An Unusual Case of Simultaneous Bilateral Trigeminal Neuralgia Due to Multiple Sclerosis

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Although several reports have indicated that trigeminal neuralgia related to multiple sclerosis may occur bilaterally in the orofacial region, trigeminal neuralgia pain usually involves the two sides in different time lapses, and the simultaneous involvement of trigeminal territories on both sides is commonly considered incompatible with its diagnosis. This case report describes a patient with bilateral trigeminal neuralgia related to multiple sclerosis that started simultaneously on both sides of the orofacial region. A 55-year-old man presented with a 16-year history of relapsing/remitting multiple sclerosis. For 1 year, the patient had been complaining of electric shock–like, paroxysmal pain of severe intensity that lasted from a fraction of a second to a few minutes and involved the first and second trigeminal divisions of both sides simultaneously. The neurophysiologic testing of trigeminal reflexes showed bilateral delayed latencies of reflex responses compatible with a trigeminal afferent pathway impairment related to multiple sclerosis. A dedicated 3T magnetic resonance imaging scan revealed pontine demyelinating plaque and a bilateral neurovascular conflict at the trigeminal root entry zone. The finding of an unusual case of simultaneous bilateral trigeminal neuralgia due to multiple sclerosis should prompt neurologists to consider a diagnosis of trigeminal neuralgia in patients with multiple sclerosis in cases of simultaneous involvement of trigeminal territories on both sides. *J Oral Facial Pain Headache 2017;31:e4–e6. doi: 10.11607/ofph.1831*

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Trigeminal neuralgia (TN) is a clinical condition characterized by sudden unilateral, electrical shock–like recurrent pain with a distribution consistent with one or more divisions of the fifth cranial nerve.[1,2]

The new classification and diagnostic grading for practice and research from the Special Interest Group on Neuropathic Pain (NeuPSIG) of the International Association for the Study of Pain (IASP) distinguishes between classical TN caused by vascular compression producing morphologic changes in the trigeminal nerve root, secondary TN due to an identifiable underlying neurologic disease, and idiopathic TN.[3]

Secondary TN is frequently related to multiple sclerosis (MS). Patients suffering from MS have a 20-fold increased risk of developing TN.[4] MS-related trigeminal neuralgia has long been attributed to a demyelinating plaque affecting the trigeminal root entry zone in the pons. Nevertheless, a recent study showed that in many patients with MS-related TN, a symptomatic neurovascular compression concurs with the trigeminal root entry zone plaque in the manifestation of TN.[5]

Although MS-related TN is usually unilateral, some cases of MS-related bilateral TN have been reported in the literature.[5,6] In all these cases, however, TN affected both sides at different time lapses (years); ie, without simultaneous involvement of the right and left trigeminal nerves. The present report describes an unusual case of a patient with MS and simultaneous occurrence of bilateral TN.

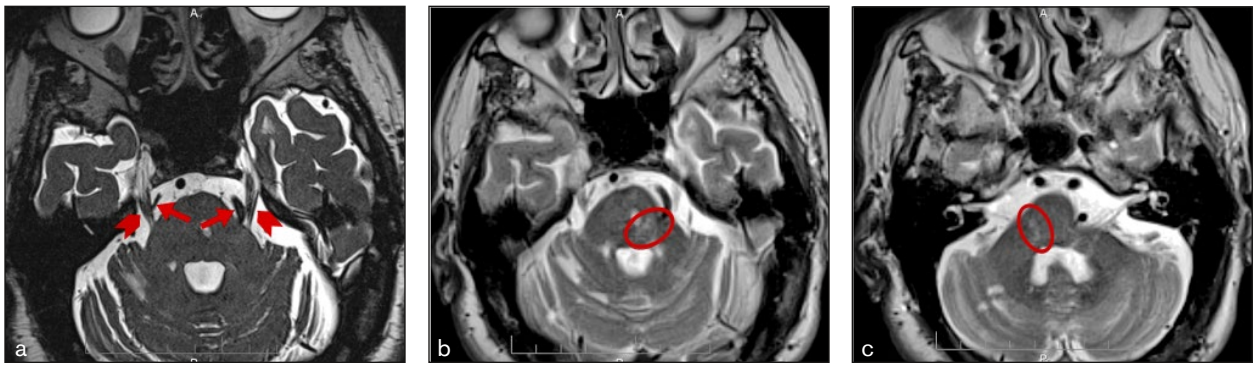

Fig 1 The MRI 3D constructive interference in steady state (3D-CISS) image **(a)** shows the trigeminal nerve course (*marked by arrowheads*) and an arterial loop consisting of the superior cerebellar artery on the right side and the vertebral artery on the left side (*arrows*). The MRI TSE T2-weighted images show the demyelinating plaque (*circle*) involving **(b)** the trigeminal root entry zone and **(c)** the caudal and posterior pontine portion.

Case Presentation

A 55-year-old man presented with a 16-year history of relapsing/remitting MS. His medical history was unremarkable for other concomitant diseases. At onset, MS was associated with paresthesia of the left leg. He is currently unable to walk and needs a wheelchair, and is being treated with glatiramer acetate.

For 1 year, the patient had been complaining of electric shock–like, paroxysmal pain lasting from a fraction of a second to a few minutes that was characterized by severe intensity and distributed in the first and second trigeminal divisions on both sides simultaneously. Paroxysms manifested spontaneously or when evoked by innocuous tactile stimuli in specific facial or intraoral areas when the patient was eating or speaking.

