no history of premonitory sensation or relief associated with the spasms. He reported no difficulty swallowing.

Neurologic examination demonstrated decreased sensation in all 3 trigeminal divisions on the left and a reduced corneal reflex on the left. Masseter bulk, tone, and strength were normal bilaterally. Contractions were noted in the jaw-closing muscles. Mouth opening was limited during speech. There was no facial weakness. There was no palatal tremor. Head rotation, shoulder shrugging, and tongue movements were normal. With the exception of mild-to-moderate distal sensory deficit, the remainder of the neurologic examination was unremarkable.

Questions for consideration:

1. What is the localization?
2. What are potential etiologies?

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SECTION 2

This patient's examination revealed 2 localizing abnormalities: decreased left facial sensation with diminished corneal reflex and frequent contractions of the left jaw muscles. Both findings localize to the trigeminal nerve. The sensory fibers of the trigeminal nerve originate at the trigeminal nucleus in the pons and the motor fibers of the trigeminal nerve originate in the pons in an area close to, but not in, the trigeminal nucleus. Sensory fibers project from the trigeminal nucleus to the trigeminal ganglion, at which point they split into 3 branch distributions: ophthalmic (V1), maxillary (V2), and mandibular (V3). Motor fibers project from the pons, decussate, track adjacent to the trigeminal nucleus, and eventually exit the cranium adjacent to V3 through the foramen ovale. From the foramen ovale, motor fibers separate from sensory fibers and radiate to the masseter and other jaw-moving muscles.

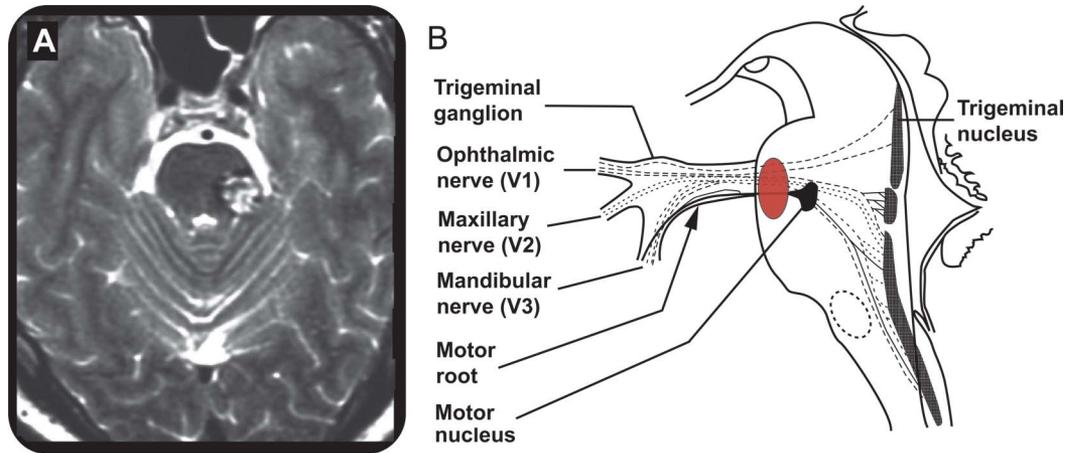
The sensory and motor branches of this patient's nerve were both affected. We therefore infer that an intracranial lesion of the trigeminal nerve must be causing the patient's difficulties. Possible etiologies of the lesion include vascular, infectious, autoimmune, trauma, or a mass. The slow onset of this patient's difficulties decreases the likelihood of vascular lesions and there was no history of trauma. A lesion isolated to the trigeminal nerve would be an unusual presentation of an infectious or autoimmune etiology such as multiple sclerosis, Devic disease, or *Listeria*, and the patient had no current or past symptoms consistent with these illnesses. Considering these factors, the most likely etiology of this patient's isolated trigeminal nerve problem is a central tumor or mass.

Questions for consideration:

1. What testing will narrow this differential?
2. What can be done to help the patient's symptoms?

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Figure MRI and schematic representation of the patient's intracranial lesion



(A) Axial cut at the level of pons, using T2 sequence. There is a left pontine space-occupying lesion. The popcorn appearance of the lesion and the dark rim of signal loss resulting from hemosiderin deposition are consistent with a diagnosis of hemangioma. (B) Diagram represents how an intracranial lesion (in red) would affect both sensory and motor branches of the trigeminal nerve.

