Trigeminal Neuralgia Triggered by Auditory Stimuli in Multiple Sclerosis

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Objectives: To describe a patient with a demyelinating brainstem lesion who developed right-sided trigeminal neuralgia triggered by auditory stimuli and to discuss the pathophysiological mechanisms underlying this unusual phenomenon.

Design: Case report.

Setting: Referral center.

Patient: A 27-year-old man who presented with clinical signs of a brainstem lesion developed right-sided trigeminal neuralgia triggered by auditory stimuli to the right ear. Magnetic resonance imaging and electrophysiological studies demonstrated a demyelinating lesion in the pons affecting the right lateral lemniscus and the right trigeminal pathway. This phenomenon completely subsided within 4 days. After a relapse, the diagnosis of clinically definite multiple sclerosis was made.

Conclusion: Lateral spread of impulse activity within the demyelinating pontine lesion is the likely explanation for the unusual phenomenon of trigeminal neuralgia triggered by auditory stimuli.

Arch Neurol. 1999;56:731-733

Trigeminal neuralgia (TN) is an uncommon, but well-recognized symptom of multiple sclerosis (MS). The pathogenesis of TN in patients with MS is still controversial. We describe a patient with MS who presented with signs of brainstem involvement and an unusual form of TN triggered by auditory stimuli. Magnetic resonance imaging (MRI), electrophysiological findings, and pathophysiological implications are discussed.

REPORT OF A CASE

A 27-year-old, previously healthy man with a 5-day history of unstable gait was admitted to the hospital. One day before admission, he had noticed a numbness of the right side of his face and a disturbance of taste. On examination, he had a horizontal spontaneous nystagmus to the left side, hypesthesia and tingling paresthesias in the territory of the second and third division of the right trigeminal nerve, and a loss of taste for sweet and salty on the anterior aspect of the right side of the tongue. There was no facial weakness. Tendon reflexes, strength, and muscle tone were normal. When standing and walking, the patient tended to fall to the right side, and his right extremities were ataxic. No sensory dysfunction of the limbs or trunk was detected. The findings of the general examination were unremarkable.

Based on clinical, laboratory, MRI, and electrophysiological findings, the diagnosis of a disseminated encephalitis was made, and the patient was treated with intravenous high-dose prednisolone hydrochloride (500 mg/d) for 5 days, without any improvement. Three days after beginning therapy, the patient had an acute attack of electrifying, shooting pain radiating from the right temple to the right lower jaw area that occurred when the telephone in the patient's room began to ring. The pain lasted for approximately 1 second and recurred every time the telephone rang. It was also triggered by the ringing of the hospital's bell indicating the time when visitors have to leave the hospital. The trigeminal pain did not occur spontaneously and could not be triggered by speaking, chewing, or touching. When the right ear of the patient was covered, the phenomenon could not be reproduced. Three days after onset, the facial pain improved, and the following day, it subsided completely and did not recur during the next 3 months.
The patient's general neurologic condition progressively improved after the corticosteroid therapy. Four weeks after his admission to our hospital, the patient was neurologically asymptomatic except for a hypesthetic area on the right side of the chin. Two weeks later, he suffered a relapse, with hypesthesia of the left side of his face and burning dysesthesias in the distal aspect of the right limbs. The patient was again treated with high-dose prednisolone and recovered completely.

The results of routine laboratory studies were normal. Examination of a cerebrospinal fluid (CSF) sample disclosed a mild pleocytosis (6 lymphocytes per cubic millimeter), with normal protein content and normal CSF IgG index; oligoclonal bands were not detected. Serologic tests of CSF and serum samples were negative for *Borrelia burgdorferi*, human immunodeficiency virus I and II, and *Treponema pallidum*.

An MRI study was performed on a 1.5-T unit. The T2-weighted turbo–spin-echo and fluid attenuated inversion recovery (FLAIR) images showed multiple hyperintense lesions in the white matter of the paraventricular area. Also, a large plaque was seen in the right pons near the middle cerebellar peduncle (Figure). Brainstem auditory evoked potentials (BAEPs) were performed using standard methods. Waves I-V were identified with normal absolute latencies and normal absolute interwave latencies I-III, III-V, and I-V, but on side-to-side comparison, interwave latency III-V (normal absolute latency, <2.3 milliseconds [msec]; normal side difference, <0.4 msec) was delayed on the right side (1.52 msec on the left side vs 2.24 msec on the right side; side difference, 0.72 msec). Trigeminal somatosensory evoked potentials were obtained by unilateral stimulation of both lips and recording from the scalp contralaterally at C5 and C6 (10-20 system). The P19 response (normal absolute latency, <22.3 msec; normal side difference, <1.9 msec) had a normal latency on stimulation of the left side (17.0 msec) and was delayed on stimulation of the right side (24.0 msec). Visual evoked potentials were significantly delayed on the right side (P100, 158 msec on the right side vs 90 msec on the left side). The findings of the otolaryngological and audiometric examinations were normal, as was the stapedius reflex.

