



PROGRESSIVE FACIAL HEMIATROPHY

Parry-Romberg Syndrome A Case Report



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Summary

Progressive facial hemiatrophy or Parry-Romberg Syndrome is a rare disease characterized by atrophy in subcutaneous tissues on skin on one half of the face and some accompanied systematic findings. Generally, it develops before the age of 20. It is quite difficult to discriminate it from linear scleroderma localized in fronto-parietal area as they have similar clinical characteristics. In this article, 12-year-old case with atrophy on the left half of her face is presented. (Figure 1)

Case

The patient was a 12 year-old girl. She came to the Department of Dermatology with the complaint of hyperpigmentation and deformation on the left half of her face. The first brown hyperpigmentation developed on her chin 5 years ago then, depression on the left side of her face and tongue have been observed in time. (Figure 2) Gradually, a yellow-brown hyperpigmentation developed on the left of her nose, on her forehead and temples. Deformation on her chin started 2 years ago. Hyperpigmentation and deformation were always confined to the left half of the face. The patient had not admitted to any other physician before because of these complaints. There were depressed atrophic areas with the hyperpigmentation on the chin and on the edge of the mouth. Lesions had caused asymmetric appearance on the face. On palpation, skin was soft and there was no sclerosis. The patient had neither a history of convulsion nor subjective complaints such as pain or hypoesthesia. There was no history of major trauma. There was nobody in the family who has similar complaints or collagen tissue disease. There were no abnormality in ophthalmologic, neurological, systemic examinations.

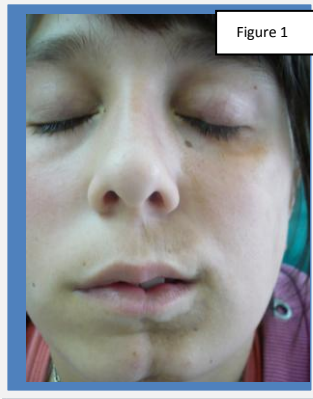


Figure 1

Routine blood count and biochemical tests were normal. The patient was negative for antinuclear antibody, anti DNA and Borrelia IgM. Thyroid function tests were normal. There was no anomaly in the soft tissues or bones in the direct head graphy. In the histopathological examination of the punch biopsy taken from the chin area, there was hyperkeratotic, irregular akantotic squamous epithelium; vacuolization in cells in epidermis; edema and dispersed, rare, mononuclear inflammatory cells in dermis. No dermal fibrosis in cross-sections and no atrophy in skin parts were observed.

Discussion

Progressive facial hemiatrophy (PFH) or Parry-Romberg Syndrome (PRS) was originally described by Parry in 1825 and by Romberg in 1846 (1-3) (Figure 3) The etiology of this rare syndrome still remains unclear. Etiologic factors accused for the disease are familial tendency which was suggested to be autosomally dominant but not much supported in literature, viral infections, peripheral trigeminal neuritis, lenfositic neurovasculitis, localized scleroderma and endocrine autoimmune disorders (3, 4). In literature it is emphasized that PFH accompanied by segmental vitiligo may be due to autoimmunity (5).

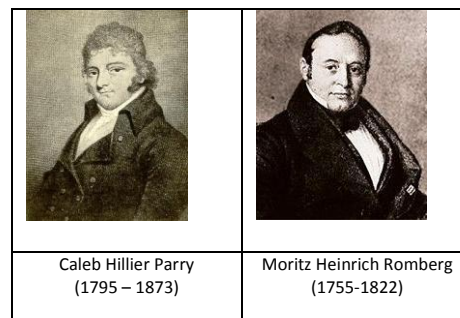
Linear scleroderma is a form of localized scleroderma proceeding with an only unilateral lesion. It emerges in early ages (1st and 2nd decade.) (2, 3). In its etiology, autoimmunity, genetic factors, viral and bacterial infections (such as Borrelia burgdorferia) are the most associated factors. The most frequently, the lower extremities are involved, which is followed by upper extremity, frontal area and frontal chest. Frontal or fronto-parietal linear scleroderma, also called as "en coup de sabre" (LSCS), develops enduring skin lesions with band-like form in this lesions (2, 3). Depression in the affected areas appeared in the form of groove and causes facial hemiatrophy. It causes scartial alopecia on hairy skin. It generally emerges unilaterally and do not extend to the lower part of the eyebrow. Sometimes, atrophy may emerge on ipsilateral tongue. Active period lasts in 2 or 5 years. In an early period, perivascular mononuclear cell infiltration, later thickness in the collagen bands in dermis are observed histopathologically. Inflammatory and fibrotic structure may also involve subcutaneous tissue, muscle, periosteum and bone. It may cause serious cosmetic problems and may be accompanied by neurological and ophthalmological anomalies (2, 7).



Figure 2

There are a lot of clinical, histopathological, and serological similarities between LSCS and PRS, and absolute diagnosis is not often possible. Some authors support for differentiating these diseases, however some of them think it is not true to identify these diseases separately. Currently, most authors accept the existence of overlapping diseases with clinical and laboratorial findings of PRS and LSCS. If PRS is evaluated as for its clinical properties, a classical PRS affects one half of the face, sclerosis is not seen on the skin, atrophy is mainly in the subcutaneous tissue. LSCS also affect one half of the face; it is usually observed on the area beginning from the line of eyebrows extending to the hairy skin; the skin is depressed and adherent, and also there may be scartial alopecia on the hairy skin (2,3,6,7).

Figure 3:



Caleb Hillier Parry
(1795 – 1873)

Moritz Heinrich Romberg
(1755-1822)

The histopathological findings of our case are not specific for both diseases. Clinical findings of our case led us to think that it was the Parry-Romberg syndrome rather than en coup de sabre type of linear scleroderma since there was a common involvement on the left side of the face from the frontal zone to the mandibular zone, there was not a scartial alopecia and sclerosis on the lesions.

In the treatment of PRS; immunosuppressive medicines such as cyclophosphamide, prednisolone, methotrexate are used, as LSCS treatment (4). In cases, where an advanced atrophy exists, cosmetic plastic surgery is applied. For this reason, mostly grafts together with dermis and fatty tissue are preferred (1). We are still going on following the treatment of our case, which we applied 5 mg/day of prednisolone.

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