Parry-Romberg Syndrome and Sympathectomy—A Coincidence?

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Parry-Romberg syndrome is a clinical entity consisting of progressive hemifacial atrophy appearing at a young age. Animal studies indicate that sympathectomy can produce hemifacial atrophy. To our knowledge, this is the first report of a patient with a possible association between Parry-Romberg syndrome and thoracoscopic sympathectomy.

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Parry-Romberg syndrome, or progressive hemifacial atrophy, is a clinical entity consisting of facial changes involving the paramedian face that usually appear in the first decades of life. The atrophy tends to slowly progress to involve the muscle, bone, and cartilage. There may be neurologic sequelae such as ptosis, inability to close the eye, trigeminal neuralgia, migraine, and, at a later stage, even epilepsy. In extreme cases, facial hemiatrophy may spread to lower ipsilateral body regions. The overlying skin often becomes hyperpigmented.[1] Diagnosis is based on clinical features, and radiographic results showing jaw atrophy. Some authors consider the disease a variant of morphea because the histologic changes are identical to deep scleroderma.[2] The possible etiologies include sympathetic denervation, trauma, vascular malformations, immunologic abnormality, hereditary disease, or infection by a slow virus.[3] To our knowledge, this is the first report of a young patient with a possible association between Parry-Romberg syndrome and thoracoscopic sympathectomy.

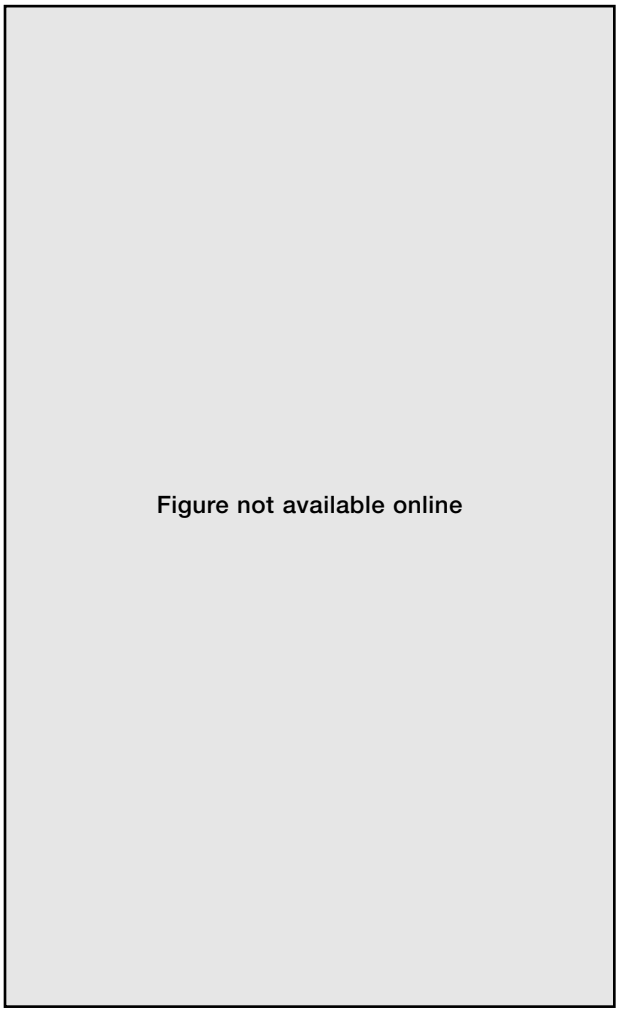

Figure 1. Left hemifacial atrophy with sunken cheek and jaw asymmetry.

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Case Report

A 17-year-old boy presented with progressive facial asymmetry. At age 13 years, he underwent thoracoscopic sympathectomy for palmar hyperhidrosis. The patient had slight facial asymmetry since childhood, but a few months after the operation, his mother noted a rapid progression of the asymmetry. His medical history was otherwise unremarkable.

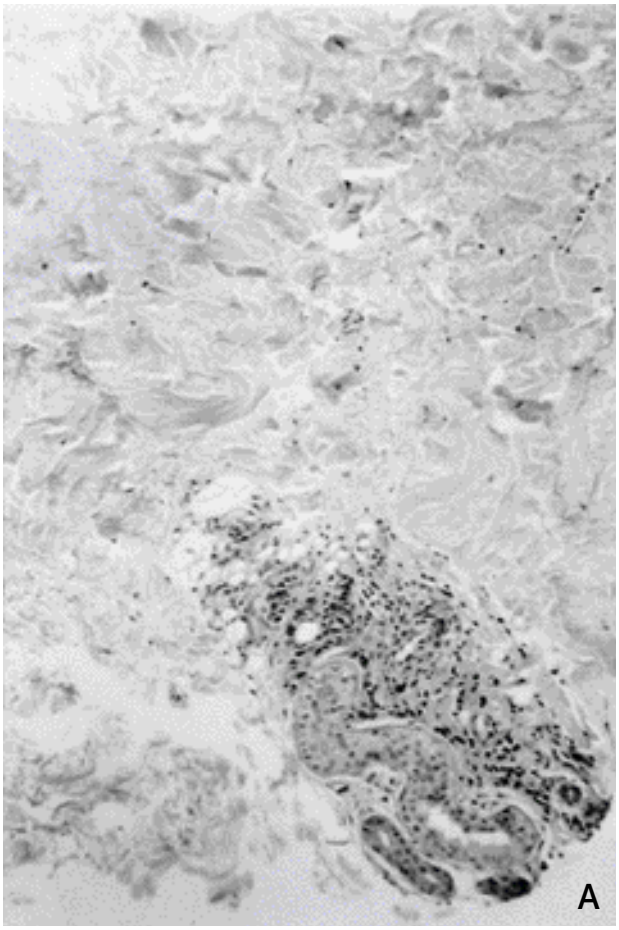
Results of a physical examination revealed left hemifacial atrophy of all soft tissue, with temporal wasting, sunken cheek, and smaller hemijaw (Figure 1). On the anterior left neck, hyperpigmented plaques were notable. Neurologic examination results were within reference range.

Results of a biopsy specimen from the temporal area revealed fibrosis extending from the reticular dermis to the temporal muscle with encroachment on the sweat glands and nerve fibers (Figure 2). The pathologic changes were compatible with deep scleroderma, supporting the clinical diagnosis of Parry-Romberg syndrome. Laboratory investigations, including tests of complete blood count, serum chemistry, antinuclear antibody, complement level, Scl-70 antibody, and thyroid function, were all within reference range.

Comment

The pathogenesis of Parry-Romberg syndrome may involve an alteration in peripheral sympathetic stimulation.[1] Resende et al[4] ablated the superior cervical sympathetic ganglion unilaterally in 1-month-old rabbits, cats, and dogs. After a year of follow-up, the researchers noted ipsilateral hemifacial atrophy, slight bone atrophy, as well as ocular atrophy, keratitis, and alopecia. The authors concluded that cervical sympathectomy in animals produced clinical alterations similar to Parry-Romberg syndrome.[4] Sherman and Chole[5] performed unilateral cervical sympathectomy in animals and showed an increase in unilateral bone resorption due to enhanced osteoclast activity. Pai-Silva et al[6] showed that cervical sympathectomy caused fibrosis and atrophy of the rabbit masseter muscle.

Theoretically, thoracoscopic sympathectomy may cause 2 of the aforementioned etiologies of Parry-Romberg syndrome: sympathetic denervation and trauma. Thoracoscopic sympathectomy is a surgical technique for the treatment of palmar hyperhidrosis.



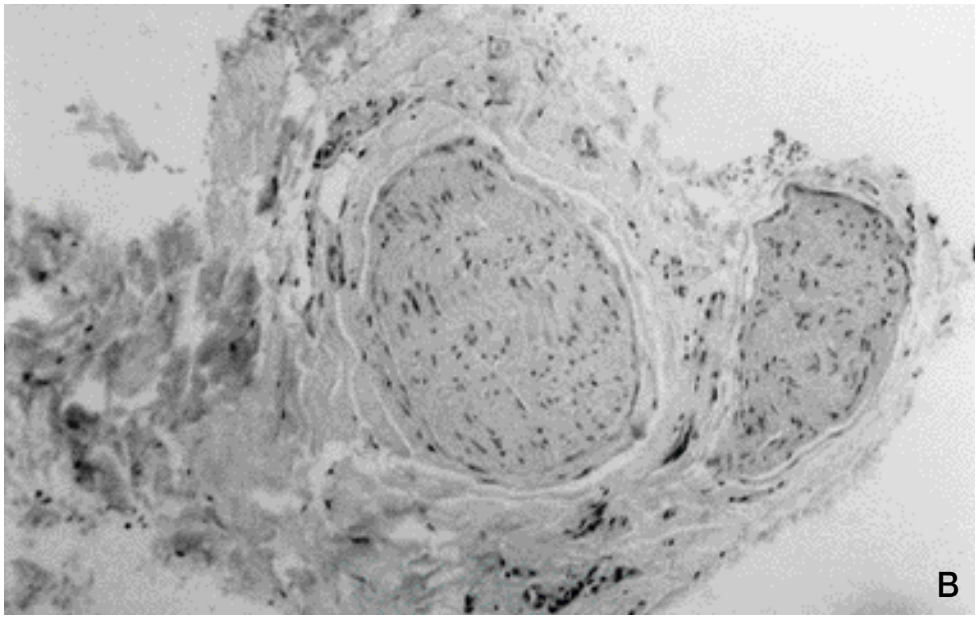


Figure 2. Fibrosis with encroachment on the sweat glands (A) and nerve fibers (B) (H&E, original magnifications ×100).

CONTINUED ON PAGE 346

CONTINUED FROM PAGE 344

The operation ablates the upper thoracic sympathetic nerve ganglions responsible for nerve stimulation of the sweat glands of the upper limbs. The most significant complication is Horner's syndrome, which results from injury to the stellate sympathetic ganglion.[7] In a summary of sympathectomies in 67 children and adolescents, complications included Horner's syndrome in 1 patient (1%) and varying degrees of compensatory sweating in 30 patients (45%).[8] Despite the evidence from animal studies that sympathectomy can result in facial atrophy, to our knowledge, there were no previous reports of such an association in humans.

The appearance of Parry-Romberg syndrome in our patient following thoracoscopic sympathectomy may have been coincidental or may have been causative. The patient had a slight facial asymmetry since early childhood, so it is possible that hemifacial atrophy actually began in the first decade of life. However, the mother noted rapid progression of the asymmetry after the operation. Thus, the operation may have influenced facial sympathetic innervations, thereby exacerbating or even initiating the hemifacial atrophy, as suggested by the animal studies. Whether there is a true association between surgery and Parry-Romberg syndrome remains to be studied further.

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