SECTION 3

EMG recordings demonstrated fasciculation potentials in the left masseter muscle. Spasms in the left masseter were characterized by increasing frequency of fasciculation discharges, often in bursts, followed by high-frequency discharges of multiple units. Individual motor units fired at frequencies up to 80 Hz (only identifiable with certainty at the beginning and end of spasms). No spasms or abnormal discharges were seen on the right. These EMG results confirmed excess activity of the left trigeminal motor nerve.

Blink reflex was elicited by stimulating the supra-orbital nerve, and blink reflex and jaw jerk studies were normal bilaterally. The masseteric inhibitory reflex was elicited by stimulating the right mental nerve at the mental foramen with a 1-cm bipolar stimulator with

a 0.2-msec duration stimulus at 28.6 mA. Masseteric inhibitory reflexes were tested during voluntary activation and involuntary spasms. During voluntary activation of the masseter, normal S1 and S2 silent periods were evoked by mental nerve stimulation. During involuntary spasms, when the same stimulation was given, there were no silent periods. Occasionally, high-frequency motor unit potentials could be elicited by voluntary muscle activation.

Brain MRI revealed a hemangioma located adjacent to the left pons in the area of the trigeminal nerve exiting zone (figure, A). The lesion was considered inoperable, but the patient experienced substantial relief with a combination of duloxetine and gabapentin. His jaw twitching almost completely resolved and pain was significantly reduced.

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SECTION 4

Given the history, examination, and these electrophysiologic findings, the patient was given a formal diagnosis of hemimasticatory spasm (HMS) caused by a central lesion. HMS is a rare disorder of involuntary brief contractions of masticatory muscles on one side and is thought to result from irritation of the ipsilateral trigeminal nerve. Our case differs from most reported cases in having a central site of origin.¹⁻³

It has been suggested that the pathophysiology of HMS lies in ectopic activation of trigeminal motor axons due to their peripheral irritation and subsequent ephaptic transmission.^{1,4} The notion of peripheral nerve pathology is supported by slowed trigeminal motor nerve conduction as evidenced by delayed jaw jerk reflex latencies,^{1,4} ipsilateral hemiatrophy in some cases,^{5,6} presence of fasciculation potentials, and absent silent period during spasms while being present during voluntary activation.^{1,4} Pathologic involvement of CNS pathways has been suggested in HMS,^{4,7} but on a functional (i.e., remodeling of brainstem pathways by abnormal sensory input), rather than structural (i.e., a CNS lesion), basis. However, in our case, we observed HMS with confirmed structural central origin.

Similarities and differences in clinical presentation between this case and previously reported cases of HMS can be explained by the central origin of the structural abnormality. Similar to our patient, most previously reported cases of HMS had normal blink reflex studies, indicating that ophthalmic branch (V1) of the trigeminal nerve was not affected.^{2,4,5,8,9} Our patient was different in that he had clear sensory deficits that are also attributed to trigeminal nerve lesions—specifically the decreased facial sensation and corneal reflex (figure, B).

Our patient differed from most previous cases of HMS in that he had a reduced corneal reflex and normal jaw jerk. This finding reinforces our conclusion of unique HMS etiology in our patient, as the trigeminal nerve is likely unaffected in the periphery. A focal irritation of the central portion of the trigeminal motor axons in the pons by the hemangioma produces neither measurable delay in reflex latency nor loss of axons, as was demonstrated in this patient.

Dull, long-lasting, and aching pain may accompany HMS and is usually correlated with spasms. However, our patient experienced transient, shock-like pain. We hypothesize that dull pain typically reported in HMS is a consequence of spasms and independent of etiology of cases with peripheral origin. The pain reported by our patient was not associated with spasms, and was likely caused by the direct compression of the central trigeminal sensory axons by the tumor.

The MRI confirmed that the patient had a left pontine hemangioma. We conclude that this case of

HMS was caused by compression of the fifth nerve at its exit from the pons due to that hemangioma. Although the etiology of HMS commonly relates to trigeminal pathology outside the CNS, central causes should also be suspected in patients with HMS. Brain imaging should be recommended in all cases, with particular attention paid to the posterior fossa.

AUTHOR CONTRIBUTIONS

Dr. Mari drafted/ revised the manuscript for content, acquired data, analyzed and interpreted data, and supervised and coordinated study. Dr. Rosenthal drafted/ revised the manuscript for content and analyzed and interpreted data. Ms. Darwin drafted/ revised the manuscript for content and analyzed and interpreted data. Dr. Hallett drafted/ revised the manuscript for content, analyzed and interpreted data, and supervised and coordinated study. Dr. Jinnah drafted/ revised the manuscript for content and analyzed and interpreted data.

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DISCLOSURE

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