Parry-Romberg Syndrome

A six-year-old boy presented with skin lesions, initially started on the left cheek at 2 years of age, then spread to left lower eyelid and forehead. On examination, diffuse sclerosis on the left cheek extending to left lower eyelid was noted. Hypoplasia of left half of the face and deviation of mouth and lips to left side were noted, with loss of eyelashes in the left lower eyelid (***Fig.*** 1 and 2). Investigations show normal blood counts, renal function tests, and liver function tests. Rheumatoid factor and antinuclear antibody were negative. Skin biopsy shows features suggestive of early morphea. CT scan of brain was normal. CT scan of face shows hypoplastic left maxillary sinus, left hemi-mandible, atrophy of the left hemi-facial muscles and subcutaneous fat.

Parry-Romberg syndrome is an uncommon degenerative condition characterized by a slow and progressive atrophy of facial tissues. A sharp demarcated line between normal and abnormal skin called *coup de saber* develops. The diagnosis is clinical and based on characteristic cutaneous and soft tissue findings.

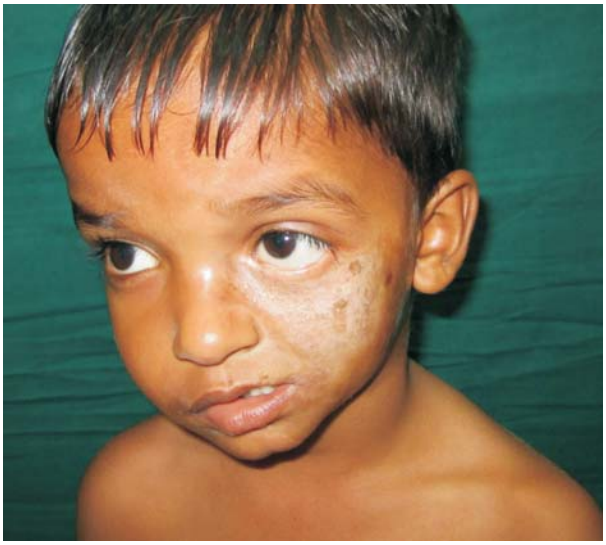

FIG.1 *Marked hypoplasia of the left half of the face with deviation of lips toward left side and loss of eyelashes in left lower eyelid.*

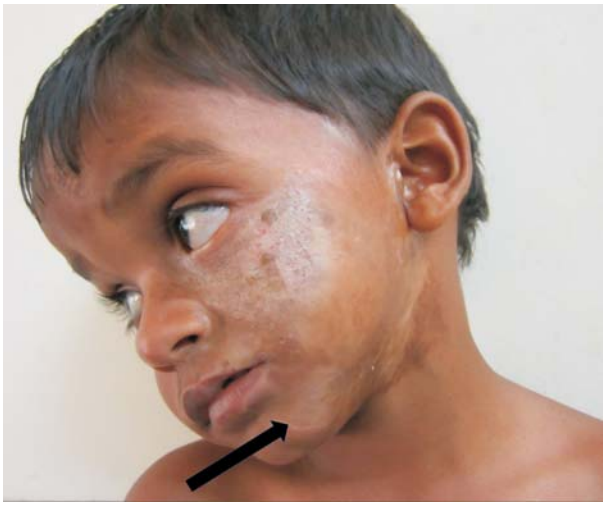

FIG. 2 *A big linear scar (coup de sabre) in the left side of mentum region.*

Differential diagnoses include hemifacial microsomia (first and second branchial arch syndrome) and its variants, such as Goldenhar syndrome, post-traumatic atrophy and partial lipodystrophy (Barraquer-Simon syndrome). Hemifacial microsomia and Goldenhar syndrome are congenital and non-progressive. In post-traumatic atrophy, history of trauma will be present. Lipodystrophy is usually bilateral and involves primarily the adipose tissue. Parry-Romberg Syndrome and localized scleroderma may represent differential spectra of same disease. Parry-Romberg syndrome will have hemifacial atrophy of the skin and tissue below the forehead, where as localized scleroderma is generally located in the fronto-parietal scalp and/or the paramedian forehead; it may extend down the face as well.

It is an auto-limitable condition and there is no cure. The treatment is usually based on reposition of adipose tissue.

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