Parry Romberg Syndrome: A Close Differential Diagnosis of Linear Scleroderma en coup de sabre

This 45 years old lady complained of asymmetry of her face since the age of 15 years. She also complained of decreased sweating on the left side and an intermittent left hemicranial headache. Examination revealed marked atrophy of the left side of the face involving the skin, subcutaneous tissue and muscles of the orbital, maxillary and mandibular regions. There was no induration or tightening or tethering of the overlying skin. The left half of the tongue was atrophic. Complement levels were normal and ANA was negative.

Facial hemiatrophy (Parry Romberg syndrome) is a slowly progressive disorder occurring mainly in females and usually involves the left half of the face. It begins in the second decade and may cause atrophy of all the tissues of the face, from skin to bones. The tongue and soft palate are often affected. There is no skin thickening or tethering. Treatment is cosmetic, consisting of transplantation of skin and subcutaneous fat.

Differentiating features between linear scleroderma en coup de sabre and Parry Romberg syndrome include paramedian atrophy in Romberg syndrome without induration of the skin overlying the scalp and with atrophy extending down the side of the face, often involving the tongue. In linear scleroderma, there is induration of the skin in the region of the scalp; it usually does not extend below the forehead and is usually not associated with neurological abnormalities. Prognostically, Parry Romberg syndrome is self-limiting but is associated with neurological abnormalities. A correct diagnosis of facial hemiatrophy will help avoid unnecessary therapy for linear scleroderma.

REFERENCES


A Wakhlu, V Agarwal, Amita Aggarwal, R Misra
Department of Immunology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India.
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