Abstract:
Parry Romberg syndrome (also known as progressive Hemifacial atrophy) is a degenerative condition characterized by atrophic changes affecting one side of the face. An Autoimmune mechanism is suspected, may be a variant of localized scleroderma, but the precise etiology and the pathogenesis of this acquired disorder remains unknown. Here, we report a case of 19 year old female with progressive hemifacial atrophy involving the left side of the face. There is no definitive treatment for this condition, but an attempt to use restorative plastic surgery which includes fat or silicone implants, flap or pedicle grafts or bone implants can be done to improve facial disfigurement.

Keyword: Progressive Hemifacial Atrophy, Parry Romberg syndrome

INTRODUCTION:
Parry Romberg syndrome also known as Progressive Hemifacial Atrophy is characterized by a progressive but self-limited atrophy of the skin and subcutaneous tissue on one side of the face. There is a progressive wasting of subcutaneous fat, sometimes accompanied by the atrophy of skin, cartilage, bone and muscle. It overlaps with a condition known as Linear Scleroderma “en coup de sabre”. In most of the instances atrophy is usually confined to one side of the face and cranium; however it may occasionally spread to the neck and one side of the body. The onset is insidious, and the condition usually manifests in the first or second decade of life. Progression of the disease is rapid in the two to ten years following onset and then stabilizes. The etiology of this condition is unclear. Many theories have been postulated to explain this rare disease like autoimmunity, alterations in the peripheral sympathetic nervous system (perhaps as a result of trauma or infection involving the cervical plexus and/or the sympathetic trunk), disorders in migration of cranial neural crest cells or chronic cell-mediated inflammatory process of the blood vessels. Hemifacial atrophy occurs sporadically and some familial distribution has been found.
It is common in females than males with a ratio of 1.5 : 1.

**HISTORY AND REVIEW OF LITERATURE**
The disease was described in 1826 by Caleb Hillier Parry(1755-1822), in a collection of his medical writings which were published posthumously by his son Charles Henry Parry(1779 – 1860). It was described a second time in 1846 by Moritz Heinrich Romberg(1795 – 1873) and Eduard Heinrich Henoch(1820 -1910). German neurologist Albert Eulenberg (1840 – 1917) was the first to use the descriptive title “progressive hemifacial atrophy” in 1871. Fat grafts have been described for use in contour reconstruction of the face since van der Muelen's account in 1889. Multiple reports over the past century- Neuber 1893, Lexer 1910, Peer 1956, Ellenbogen 1986, Billings and May 1989, have documented excellent aesthetic results. Fat auto grafts are said to vascularise at day 4 and after a process of inflammation and fibrosis, resorption occurs until roughly 40 to 70% of the graft remains in patients followed from 1 to 3 years. Dermis fat grafts have also been employed with excellent contour restoration and survival. Fat and dermis fat grafts would appear to be appropriate for mild to moderate contour deformities of the hemiface in Romberg's disease. Facial contour restoration with free transfer was described by Harashina in 1977. Following his description of free groin transfer others have used omentum, parascapular,rectus abdominis,lattismus dorsi flaps. Unlike fat grafting the free vascularised flaps transferred provided large volumes of reliable tissue.

**CASE REPORT:**
19 year old female reported with the complaints of atrophy of the left half of the face. Patient was apparently normal at birth. Later, at eight years her mother noticed atrophy of the face which had progressed upto sixteen years. The past three years showed no progression of symptoms. There were no associated symptoms.

The patient’s medical and family history was non contributory.

**Fig. 1 HEMIFACIAL ATROPHY OF LEFT SIDE OF FACE**

On physical examination facial asymmetry was detected due to the hollowing of cheek and chin on the left side, the left eye was depressed in the socket(enophthalmos) along with deficient eyebrows, the malar hypoplasia, atrophy of facial skin and the upper lip on the left side. Loss of subcutaneous fat with prominent bony ridges on the ipsilateral side was observed when compared to the normal side. Pigmentation was noticed on the left side of the forehead and cheek. There was atrophy of temporalis,buccinators, masseter muscles and prominent zygomatic arch on the left side. Maxilla and mandible appeared hypoplastic on the affected side(Fig 1). Routine blood investigations were carried-out which revealed all values were within normal limits. Based on the clinical features, a diagnosis of Parry-Romberg syndrome was made. Patient opted for soft tissue reconstruction of the affected side. Dermal fat graft was planned.
An elliptical incision was made in the left side buttock (Fig 2), overlying skin was deepithelialised (Fig 3) and dermal fat graft was harvested (Figs 4, 5).

Through a left preauricular incision, pocket was created (Fig 6) and dermal fat graft inserted and secured with sutures (Fig 7). Skin was closed with suction drain (Fig 8). Post operative period was uneventful (Fig 9). Sutures were removed on the tenth post-operative day. Patient was followed up for one year. During follow up, she had forty percent of fat resorption (Fig 10).

**Fig. 2 SKIN INCISION OVER LEFT BUTTOCK**

**Fig. 3 DE-EPITHELIALISATION DONE**

**Fig. 4 DERMAL FAT GRAFT HARVEST**

**Fig. 5 DERMAL FAT GRAFT**

**Fig. 6 PREAURICULAR POCKET CREATION**
Progressive hemifacial atrophy is not a congenital disorder, with the typical onset being in the first or second decade of life. The hallmark of the disorder is a slowly progressive course with an "active phase" of disease characterised by involution or "wasting away" of the skin, subcutaneous tissue and muscle. This "active phase" lasts from two to ten years. The subcutaneous tissue is the most severely involved, followed by substantial involvement of the skin and muscle. Early onset cases have hypoplasia of mandible and maxilla. Important features of this disease are enophthalmos, deviation of mouth and nose to the affected side and the unilateral exposition of teeth (when lips are involved). Clinically the skin can be dry and hyperpigmented. Some patients present a demarcation line between normal and abnormal skin, known as "coup de sabre" (french term which means "cut of the sword"). Neurological complication such as trigeminal neuralgia, facial paraesthesia, severe headache and contralateral epilepsy can also be present. There seems to be no evidence that systemic treatment for the true Romberg’s disease pattern of illness is of benefit. The treatment of true Romberg is symptomatic. The timing of any surgical intervention is generally agreed to be the best following...
exhaustion of the disease course and completion of facial growth. Photographic documentation and confirmation that the disease is actually burnt-out is actually important and most authors concur on a waiting period of one or two years before proceeding with reconstruction. Facial contour restoration have been described including fat grafts, dermis fat grafts, Lipo injections, pedicle flaps, bone and cartilage grafts, microvascular free tissue transfer, orthognathic corrective maxillary surgery and alloplastic implants probably all of these have a place if properly utilized in the appropriate individual case.

CONCLUSION:
Parry Romberg syndrome is an uncommon condition which manifests as atrophy of one side of the face. There is no definitive treatment for this condition but an attempt to use restorative plastic surgery includes fat or silicone implants, flap/pedicle grafts or bone implants can be done to improve facial disfigurement.

REFERENCES:


