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Parry-Romberg Syndrome: A Case Report

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Abstract: Progressive hemifacialatrophy ,also known as Parry-Romberg syndrome, is an uncommon, degenerative and poorly understood condition. It is characterized by a slow and progressive atrophy affecting one side of the face ,generally left . An autoimmune mechanism is suspected and the syndrome may be a variant of localized scleroderma .A cerebral disturbance of fat metabolism has been proposed as a primary cause . This can be result of a atrophic malformation of cerebral sympathetic nervous system. Possible factors that are involved in the pathogenesis are trauma, viral infections ,heredity, endocrine disturbances ,and autoimmunity ,among others. Characteristically, atrophy progresses slowly for several years and, soon after, it become stable. The purpose of this work is to report a clinical case with right sided progressive hemi facial atrophy and affecting a middle age group male which itself is a rare presentation of this uncommon condition.
Keywords: Hemifacialatrophy, Parry- Romberg Syndrome, scleroderma

INTRODUCTION

Progressive hemifacial atrophy, also known as Parry-Romberg syndrome, is a rare uncommon degenerative condition characterised by a slow and progressive atrophy, generally unilateral, of facial tissues, including muscles ,bones ,and skin[1]. It was first described by Caleb Hillier Parry in 1825 and later in more detail by Moritz Heinrich Romberg in 1846 [2]. More than an aesthetic trouble, this illness brings several functional and psychological problems, when a "symmetric" face loses its identity.

Characteristically, there is regional atrophy of skin, subcutaneous tissue, and musculature. When the onset is before the second decade of life, the underlying bone and cartilage may also be involved [3]. A sharply demarcated line between normal and abnormal skin develops ,so called coup de saber,and the involved area varies from a discrete lesion to a widespread, extensive malformation .Alopecia and pigmentation of the involved skin often appear[3].

In addition to the obvious facial atrophy , a variety of accompanying conditions have been reported ; ocular changes in 10% to 35% of cases; neurologic disturbances, which include focal epileptiform seizures and trigeminal neuralgia; and ipsilateral progressive body atrophy of half of the lip and tongue, shortening of the body of mandible and/ or ramus and retarded tooth eruption [3].

The objective of this work is to report a clinical case of a middle aged male patient with features of progressive hemifacial atrophy of right side along with discussion concerning general characteristics, aetiology , pathophysiology ,and treatment of progressive hemifacial atrophy.

CASE REPORT

A 49 –year-old ex army man was referred to medicine OPD of our tertiary care Hospital for general physical check-up from Department of Dermatology. He had presented to the Dermatologist for complaints of hyper pigmentation of right half of face. During the physical examination it was noted that this patient presented with a facial asymmetry with marked hypoplasia of right side of the face(**Fig-1**) in maxillary region and atrophy of right temporal region .A big linear depression (coup de sabre) was present on right side of face producing an obvious demarcation of bony prominence. Patches of hyper pigmentation were seen on the skin of affected area i.e. on the malar region, preauricular region and supraorbital region. No history of preceding trauma, itching, photosensitivity, stiffening over affected site was present. On proper history taking patient revealed that he had first noticed this asymmetry of the face(**Fig-2**) 4 to 5 years back and it gradually progressed since then. At present it had become an obvious deformity producing an aesthetic concern to the patient for which he had previously consulted many local doctors.

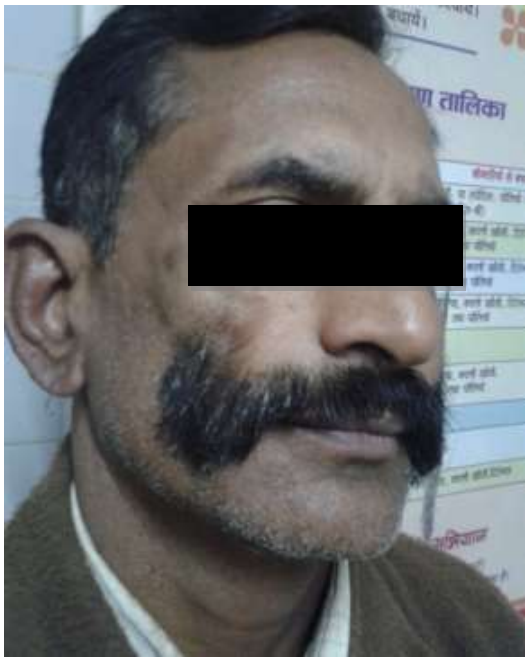

Fig. 1 – Hypoplasia and atrophy in right side of face demarcating bony prominences.

