TRIGEMINAL SENSORY NEUROPATHY ASSOCIATED WITH SYSTEMIC SCLEROSIS

Report of three brazilian cases

Reinaldo Teixeira Ribeiro¹, Libânia Melo Nunes Fialho¹,
Larissa Teles de Souza¹, Orlando Graziani Póvoas Barsottini²

Systemic sclerosis (SSc) is a connective tissue disease manifest by fibrotic tissue changes, microvascular disease, and autoimmune abnormalities. The prevalence of different neurological manifestations in SSc has ranged from 0.8%¹ to 18.5%² according to the adopted criteria.

Trigeminal sensory neuropathy (TSN) causes numbness in the mandibular or maxillary divisions of the nerve in about 2/3 of the cases, and in the distribution of all divisions in the remaining cases. The sensory abnormalities evolve slowly and usually spread contralaterally in an asymmetric pattern. The numbness may be accompanied by burning dysesthesia that is distinct from trigeminal neuralgia³.

TSN is an infrequent complication of SSc. Although epidemiological studies are scarce, the prevalence of TSN associated with SSc in the largest series available was 4%⁴. A fairly diligent review of the literature revealed no previous report of TSN as a complication of SSc in Brazil. We present three cases of such association.

CASES

Case 1

A 42-year-old male had presented progressive facial hypoesthesia and dysesthesia for the last three years and was diagnosed with the diffuse form of SSc two years ago. At his first neurological evaluation, he had sclerodermic fascies, sclerodactyly, Raynaud's phenomenon, bilateral facial and lingual hypoesthesia, left hemifacial dysesthesia, and an absent left corneal reflex. Nailfold capillaroscopy was performed and showed devascularization and ectasias. ANA was positive (1/640) and anti-RNP was also positive. Nerve conduction studies (NCS) revealed sensory myelinic polyneuropathy in all limbs and face. Blink reflex studies (BRS) demonstrated absent responses on both sides to left supraorbital nerve stimulation. Electromyography (EMG) was

normal. He was on methotrexate and prednisone without improvement of the TSN. Gabapentin 800 mg daily and nortryptilin 100 mg daily were introduced with only a partial response at their maximum tolerable doses.

Case 2

A 47-year-old female had presented Raynaud's phenomenon and arthritis in her hands for the last three years and was feeling progressive numbness in the inferior half of the face for the last two years, without any previous diagnosis. At her first neurological evaluation, she had sclerodermic fascies, sclerodactyly, hypoesthesia in the maxillary and mandibular divisions territory bilaterally. ANA was positive (1/80), an esophagogram showed an increased esophageal caliber and a thoracic tomography confirmed this finding and revealed signs of pulmonary fibrosis. All electrophysiological studies performed (NCS, BRS, EMG) were normal. She had never been on medication for TSN or SSc yet.

Case 3

A 50-year-old female had presented hypoesthesia only in the maxillary division of the right trigeminal nerve about three years before her first neurological evaluation. Six months later, she started presenting hypoesthesia in the mandibular division, effort dyspnea, Raynaud's phenomenon and myalgia. She was diagnosed with SSc in overlap with myositis 10 months before our first evaluation. At that time, she presented digital ulcers, sclerodactyly, right hemifacial and lingual hypoesthesia, and an absent right corneal reflex. NCS were normal. BRS demonstrated absent responses on both sides to right supraorbital nerve stimulation. EMG showed polyphasic, low amplitude, and short duration action potentials of the motor units within the orbiculari oris muscles. She had to stop methotrexate treatment due to adverse effects, and then she was started on prednisone 20 mg daily and amitriptyline 50 mg daily without improvement.

NEUROPATIA SENSITIVA TRIGEMINAL ASSOCIADA COM ESCLEROSE SISTÊMICA: RELATO DE TRÊS CASOS BRASILEIROS

¹MD, Department of Neurology, Federal University of São Paulo São Paulo SP, Brazil; ²MD, PhD. Department of Neurology, Federal University of São Paulo, São Paulo SP, Brazil.

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Dr. Reinaldo Teixeira Ribeiro – Rua Borges Lagoa 71 / 93 - 04038-030 São Paulo SP - Brasil. E-mail: reinaldo_ribeiro@yahoo.com.br

In all three cases, cranial magnetic resonance imaging (MRI) was normal and a written informed consent was obtained.

DISCUSSION

The increased likelihood of TSN to affect females with SSc in their fourth and fifth decade of life indicated in our small report is probably due to the prevalence pattern of SSc in the general population as shown in a large epidemiological study⁵. Furthermore, there was no significant difference in the sex distribution between the patients with TSN and those without TSN in the largest series available⁴.

Alike previous case reports, our series indicate that TSN complaints usually begin with the onset of facial numbness, with or without pain or paresthesias. This symptom may precede the first indication of clinically active SSc, but generally follows the latter by a matter of months. The associated SSc is not usually attended by rapidly progressive life-threatening neurologic or systemic disease^{4,6}.

In our three patients with TSN, the facial sensory deficit was prominent. Similarly to what is publicized in the literature, the initial involvement was confined to the distal portions of the second and third divisions of the trigeminal nerve. Although a corneal reflex was absent in two patients at the time of neurological examination, the facial paresthesias and sensory deficit were more apparent in the perioral region. The motor division of the trigeminal nerve was spared in all patients^{3,4,7,8}.

BRS is the standard test to assess trigeminal function. Although it demonstrated an afferent pattern of abnormal responses in two patients (Cases 1 and 3), normal results of BRS in TSN (as seen in Case 2) were reported previously.⁹ TSN has been observed most frequently in CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia) or SSc in overlap with other disorders, like myositis. The antinuclear and anti-RNP antibodies have frequently been found in SSc with TSN.⁴

The distribution of sensory loss found in SSc with TSN is not consistent with the somatotopic arrangement of the nucleus of the spinal tract of the trigeminal nerve and almost invariably fails to respect trigeminal division-

al and cutaneous nerve territories. It is speculated that the disease process lies in the Gasserian ganglion or sensory root of the trigeminal nerve, as reflected on MRI by abnormal contrast uptake and slight enlargement of its pre-ganglionic segment, which resolves with the resolution of the early edematous stage of SSc¹⁰. In most instances, TSN does not respond to antineuralgic or immunosuppressive therapy⁸.

We report three cases of TSN associated with SSc. We would like to emphasize that the involvement of the nervous system may occur in SSc and it is not so uncommon. TSN is probably underdiagnosed in these patients as it can easily be confused with symptoms of sclerotic skin stiffness. Careful and repeated investigation of an underlying illness should be required as TSN may antedate the active phase of many connective tissue diseases such as SSc.

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