Parry Romberg Syndrome in a Pediatric Patient

**Key Words:** Parry Romberg, facial hemiatrophy, hypoplasia maxilla

**Introduction:**
Parry Romberg Syndrome is also known as progressive hemifacial atrophy. It was first described by Parry in 1825 and Romberg in 1846. Eulenberg in 1871 termed it as Progressive facial hemiatrophy. The syndrome is characterized by slow atrophy involving one side of the face. The affected individuals are normal at birth, with insidious atrophy progression in the first two decades of life. We report a unique case of a 9-year-old girl with Parry Romberg syndrome with no neurological sequelae but with hypoplastic maxillary bones, malocclusion of teeth, atrophy of tongue, hepatosplenomegaly, and failure to thrive.

**Case Report:**
A 9-year-old girl presented with complaints of inability to gain weight, short stature, and deformity of the face. She was well up to 6 years of age followed by slowly shrinking of the right side of the face noticed at 6 years of age and then increasing progressively and now having hollowed appearance of the cheeks. The patient had deformity of the upper lips with deviation of the lip to the right side. She was well growing up to 6 years but not gaining weight and height for the last 3 years. Parents complained of short stature as compared to other peers in school and increasing hollowing of the right side of the face. No history of trauma, prolonged infection, or contact with tuberculosis. No other family sibling was suffering from similar illness. Clinical examination revealed a malnourished girl with weight 20 kg [<3rd centile], height 120 cm < 2 S.D. Vitals signs were normal. Liver 3 cm, BRSCM, spleen was palpable. She was alert, with no signs of neurological abnormality. Speech and hearing were normal. Millstones were normal for age except malocclusion of teeth noticed at 8 years age. The face was asymmetric with right-sided atrophy. The right maxilla and zygomatic region appeared hypoplastic with shrinking of skin and giving a hollow appearance of cheek on the right side (Figure 1) as compared to the normal left side (Figure 2). The right side of the upper lip showed deviation towards the side of contracture. Chin was also deviated to the right side. Ears and eyes were normal. Intra-oral examination revealed mixed dentition stage with deformity of upper teeth and atrophy of tongue. Child was investigated. Blood counts, antibody testing for autoimmune possibility, radiography, and ultrasonography were done. Antibody testing for antinuclear antibodies ANA was negative. IGA Tissue Transglutaminase antibody was normal. Thyroid profile was normal.

Ultrasound abdomen showed liver enlarged span 11 cm normal texture. Spleen was enlarged span 8.2 cm. X-ray of skull including orbit, sinuses, and facial bones was done, which revealed hypoplastic maxillary bones, small maxillary sinuses, malocclusion of teeth. Orbits were normal, mastoid bones normal (Figure 3). Based on clinical and radiological findings the diagnosis of progressive facial hemiatrophy hence Parry Romberg syndrome was made. Plastic surgeon opinion was taken, and she was planned for cosmetic surgery once the disease stabilizes.

**Discussion:**
Parry Romberg syndrome is also known as progressive hemifacial atrophy. It was first described by Parry in 1825 and Romberg in 1846. It is characterized by slow and progressive...
atrophy of skin and subcutaneous tissue, but may extend to muscles and bones of face [4],[5]. The disease is usually involves left side of face but may extend to one side of neck and whole body [5]. The disease has a prevalence of 1 in 70000 affecting mainly females [5] but in our case right side was involved. The disease has onset in first decade of life, progresses and then stabilizes [6],[7]. The patient in our case has disease onset at 6 years of life and approached to our hospital at 9 years of life. The exact etiology is unknown and various factors have been proposed including autoimmunity, viral infection, peripheral neuropathy, vascular disturbances, increased sympathetic activity causing fascial atrophy [8]. The disease can present with ocular, neurological feature like trigeminal neuralgia, epilepsy, focal paresthesia, headache [9]. Imaging findings on CT includes intracranial calcifications, vascular malformations, demyelinations [10]. Treatment aims at correction of fascial deformity after cessation and stabilization of disease process, usually at end of second decade. Plastic surgeon opinion was taken in our case and advised facial augmentation with grafting by 18 years of age.

REFERENCES: