A Case Report - Progressive Hemifacial Atrophy

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Abstract: Progressive hemifacial atrophy also known as Parry Romberg Syndrome is rare degenerative and poorly understood condition of unknown etiology. Factors involved in pathogenesis are autoimmunity, trauma, radiation, viral infection, hereditary and endocrine disturbance. Atrophic process in disease slowly progresses and then become stable after several years.

Key Words: Hemifacial atrophy, scleroderma, alopecia.

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I. Introduction

Progressive facial hemiatrophy also known as Parry Romberg Syndrome characterised by progressive but self limiting atrophy of the skin and subcutaneous tissue of face, muscle, cartilage and bone. Onset is insidious usually manifest in 1st or 2nd decade of life with skin changes resembling scleroderma. Incidence is more in female (F:M, 3:1). Usually confined to one side of face mostly to left side. It may occasionally spread ipsilaterally to neck, one side of body and scalp and may lead to cicatricial alopecia of scalp. Usually accompanied by neurological complain like trigeminal neuralgia, facial paraesthesia. Disease is thought to be unilateral inflammatory process associated with chronic vascular disturbance or neurological cause. Many evidence support autoimmune mediated inflammation by activated T Cells, plasma cell and eosinophil leading to elevated ANA, cytokine and adhesion molecule (ICAM-1, VCAM-1, E-selectin) leading to tissue and vascular damage. Extracutaneous involvement includes scalp, ocular, oral, articular, muscular and neuromuscular. Disfigurement, fatigue, pain, itch are the main complain which also affect psychosocial behaviour and quality of life.

II. Case History

- A 36 years old married female came to dermatology opd with complain of atrophy with hyperpigmentation of left side of face for 10-15 years, alopecia of the scalp and burning pain over facial and scalp lesion for 3-4 years. On history she had non sclerotic hyperpigmented patch of size 2cm-2cm just below left pinna. It was not associated with pain, burning or pruritus, but the patient had resistance over opening of jaw and difficulty during mastication. She used some depigmenting cream for that but the lesion was non responding to treatment. Gradually the hyperpigmented patch started enlarging in size and in around 15 years involved almost whole face on same side, with loss of subcutaneous fat, leading to facial asymmetry. The difficulty during mastication and spasmodic feeling during jaw opening gradually improved and not present since last 5-6 years. Since last 3 years she noticed gradual patchy loss of hair over the tempororfrontal area of left side of scalp which is increasing in size. She is also complaining of severe burning pain over the involved skin on same side of face and scalp since last 3 years and migraine attack occasionally. On examination the involved skin was hyperpigmented non sclerosed, angle of mouth was slightly pulled towards left side. Oral cavity was examined and it was normal with normal dentition. Lower eyelid was slightly everted. On closing the eye on left side a small gap between both eyelids seen just lateral to medial canthus due to localized atrophy of upper eyelid. Loss of lower eyelashes along with alopecia in left eyebrow (towards medial side) was also noticed. Cicatricial alopecia of scalp in same side was present. No history of seizure, and any other chronic illness present. No history of similar problem in any family members and the patient was in good general health condition. All routine investigation were normal. ANA was slightly positive. Anti SCL-70 antibody was negative.

- X-ray of the face was done to rule out any bony involvement but no significant bony changes was there except some hyper lucent area over left maxillary sinus.
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Figure 1 - Atrophy of left side of face with alopecia over left eyebrow.

Figure 2

Figure 3
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III. Discussion

The clinical feature of progressive hemifacial atrophy was first described by Caleb Hillar Parry in 1825 and subsequently described by Moritz Heinrich Romberg in 1846. Although exact etiology is unknown it is believed to be related to localized facial scleroderma. Pathology is related to autoimmune chronic lymphocytic infiltration around neurovascular bundle. Anatomic changes in Parry Romberg Syndrome impact the growth potential of soft tissue and bone preventing proper growth in size during active growth period. Hence the atrophy that started in the second decade of life is less noticeable as facial growth is nearly complete by this time (as in this case). Patients mostly visit hospital due to problem of bony contracture hindering jaw opening, eating, mastication and psychosocial problem due to facial asymmetry.

References


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