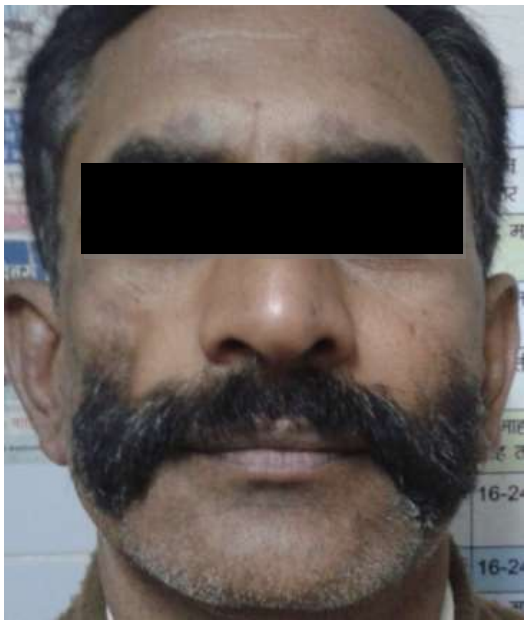

Fig. 2 - Obvious asymmetry between right and left side of face.

DISCUSSION

Progressive hemifacialatrophy,as described in the case above, is a rare progressive degenerative pathology, of unknown cause , which affects the face and produces not only aesthetic concern to the patient but also the functionality of the attained hemiface.

The pathogenesis of progressive hemifacial atrophy is unknown. A supposed neurotrophic pathogenesis was described by Cassirer in 1912 [4]. He proposed that the atrophic disease process follows the pattern of trigeminal nerve innervations[4] . Certain studies suggested that the disorder was familial one. The anatomic changes of Parry- Romberg Syndrome impact the growth potential of hard tissue, preventing an increase in size during active growth periods . The associated soft tissues shrink by loss of adipose tissue [4]. Hence, atrophy that started in the second decade of life is less noticeable because facial growth is nearly complete. Early disease onset and long duration cause greater deformity.

Most of the cases reported in literature affect the younger age group i.e. between 5-15 years and involve left side of face [5] but in our case disease process started in late forties and involves right side of face. The possible explanation for this may be that as autoimmunity being the key factor along with the genetic and environment factor superimposing on it at any time of life

The severity and specific symptoms of the disease are highly variable from one person to another. Frequently, the onset of Parry Romberg Syndrome occurs along first and second decades of life . This syndrome seems to have higher incidence among women and affect left side of face most often. Characteristically, the atrophy progresses slowly over many years and, then ,it become stable [1]. Alternatively, the condition may “burn” itself out at a very early stage and result in minimal deformity [3]. Alterations concerning involvement ,duration, and deformity can stabilize in any stage of growing and development[1]. The extension of the atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare.

Clinically , the skin can be dry and with a dark pigmentation. Some patients present a demarcation line between normal and abnormal skin, reminding a big linear scar , known as” coup de sabre”, as could be noticed in this patient [1, 6]. Occasionally, there may be some neurological complications, such as trigeminal neuralgia, facial paresthesia, severe headache, and contra lateral epilepsy [1,6] .These complications were not discerned in the present case.

The relationship between Parry-Romberg Syndrome and scleroderma is controversial. Regarding the clinical findings and clinical course, localized scleroderma and Parry –Romberg Syndrome may represent different spectra of the same disease process[7,8]. It has been suggested that the term Parry-Romberg syndrome should be used for progressive hemifacial atrophy without features of cutaneous scleroderma [7].

Besides aesthetic treatment ,symptomatic treatment of neurological disorder is indicated and

corrective surgeries should be done when the disease process ceases to progress [9]. Parry -Romberg Syndrome is an auto-limitable condition and there is no cure . Affected patient should have multi-disciplinary attendance of physicians, dentists, phonoaudiologists, and psychologists. The treatment is usually based on reposition of adipose tissue that was lost due to atrophy [10].

CONCLUSION

In conclusion ,a case of progressive hemifacial atrophy, with its archetypal feature ,was discussed. Till recently ,the exact etiology and pathogenesis of this degenerative condition has not been elucidated. Many patients present with classic clinical features and there is little intricacy in diagnosis of progressive hemifacial atrophy .Parry Romberg Syndrome, itself is a rare condition and what makes it more rarer is that it is reported on right side and in a late middle age male. A proper diagnosis and multidisciplinary treatment approach is essential for management of progressive hemifacial atrophy.

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