A precise sensory profiling was performed by using bedside tools. Touch was investigated with a piece of cotton wool, and pinprick sensation with a wooden cocktail stick. Both negative (tactile, thermal, and pricking hypoesthesia) and positive (paroxysmal pain, ongoing pain, dysesthesia, mechanical allodynia, and pinprick hyperalgesia) symptoms were examined.[7] Clinical examination revealed tactile hypoesthesia only in the left trigeminal territory. Conversely, no positive symptoms were found, with the exception of electric shock–like paroxysms evoked by touching trigger zones on both sides (the ala of the nose and the upper lip). The remaining clinical and neurologic examinations showed right hemiplegia with severe spastic hypertonia.

The patient underwent trigeminal reflex testing to detect trigeminal pathway damage, a mandatory item for diagnosing MS-related TN according to the new classification and diagnostic grading of the IASP. Trigeminal reflex testing included recordings of the blink reflex evoked by electrical stimulation of the supraorbital nerve and the masseter inhibitory reflex evoked by electrical stimulation of the infraorbital and mental nerves.[8] The methods used adhered to the Recommendations for Clinical Practice of the International Federation of Clinical Neurophysiology.[9] The latencies of all trigeminal reflex responses were delayed (R1 component of the blink reflex: 12.8 milliseconds on the right side and 16.6 milliseconds on the left; SP1 component of the masseter inhibitory reflex: 17.0 milliseconds on the right and 22.0 milliseconds on the left).

The patient underwent anatomical MRI acquired with a 3-Tesla magnet (Siemens Verio) equipped with a 12-channel head coil. A previously described magnetic resonance imaging (MRI) protocol was used.[5,10] In brief, this protocol included a T1-weighted image (repetition time [TR]: 2,300 milliseconds, repetition echo [TE]: 2.98 milliseconds, inversion time [TI]: 900 milliseconds, flip angle: 9 degrees, field of view [FOV]: 240 × 256 mm, 208 slices, 1 mm isometric voxel), turbo spin echo (TSE) T2-weighted images (TR: 6,000 milliseconds, TE: 104 milliseconds, flip angle: 150 degrees, FOV: 220 mm, slice thickness: 3 mm), a TSE double-echo proton density and T2-weighted image (TR: 4,000 milliseconds, TE: 9.6/96 milliseconds, FOV: 220 mm, slice thickness: 3 mm, gap: 0 mm, 50 slices matrix = 384 × 384), and a 3D constructive interference in steady state (3D-CISS) image (TR = 1,000 milliseconds; TE = 132 milliseconds; FOV= 200 mm; slice thickness = 0.5 mm; slice number = 56; matrix = 384 × 384), as well as a 3D time-of-flight magnetic resonance angiography (3D-TOF-MRA) scan (TR = 22 milliseconds; TE = 3.60 milliseconds; FOV = 200 mm; slice thickness = 0.5 mm; matrix = 384 × 384). The MRI scans documented on the left side a demyelinating plaque involving the trigeminal root entry zone and on the right side a relatively more caudal and posterior pontine lesion that coexisted with neurovascular conflict (ie, a direct contact between arteries and the proximal portion of the nerve) on both sides (Fig 1).

The patient started a treatment with oxcarbazepine (1,800 mg/day) and achieved satisfactory control over the paroxysms without significant adverse events.[11,12]

Discussion

Although several reports have indicated that MS-related TN may occur bilaterally, pain usually involves the two sides at different time lapses, and the simultaneous involvement of the trigeminal territories of both sides is commonly considered an exclusion criterion for the diagnosis of TN. This case report describes the previously unreported finding of a bilateral, MS-related TN with paroxysmal pain occurring simultaneously on both sides of the orofacial region. In patients with MS, therefore, a diagnosis of TN should also be considered in cases of simultaneous bilateral orofacial pain.

How MS-related TN should be defined remains a controversial topic. Although the diagnostic criteria of the latest International Headache Society classification for diagnosing TN has been taken into account, the proposed definition (painful TN attributed to MS plaque) was avoided in the present report. Rather, the recently proposed definition issued by the NeuPSIG of the IASP, which identifies secondary TN, was considered.[3] The definition of TN was considered misleading because it identifies conditions that are distinct from TN and manifest completely different clinical features, such as pain associated with acute herpes zoster. Conversely, secondary TN is a well-established and widely accepted definition that avoids confusion.

Reaching a definitive diagnosis of MS-related TN inevitably depends on detecting trigeminal pathway impairment. In the present case, trigeminal reflex testing elicited abnormal responses, showing impairment of the trigeminal pathway and confirming the diagnosis of MS-related TN. Trigeminal reflex testing has a specificity and sensitivity approaching 90% in the diagnosis of symptomatic TN.[14] Therefore, trigeminal reflex testing cannot exclude coincident classical TN, a condition that usually spares trigeminal reflex responses.

The MRI investigation of the patient revealed a pontine demyelinating plaque probably affecting the trigeminal root entry zone on the left side, as the pontine demyelinating lesion on the right side was located in a more caudal and posterior area. Since the MRI scans showed bilateral neurovascular conflict, it can be hypothesized that in this patient the pontine plaque and the coexisting neurovascular conflict caused TN through a double-crush mechanism, namely inflammatory demyelination and mechanical demyelination in the same first-order neurons.[15] This hypothesis receives support from clinical findings showing that TN started in this patient when he was 52 years old, an age potentially compatible with classic TN.[16] However, given that the MRI investigation showed demyelinating plaque in a pontine area on the right side, which is usually considered irrelevant to MS-related TN, the hypothesis that the neurovascular compression on the right side in this patient acted as the main factor causing TN cannot be excluded.

Conclusions

This case report describes the previously unreported finding of simultaneous occurrence of bilateral MS-related TN. In patients with MS, the diagnosis of TN should also be considered in cases of paroxysmal pain simultaneously involving the trigeminal divisions on both sides of the orofacial region.

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