Progressive hemi facial Atrophy- A Review

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Abstract:

Parry Romberg Syndrome (PRS) also called progressive hemifacial atrophy characterized by atrophy on one side of face typically fat, but variably skin, other connective tissue, and sometimes bone. Several clinical features have been reported like facial hemiatrophy, abnormalities of ear, eye, neurological abnormalities and oral abnormalities. This review describes the etiopathogenesis, clinical features, differential diagnosis and management of this condition.

Keywords: Parry Romberg syndrome (PRS)**,** Progressive facial hemi atrophy, en coup de sabre

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Introduction

Unilateral progressive atrophy of the face was first described by Parry in 1825 and by Romberg in 1846. Eulenberg coined the term 'progressive facial hemi atrophy' in 1871. There is involvement of the skin and subcutaneous fat, and on rare occasions also of the muscles and bones. It is uncommon and generally unilateral with a higher incidence rate in females. The extension of the atrophy is frequently limited to one side of the face, and the ipsilateral involvement of body is rare.[1] More than an aesthetic trouble, this illness bring several functional and psychological problems, like ocular problem, migraine, epilepsy.[2]

Etiopathogenesis

The aetiology of this rare condition is unknown. Various authors have linked this condition to local trauma, infection on to lesions of the ipsilateral trigeminal nerve or cervical sympathetic ganglion, but the pathology remains obscure.[3]

The skin pathology if caught at the onset shows inflammatory changes. Supportive evidence for an inflammatory hypothesis includes: a higher frequency of autoantibodies than the general population; the overlap with linear scleroderma; vitiligo; the presence of transient high signal lesions on brain MRI; a couple of neuropathological reports of intracerebral inflammation; and oligoclonal bands in the cerebrospinal fluid. Cats and rabbits with experimental lesions of the superior cervical sympathetic ganglion share some of the clinical features of Parry-Romberg syndrome. There are very rare cases (<2%) with a possible hereditary aetiology.[4]

T-cell epitope of B. burgdorferi outer surface protein A (OspA) has significant homology with a fragment of human LFA-1α and hypothesized that this could lead to an appropriately prolonged production of inflammatory mediators even after complete elimination of the spirochete. Expression of LFA-1α and ICAM-1 is increased in the course of morphea, strengthening the idea that cross-reactivity between OspA and LFA-1α could play a role in perpetuating the chronic inflammatory process, contributing to the pathogenesis of fibrosclerotic lesions.[5]

Clinical features

The main feature is hemiatrophy of the facial tissues, typically fat, but variably skin, other connective tissue, and sometimes bone.[6]

Patients range in severity from those with barely perceptible asymmetry to severe disfigurement. Patients have a more definite vertical or diagonal "line" on their forehead as a result of cutaneous sclerosis. These lines tend to follow "Blaschko's lines". When a pathological "line" is present it is called scleroderma "en coup de sabre" ("in a sabre cut"). Scleroderma "en coup de sabre" does, however, appear to be an overlapping condition with Parry-Romberg syndrome and shares a similar list of associated features. The other terms to be familiar with in this area are "morphoea" and Gower's panatrophy.[4]

The ear may become misshapen and smaller than normal or, because of lack of supporting tissues. Basilar kyphosis has been described. Early facial change, usually appearing during the first decade (average-9 years), involves the paramedian area of the face and slowly spreads, resulting in atrophy of underlying muscle, bone, and cartilage. First to be involved is usually the area

covered by the temporal or buccinator muscles. The process extends to involve the brow, angle of the mouth, neck, or even half the body. There is a marked predilection for left-sided involvement of the face. The overlying skin often becomes darkly pigmented.[7]

The osseous lesions described in Parry Romberg Syndrome (PRS) are variable and are strictly related to the age at which the condition appears. When onset in the children younger than 5 years of age, it is frequent to find the frontoorbitozygomatic area affected, whereas when onset is late, in occurring, the skeletal changes take place preferentially in the lower third of the face. The most frequently reported osseous lesions consist of maxillary and mandibulary hypoplasia, in all dimensions, with deviation of the mid-line of the face towards the affected side. The body of the mandible may be shorter than the normal. Spontaneous fractures of the mandible have been reported.[8]

Some individuals may experience other associated symptoms like neurological abnormalities such as epilepsy, migraine and facial pain, trigeminal neuralgias, facial paresthesias, ophthalmic disorders such as heterochromia, uveitis, enophthalmos due to atrophy of fat around the eye[9] and lagophthalmus.[7]

Changes in the hair may precede those of the skin. The scalp on the affected side may exhibit circumscribed but complete alopecia limited to the paramedian area, eyelashes, and median portion of the eyebrows. Poliosis, or blanching of the hair, has also been noted.[7]

Mouth and nose are deviated to the affected side, deviating also facial and dental midlines. Atrophy of superior lip led the anterior teeth to be exposed, and there may be also unilateral atrophy of tongue. Our case showed clearly those features of facial asymmetry and tongue atrophy.[10] A decreased depth and width of the retromolar region of the pharynx may occur.[9] Rarely there may be associated involuntary jaw closure.[9] Radiographically, teeth of affected side can present some deficiency in root development and, consequently, delayed eruption and dental crowding. Very often, there is unilateral posterior crossbite, as a result of jaw hypoplasia and delayed teeth eruption. The intraoral soft tissue and chewing muscles are, sometimes, normal,

without any movement, speeching or deglutition implications.[10]

Differential diagnoses

Hemifacial microsomia (first and second branchial arch syndrome) and its variants, such as Goldenhar syndrome, but these are congenital and essentially non-progressive conditions. Post-traumatic atrophy and partial lipodystrophy (Barraquer-Simon Syndrome) are also included in the differential diagnosis. However, partial lipodystrophy is usually bilateral and involves primarily the adipose tissue.[11]

Management

Various systemic treatments have been tried for PRS or "en coup de sabre" morphea including oral steroids, D-penicillamine, antimalarials, methotrexate, cyclophosphamide, cyclosporine, and azathioprine. These aggressive immunosuppressive treatments are chosen based mainly on the disease activity as well as the extracutaneous complications. For localized scleroderma, based on its clinical subtype and manifestations, there has recently been proposed a treatment algorithm, which mainly includes topical steroids, topical Vit D3 analogues, PUVA, UVA1, and methotrexate. Due to the considerable overlap of "en coup de sabre" scleroderma and PRS, and the recent classification of PRS as a subtype of linear scleroderma the same treatment approach could be followed in these patients.[12]

The treatment is usually based on reposition of adipose tissue that was lost due to atrophy. Autogenous fat grafts, cartilage grafts, silicon injections and prostheses, bovine collagen and inorganic implants are some alternatives to esthetic correction of the atrophy. However, these treatment modalities resolve just momentarily the good appearance, whereas all the structure projected in the cosmetic surgery is lost with time, due to gravity action, and the patient needs a new intervention.[13]

So, PRS is a multispecialty disorder affecting unilateral face. There is no specific treatment which is permanent. Counseling the patient with the drugs tried and surgical procedures are the only line of management. But treatments of other abnormalities like neurological disorders are difficult. As this is a progressive disorder an early diagnosis and management is essential.

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