**COMMENT**

The course of our patient’s disease fulfills the criteria of clinically definite MS. The patient presented with clinical signs of brainstem involvement and an unusual form of paroxysmal facial pain. This facial pain had clinical features of TN affecting the third division of the right trigeminal nerve, but occurred only when triggered by auditory stimuli to the right ear. To our knowledge, a similar phenomenon has not been reported previously. The pain subsided within 4 days. We cannot be certain whether the improvement was a result of the corticosteroid treatment or a spontaneous remission.

Usually, TN in patients with MS is clinically indistinguishable from “idiopathic” TN, occurring spontaneously or triggered by mechanical means such as speaking, chewing, or touching. There is still controversy about whether TN associated with MS is caused by vascular compression of the trigeminal nerve or by pontine lesions implicating trigeminal nerve fibers. Iragui et al described a patient with MS whose clinical symptoms of right-sided TN were...
accompanied by abnormalities in BAEPs from the right ear. They postulated that the cause was a pontine lesion involving the right trigeminal sensory root and the right lateral lemniscus. They concluded that, because of the close anatomic proximity of these structures, a single lesion could be responsible for both the TN and the BAEP abnormalities.

In our patient, MRI revealed a plaque in the right pons near the middle cerebellar peduncle reaching in its medial anterior portion to the region of the lateral lemniscus. In BAEPs from the right ear, interwave latency III-V was delayed compared with that from the left ear. Because waves III and V are generated in the low pons (superior olivary complex) and in the high pons or low midbrain (lateral lemniscus or inferior colliculus), respectively, and inputs of BAEP generators seem to ascend the brainstem ipsilaterally, this finding indicates a demyelinating lesion of the right lateral lemniscus. The delay of the trigeminal somatosensory evoked potentials after stimulation of the right trigeminal nerve could be due to either a central or a peripheral lesion, but the clinical picture argues strongly in favor of a central lesion. Taken together, the MRI and electrophysiologic findings point to a pontine plaque causing a demyelinating lesion of the right lateral lemniscus and the right trigeminal sensory pathway, as in the case reported by Iragui et al.9

However, in contrast to the previously reported case,9 the TN in our patient occurred only when triggered by auditory stimuli to the right ear. We assume that this phenomenon is due to cross excitation between fibers of the auditory and the trigeminal pathway within the demyelinating lesion in the right pons. Clinical observations of paroxysmal phenomena associated with MS provide evidence that lateral spread of impulse activity may occur within demyelinated plaques. Besides the observation that some paroxysmal positive symptoms are provoked by specific movements or specific sensory stimuli, the frequently sequential occurrence of positive symptoms attributable to adjacent tracts in central white matter is clinical evidence of lateral spread of excitation within areas of demyelination.10 For example, Osterman and Westerberg11 reported 2 cases of spinal sensorimotor seizures with repeated attacks of unilateral tonic spasm, either followed or preceded by a contralateral sensation of heat and pain. They concluded that this phenomenon could be explained by a transversely spreading activation of axons within a lesion in the spinal cord involving the lateral corticospinal tract and the lateral spinothalamic tract. Experimentally, the possibility of ephaptic transmission between adjacent pathologically myelinated axons was demonstrated by single-fiber studies on peripheral nerve roots.10 However, the paroxysmal trigeminal pain provoked by auditory stimuli in our patient presumably reflects simultaneous activation of large numbers of fibers. Whether such multifiber interactions within the central nervous system are mediated electrically, i.e., by ephaptic transmission in the strict sense, or chemically, remains to be determined experimentally.10

Although TN associated with MS usually occurs years after the onset of MS, it can also be the only presenting symptom,1,2 thus masquerading idopathic TN. Triggering of the facial pain by auditory stimuli, as in our patient, may suggest central involvement and requires further neurologic evaluation.

Accepted for publication March 2, 1998.

We thank Manfred Stoehr, MD, and Lea Averbuch-Heller, MD, for helpful discussions and for critically reading the manuscript.

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REFERENCES