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# Bilateral painful trigeminal neuropathy as an initial manifestation of systemic sclerosis: a case report

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**Introduction**
Painful trigeminal neuropathy is characterized by signs of trigeminal nerve dysfunction combined with chronic pain, which is typically unilateral and non-neuralgiform. Bilateral painful trigeminal neuropathy is uncommon and should raise suspicion for connective tissue diseases, especially systemic sclerosis.

**Case presentation**
A 67-year-old woman presented with a history of burning pain and paresthesia in the left malar region, with gradual progression over the years to bilateral involvement of the lower two-thirds of the face. Neurological examination revealed bilateral facial hypoesthesia without other focal deficits. Brain MRI showed no structural abnormalities. Systemic examination demonstrated Raynaud phenomenon, sclerodactyly, and microstomia. Further investigation confirmed the diagnosis of systemic sclerosis and pulmonary arterial hypertension. Pain symptoms were satisfactorily managed with gabapentinoids.

**Discussion**
Trigeminal neuropathy associated with systemic sclerosis is rare but well described, typically presenting with sensory-predominant features and an insidious course. Bilateral involvement, although uncommon, is an important clinical clue suggesting a systemic etiology. The underlying pathophysiology involves microvascular changes and collagen deposition, leading to peripheral nerve damage. Recognition of associated clinical features, such as Raynaud phenomenon and sclerodactyly, is essential to guide further investigation. Normal neuroimaging may contribute to diagnostic delay, reinforcing the importance of comprehensive clinical assessment. In addition to systemic sclerosis, other connective tissue diseases should be considered in the differential diagnosis of painful trigeminal neuropathy, such as sarcoidosis and Sjogren's syndrome.

**Conclusion**
The presence of bilateral painful trigeminal neuropathy should prompt investigation for connective tissue diseases. Early recognition of systemic features allows timely diagnosis of systemic sclerosis and appropriate management of its associated complications. The clinical relevance of this case lies in the neurologist's role in suspecting systemic sclerosis due to the facial pain phenotype. This has led to early identification of pulmonary arterial hypertension, a potentially life-threatening visceral complication of systemic sclerosis. Neurologists and headache and facial pain specialists should actively pursue systemic signs in individuals with bilateral trigeminal neuralgia. Raynaud phenomenon, sclerodactyly and microstomy exemplify relevant findings to look for.

**Keywords:**
Trigeminal neuropathy
Systemic sclerosis
Connective tissue diseases
Facial pain

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