To the Editor:

Trigeminal motor neuropathy is a rare condition presenting with muscle weakness and atrophy in the distribution of the trigeminal nerve without sensory changes. We present a challenging case with clinical features that mimic progressive hemifacial atrophy (PHA), a disease characterized by slowly progressive, unilateral facial atrophy that can be accompanied by inflammation, fibrosis, or atrophy of the skin or subcutaneous fat as early features.

A 55-year-old man presented with right-sided ptosis and progressive right-sided facial atrophy of 4 years' duration. A clinical diagnosis of PHA was made by the rheumatology department, and the patient was referred to the dermatology department for further evaluation. Examination at presentation revealed right-sided subcutaneous atrophy of the cheek, temple, and forehead extending to the scalp with absence of sclerosis, pigmentary alteration, or typical linear morphea lesions (Figures 1 and 2). The patient had no sensory changes in the affected area.

Workup by the dermatology department included magnetic resonance imaging (MRI) of the face and scalp, which demonstrated denervation muscle atrophy exclusively in the distribution of the third branch of the right trigeminal nerve, including severe atrophy of the right temporalis and masseter muscles and moderate atrophy of the pterygoid muscles. No signs of inflammation, fibrosis, or atrophy of the skin or subcutaneous fat were found, ruling out a diagnosis of PHA.

PRACTICE POINTS

• The differential diagnosis of progressive hemifacial atrophy includes disorders of the trigeminal nerve.
• Trigeminal motor neuropathy presents with muscle weakness and atrophy without involvement of the skin, subcutis, or bone.
The patient was referred to the neurology department where he was found to have a normal neurologic examination with the exception of right-sided ptosis and temporalis and masseter muscle atrophy. Notably, the patient had normal sensation in the distribution of the trigeminal nerve and normal strength of the masseter and temporalis muscles.

An extensive workup by the neurology department was completed, including magnetic resonance angiography, eyelid testing, and testing for causes of neuropathies (e.g., infectious, autoimmune, vitamin deficiencies, toxin related). Of note, magnetic resonance angiography showed no abnormalities within the cavernous sinus or trigeminal cave but showed potential vascular compression of the trigeminal nerve, which was believed to be an incidental finding. The remainder of the workup was unremarkable. Based on muscle denervation atrophy in the distribution of the third branch of the trigeminal nerve in the absence of sensory symptoms or deficits, the patient's presentation was consistent with trigeminal motor neuropathy.

In reported cases, the pathogenesis of trigeminal motor neuropathy is attributed to tumors, trauma, stroke, viral infection, and autoimmune reaction. In other reported cases, the cause is unknown as was the case in our patient. Magnetic resonance angiography revealed potential vascular compression of the trigeminal nerve, which has been previously reported to cause trigeminal neuropathy. However, patients with trigeminal neuropathy presented with sensory changes in the distribution of the trigeminal nerve as opposed to motor symptoms and muscle atrophy.

We present a case of trigeminal motor neuropathy presenting as PHA. Progressive hemifacial atrophy is a rare, slowly progressive disease characterized by unilateral atrophy of the skin, subcutis, muscle, and bony structures of the face. Onset usually is during childhood, though later onset has been reported. The pathogenesis of PHA is not well understood, though trauma, infection, immune-mediated causes, sympathetic dysfunction, and metabolic dysfunction have been proposed. Diagnosis of PHA typically is based on clinical presentation, but histology and imaging are useful. In contrast to trigeminal motor neuropathy, MRI findings in PHA demonstrate involvement of the skin.

Differentiation between PHA and trigeminal motor neuropathy is important because treatment differs. Treatment of trigeminal motor neuropathy depends on the etiology and may include removal of underlying neoplasms, while treatment of PHA depends on disease activity. The initial goal when treating PHA is to improve symptoms and slow disease progression; immunosuppressants may be considered. Facial reconstruction is an option when PHA is stable.

In this case, the features differentiating trigeminal motor neuropathy from PHA include age of onset and MRI as well as clinical findings of muscle atrophy limited to the distribution of the third branch of the trigeminal nerve. Although PHA is a rare disorder, this case demonstrates the importance of including trigeminal motor neuropathy in the differential diagnosis.

REFERENCES