PROGRESSIVE FACIAL HEMIATROPHY: PARRY-ROMBERG SYNDROME – A CASE REPORT

Chandra Shaleen*, Raj Vineet**

Progressive facial hemiatrophy or Parry-Romberg syndrome is a rare disease characterized by atrophy of subcutaneous tissues including hard tissues of one side of face. It is a rare condition which generally develops before the age of 20 years. In this article a case of this disease is presented and treatment options are discussed.

Keywords: Parry-Romberg syndrome, facial hemiatrophy, Progressive facial hemiatrophy.

The patient, a 22 year old right handed girl, reported to the Out Patient department of Saraswati Dental College, Lucknow, with chief complaint of deformation of her left side of face. Deformation of face was first noted by her parents when she was 8 years old, which increased with age. Before this the patient had not consulted any physician. The patient had two siblings, one younger brother aged 18 years and a younger sister aged 15 years. Both were healthy and symptom free. Her parents were unrelated and healthy with no evidence of tissue atrophy. There was no family history with similar complaints or with collagen tissue disease. The patient had been delivered normally at full term following a normal pregnancy. She was a pleasant, intelligent, 152 cm. tall girl and weighing 48 kg. General physical examination did not reveal any abnormality. There was a remarkable difference in the length of body of mandible on left and right side with shortening seen on left side (Fig. 1). The left oral commissure was more superiorly placed as compared to the right and the pinna and ear canal formation on left side was incomplete (Fig. 2). The eyes, eye lashes, eyebrows and hair were unaffected. On palpation, the skin was soft and there was no sclerosis present. The patient had no history of convulsions. No subjective complains such as pain or paraesthesia or anaesthesia was present. There was no history of major trauma. There were no abnormalities in ophthalmologic, neurological and systemic examinations. Routine blood counts and biochemical tests including thyroid function tests were normal. Based on the above features, a diagnosis of progressive facial hemiatrophy was made. Patient was advised cosmetic plastic surgery.

**Department of Oral Pathology, Saraswati Dental College,Faizabad Road,Lucknow- 227105*

Discussion:

Progressive facial hemiatrophy or Parry-Romberg syndrome was first described by Parry in 1825 and later by Romberg in 1846[1-3]. The etiology of this rare syndrome is unclear. Etiological factors implicated with this disease are hereditary (suggested to be autosomal dominant) viral infections, peripheral trigeminal neuritis, lymphocytic neurovasculitis, localized scleroderma and endocrine autoimmune disorders[3,4].It has been emphasized in literature that progressive facial hemiatrophy accompanied by segmental vitiligo may be due to autoimmunity[5].

The wasting in progressive facial hemiatrophy involves subcutaneous tissue, fat, cartilage and bone. The muscles may appear to be affected but this is due to atrophy of fat and connective tissue in the muscle; the muscle fibres themselves remain intact. Many related structures such as the tongue, soft palate, mucous membranes, cartilage and some of the facial bones may be affected. There may be pains of neuralgic type in the affected area. Sweating and lacrimation on the affected side may be increased or diminished.

In some patients, the facial hemiatrophy has been part of a generalized hemiatrophy involving limbs on the same side[6]. Cases of 'crossed hemiatrophy' are also reported[6] in which the wasting involved one side



Fig. 1: Frontal view, showing difference in length of body of mandible



Fig. 2: Lateral view, showing lack of development of Pinna and ear canal

of face and limbs of the opposite side. Facial hemiatrophy with associated atrophy of the ipsilateral breast has also been reported[7].

Due to the striking appearance, the diagnosis of this disease is easy, however scleroderma and lipodystrophy have to be considered in differential diagnosis.

The atrophic process usually subsides with attainment of adulthood. Though there is no specific treatment, cosmetic surgery is helpful once the atrophic process has subsided. Excellent results have been obtained by implanting fat from lower abdominal wall or from the omentum into the subcutaneous tissues of the cheek[8]. Recently artificial fillers have made great advances. Poly-L-Lactic acid derived fillers[9] like Sculptra© and other fillers like Restylane© have great potential to be used for treatment of such patients.

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