Parry-Romberg Syndrome: Vasculopathy and its Treatment With Botulinum Toxin

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Abstract: Parry-Romberg syndrome is a rare condition characterized by progressive, hemifacial atrophy, hair loss, enophthalmos, retinal vasculopathy occasionally associated with hemicranial pain syndrome (secondary trigeminal neuralgia). The cause of the condition is unknown; however, substantial evidence suggests that vasculopathy plays a significant role in the genesis of the neurologic damage and facial lipodystrophy. Herein describes a case of Parry-Romberg syndrome treated with repetitive botulinum toxin A injections, with almost complete resolution of severe chronic pain.

Parry-Romberg syndrome is characterized by dermal sclerosis and lipodystrophy of the face, orbit, and scalp. The syndrome has also been associated with loss of scalp hair associated with linear scalp furrowing, otherwise described as “saber-cut deformity.” This deformity has also been described as a local form of scleroderma (linear scleroderma). Comorbidity in central nervous system includes secondary form of trigeminal neuralgia and intractable migraine syndrome. Herein describes beneficial application of botulinum toxin for pain relief, which improved hair growth over injection regions and mentation. Informed consent was obtained for procedure according to the principles of the Declaration of Helsinki.

CASE REPORT

A 62-year-old nurse presented with severe debilitating pain of the right brow, forehead, and scalp for the past year associated with progressive hair loss over a linear region on the right portion of the scalp and atrophy over the right forehead and brow (Fig. 1A, B). The enophthalmos started at age 31. A biopsy showed focal inflammation in muscle and skin consistent with the diagnosis of linear scleroderma (“En coup de Sabre”). At age 53 (10 years prior to presentation), the pain which had been at first intermittent became constant and rated as a 9 of 10 in severity causing inability to maintain gainful employment. At age 53, the pain was rated as a 9/10 severity causing inability to maintain gainful employment. A local anesthetic block of the right temporalis, lateral pterygoid, and masseter muscles was performed to determine pain origin and to rule out any contribution from dental origins. A diagnosis of hemicranial pain syndrome was made.

FIG. 1. An axial T1-weighted image through the mid orbital level (A) shows severe enophthalmos, OD, secondary to atrophy of the right orbital fat, and atrophy of the periorbital and preauricular subcutaneous soft tissues. An axial T1-weighted image through the infratemporal fossa (B) shows atrophy of the right temporalis, lateral pterygoid, and masseter muscles. Enophthalmos with loss of hair in a linear pattern over the affected side are apparent (C, D).

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the sympathetic fibers’ effect on vasculature of the fingers. The regional effect creates a chemical sympathectomy. Laser Doppler studies have indicated an increased blood flow after the neurotoxin injections at therapeutic doses (Fig. 3). Botulinum toxin has been shown to be effective in relaxing smooth muscles in multiple organs and to have a salutary effect on inflammation and certain inflammatory mediators.

In summary, this case is the first well-documented example of Parry-Romberg neuropathy effectively treated with botulinum toxin over an extended period. Because of botulinum toxin-induced vasodilatation in hand vessels, it is postulated that increased perfusion may be the mechanism of pain relief. If increased tissue perfusion can be accomplished with repeated botulinum toxin, it is feasible that atrophic changes induced by vasculopathy can be mitigated by repeated botulinum injections for this condition.

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REFERENCES