Paroxysmal facial pain in a patient with Parry Romberg Syndrome

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Objective

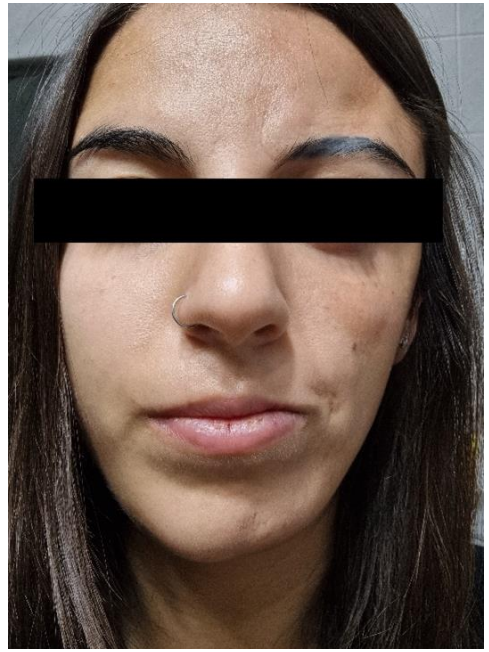
Parry Romberg Syndrome (PRS) is a rare disease, characterized by hemiatrophy of the skin and facial soft tissues, it can affect muscles, cartilage and bones.
The cause has not been identified. It has been related to trauma, vascular malformations, infections, immune-mediated processes and alterations of the sympathetic nervous system.
Neurological clinical manifestations occur in approximately 20% of cases, headaches and facial pain are some of them.
The objective is to describe the clinical characteristics of facial pain in a patient, report its association with PRS and response to treatment.

Case Description

A 27-year-old woman with a diagnosis of PRS and a history of bruxism and episodic migraine without aura, controlled with ibuprofen, referred stabbing and paroxysmal pain of 7 years of evolution. Pain was in the left temporal and maxillary region, lasting from seconds to 1 minute, moderate intensity and some episodes associated with homolateral temporal and masseter muscle spasm. Frequency of 5 times a day. She referred another sharp and severe left eye pain, lasting 2 hours with ipsilateral congestion and rhinorrhea lasting 24 hours, up to 2 crises per day and 10 crises per month, with response to indomethacin.
Physical examination revealed left facial hemiatrophy, episodic homolateral temporal and masseter muscle spasm and preserved sensitivity in the trigeminal territory.
Brain magnetic resonance imaging (MRI): isolated nonspecific white matter punctiform lesions. Magnetic resonance angiography of intracranial arteries was normal. Magnetic resonance neurography of the cranial nerves showed hypertrophy of the left muscles of mastication, except for left masseter which was smaller on the left side., decreased volume of the submandibular gland and decreased thickness of the soft tissues. Thinning of the hemifacial and hemicranial bone structures as well as of the left soft tissues was observed.
Cranial nerves presented normal caliber, course and signal. Temporomandibular joint (TMJ) MRI: dysfunction of both TMJs. Electromyogram of the left masseter and temporal muscles: when presenting the masticatory spasm, contraction of the temporal muscle was observed, followed sequentially by the masseter before ending.
Gabapentin treatment was started without improvement. Treatment with botulinum toxin, under EMG guidance of the left masseter and temporal muscles was started with 80% improvement in pain and spasm.

Conclusion

The association between PRS and hemimasticatory spasm is rare and is associated with compromise of the trigeminal nerve.
It is described that the sympathetic hyperactivity produced by an inflammatory process affects the blood vessels and the cranial nerves and this would cause tissue damage. Due to the atrophy of the soft tissues, mainly of the masseter and temporalis muscles, there is a focal demyelination of the peripheral branches of the trigeminal nerve, which causes an abnormal excitation of the fiber and consequently facial pain.
This patient had two different kinds of pain, paroxistic facial pain and a longer lasting ocular pain with autonomic signs probably as a consequence of trigeminal involment. More studies are needed to elucidate the cause.



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