

## Progressive facial hemiatrophy with associated osseous lesions

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### ABSTRACT

Progressive facial hemiatrophy (PFH) is a rare condition characterized by the slow, progressive appearance of a unilateral facial atrophy that affects the skin, subcutaneous tissue, muscle and bone. We report the case of a 60-year-old female patient whose cutaneous symptoms commenced in 1987 in the form of a purplish erythema on the left side of her face and neck, which subsequently remitted giving rise to an indurated region in the left maxillary region. Since 1995 until the present day, she has developed facial hemiatrophy on the left side accompanied by progressive osseous reabsorption of the upper maxilla and left mandible with atrophy of soft tissue. The association of the onset of PFH with progressive osteolysis of the maxilla has not been previously reported in an adult patient.

**Key words:** Progressive facial hemiatrophy, Parry-Romberg syndrome, bone.

### INTRODUCTION

Progressive facial hemiatrophy (PFH), or Parry-Romberg's syndrome, is a rare condition of unknown aetiology characterized by the slow and progressive appearance of unilateral facial atrophy that affects the skin, subcutaneous tissue, muscle and bone (1-3). Hemilateral extension to the trunk and members has been reported in isolated cases (1,4). It frequently commences in the first or second decade of life, progresses at a variable rhythm and stabilizes within a period of between 2-10 years.

The osseous lesions described in PFH are variable and are strictly related to the age at which the condition appears (4). When the condition commences after the age of 15 years, the lesions are considered to appear exclusively in soft tissue (5).

We present a late appearing case of PFH with important associated osseous lesions involving maxilla and left hemimandible, that we have had the opportunity to study in recent years. As we know, it is the first case reported with this particular development.

### CASE REPORT

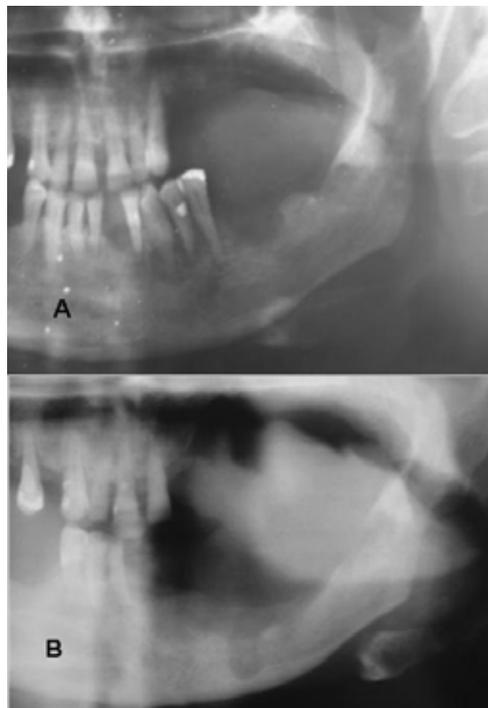
65-year-old female patient, the outstanding event in whose personal case history is having been operated on due to a hypophysial adenoma in 1979 and subsequent radioterapy. The cutaneous manifestations commenced in 1987 in the form of purplish erythematous maculae and some telangiectasia on the left side of the face and neck. The histopathological study showed a moderate inflammatory perivascular infiltrate, predominantly lymphocytic. This process evolved spontaneously in 2-3 years towards an indurated area, with the color of normal skin, localized in the left maxillary region. A cutaneous biopsy found a flattened epidermis, without rete ridge, and an increase in collagen throughout the entire thickness of the dermis. This surrounded the hair follicles and eccrine glands, which were found to be compressed by the fibrosis. Perivascular infiltration of lymphocytes and plasmatic cells was also observed. These findings led us to diagnose morphea. Since 1995 until the present day, the patient has slowly and progressively developed an atrophy on the left side of her face. This started off in the form of a depressed area in the maxillary region with atro-

phied, waxy-yellowish skin and accentuated folds. A slight upward turning of the corner of the mouth was observed on the affected side. The left half of both lips presented a decrease in thickness that resulted in poor occlusion of the orifice of the mouth with exposal of some teeth (Fig. 1). The anatomopathological study showed atrophy of the epidermis and replacement of the subcutaneous tissue by collagen. The diverse analyses carried out throughout the process, such as Hemogram, VSG, Hematic Biochemistry, ANA and anti-DNA, Complement, Immunoglobulins and Cryoglobulins, were either normal or negative. The serology for *Borrelia burgdorferi* was negative.

The patient was simultaneously assessed in the Maxillo-facial Surgery Service from 1994 onwards due to lack of cicatrisation of the left maxillary alveolar layers after dental exodontia carried out one year previously. The orthopantomographs (Fig. 2) showed progressive reabsorption of the left upper maxilla and of the left mandible (body and ascending ramus). Facial CT (1998 and 2002) evidenced importante atrophy of affected bone end soft tissue. Last CT showed a pathologic mandibular fracture, resolved without surgery (Fig. 3). Bone biopsies were inespecific. Nowadays, the patient has a good quality of life.



**Fig. 1.** Depressed area in the left maxillary region and accentuated folds. Upward turning of the corner of the mouth was observed on the affected side.



**Fig. 2.** Orthopantomographs showed progressive left mandibular reabsorption. Upper: Year 1994; Lower: Year 2002.



**Fig. 3.** CT (1998) with axial (A) and coronal (B) cuts showing bimaxillary and soft tissues atrophy. Tridimensional 2002 CT (C and D) showed a pathologic mandibular fracture and severe left side bimaxillary atrophy.

## DISCUSSION

The diagnosis of PFH in our case is based on the clinical and histopathological history, presenting atrophy of the skin and subcutaneous tissue as the most significant facts. Its onset was both delayed (in a 45-year-old patient) and insidious with purplish-erythematous blemishes that evolved to an indurate plaque suggestive of morphea, finally developing into facial hemiatrophy.

The relationship between localized scleroderma and PFH is the subject of debate. Some authors consider linear scleroderma to be an abortive form of PFH (6), and even the same disorder (7), while others suggest that an overlap exists between them (8). Sakuraoka et al. (9) reported two types of clinical presentation of PFH: the first, in the form of non-adherent depressions on the cheek without any other cutaneous manifestations; and in contrast to this, a second indurated form with marked dermal fibrosis, often associated with scleroderma in other areas of the body. For these authors, both types would represent one and the same connective tissue disorder, similar to localized or generalized scleroderma and whose differences in clinical presentations would depend rather on the region of the dermis affected. Our case would be included in this second form of presentation of PFH.

The osseous lesions described in PFH are variable and are strictly related to the age at which the condition appears (2,4). When onset occurs in 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 (4). The most frequently reported osseous lesions consist of maxillary and mandibular hypoplasia, in all dimensions, with deviation of the mid-line of the face towards the affected side (4). The mandibular body may be shorter than normal, presenting deficient verticality and a delay in the development of the mandibular angle (3). Spontaneous fractures of the mandible have been reported (10). It is common to observe eruptive delays and alterations in dental development accompanying these changes, with dental hypoplasia both in the maxillary and mandibular arch (3,4).

When the onset of the condition occurs at a later age, after the age of 15 years, the lesions are considered to appear exclusively in the soft tissue (4,5). The present study reports a case of PFH developed over an area of prior induration in an adult patient with substantial progressive atrophy of the maxillomandibular area and accompanying osseous lesions of the osteolytic type. This peculiar association of atrophy of the soft tissues with progressive osteolysis of the maxillae has not been previously reported.

The aetiology of the PFH is unknown, but many authors associate this entity and Lyme disease, supported on isolated cases (11). In our case the serology for *Borrelia burgdorferi* was negative, so this cause was excluded.

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. The cosmetic treatment is just recommended when the illness stops its evolution (12).

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