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*Case Report*

# PERIODONTAL RECESSION IN PARRY ROMBERG'S HEMIFACIAL ATROPHY

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## ABSTRACT

Parry-Romberg syndrome (PRS) is a rare disorder characterized by slowly progressive hemifacial atrophy, including the dermis, subcutaneous tissue, fat, cartilage and bone. The facial deformity is usually more severe if atrophy begins in the first decade, as growth is rapid. A 32-year-old male came for a dental visit because he was concerned about gingival recession on the central incisor and partial edentulism. Extraoral examination revealed a facial asymmetry with the midline deviated to the left and the labial commissure stretched upwards to the left. In addition, a linear morphea "en coup de sabre" on the skin of the forehead was appreciated. Intraoral examination revealed a partial edentulousness, gum recession and a diastema between the upper central incisors. In particular, the gingival recession at the upper left central incisor level was located precisely at the level of the atrophic line of the upper lip. Only two additional cases of gingival recessions at the level of the upper incisors in PRS patients have been previously reported. It is important that gingival recession is known by periodontists as well as by orthodontists and maxillofacial surgeons.

**KEYWORDS**: *gum, periodontium, recession, atrophy, gingiva*

## INTRODUCTION

Parry-Romberg syndrome (PRS) is a rare disorder characterized by slowly progressive hemifacial atrophy, including the dermis, subcutaneous tissue, fat, cartilage and bone (1). Bilateral manifestations have been reported in 5 to 10% of cases (2). Aetiology is not well understood, but the occurrence of PRS in one of two identical twins makes the possibility

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that hereditary factors are involved in its aetiology unlikely (3). Symptoms and physical findings associated with PRS usually become apparent during the first decade or early during the second decade of life (4). In some cases, the disorder is apparent at birth. The facial deformity is usually more severe if atrophy begins in the first decade, as growth is rapid. Extraoral manifestations are facial asymmetry (5). In addition, blanching of the hair or bald patches on the scalp and loss of eyelashes and eyebrows may occur (6).

Tissue atrophy can result in a deviation of the middle and lower third of the face to the affected side. The mouth may be drawn upward because of skin and subcutaneous tissue atrophy, often deviating to the affected side. Intraoral manifestations can involve both the upper lip and the tongue (7), and the soft palate can also be deficient on the affected side (8). The delayed eruption, missing teeth, deficient root development, or resorption of the roots of teeth on the affected side have been reported (9). The teeth appear clinically normal, have regular enamel, dentin, cementum and pulp, and test vital. The mandible and alveolar ridge may also be smaller on the affected side (8). The mandibular body may be shorter than average, the ramus can be deficient vertically, and the mandibular angle development can delay. The jaw disturbances can result in a unilateral malocclusion on the affected side and deviation of the facial and dental midlines (7).

*Case presentation*

A 32-year-old male came for a dental visit because he was concerned about gingival recession on the central incisor and partial edentulism. The patient reported the onset of changes on his face shortly after he was 10 years old. Extraoral examination revealed a facial asymmetry with the midline deviated to the left and the labial commissure stretched upwards to the left. A linear morphea "en coup de sabre" on the skin of the forehead, left eyebrow, and the upper lip's midline was appreciated. Furthermore, the absence of eyebrows in the medial part of the left eyebrow arch and of the upper lashes and the total absence of the lower lashes always in the left eye stood out. The upper lip at the level of the median line was considerably thinned, while in its left part, a discoloured curvilinear line was appreciated, as well as the absence of a moustache. The patient reported that he was unfamiliar with periodontal disease and lost teeth due to carious lesions. An intraoral examination revealed partial edentulousness, gum recession and a diastema between the upper central incisors.

In particular, the gingival recession at the level of 2.1 (class IV of Miller's classification) (10), which also presented root caries, was located precisely at the level of the atrophic line of the upper lip (Fig. 1).

Periodontal probing and bleeding on probing did not show significant alterations except for the recession on 2.1, which presented a two-wall defect with a 4mm periodontal probing. The upper left central incisor was vital and not mobile. The patient had performed an orthopantomography and intraoral radiographs. The patient had reported occasional clicks in the left temporomandibular joint (TMJ), especially during chewing.

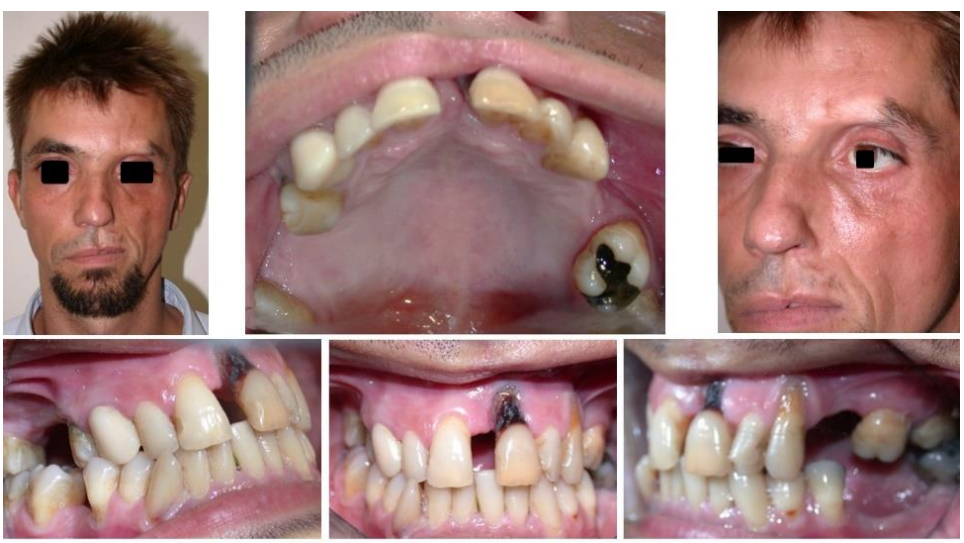

**Fig. 1.** *Intra and extraoral images highlight the characteristic skin lesions and gingival recession.*

A face and TMJ magnetic resonance imaging (MRI) scan were required. These examinations revealed sinusitis of the right maxillary sinus, a slight reducible disc dislocation of both TMJ and an asymmetry of the condyles since the left was smaller than the right. Moreover, a smaller thickness of the soft tissues of the left cheek was appreciated compared to the right one (Fig. 2).

## DISCUSSION

All the characteristic extraoral signs of PRS are present in our case. Atrophy caused by scleroderma determines alterations of the soft and hard tissues, which are more marked when it arises earlier. Therefore, the reduced involvement of facial skeletal structures is due to the late onset of the disease. In this case, the asymmetry of the condyles detected by the TMJ MRI has to be referred indirectly to the disease since a tilting of the occlusal plane determined an asymmetrical chewing function and, therefore, an asymmetric condyle different morphology. In addition, the face MRI showed an asymmetry of the soft tissues of the cheeks.

The peculiarity of this case is due to the gingival recession of the upper left central incisor since the scleroderma lesion probably caused a recession. The position coincides perfectly with the alteration present in the upper lip. Only two other cases of gingival recessions at the level of the upper incisors in the presence of localized scleroderma “en coup de sabre” involving the upper lip have been reported in the literature (11, 12).

## CONCLUSION

A rare case of a gingival recession of the upper left central incisor in a patient with a PRS and a localized scleroderma “en coup de sabre” is presented. These soft tissue diseases must be known by periodontists, orthodontists, and maxillofacial surgeons.

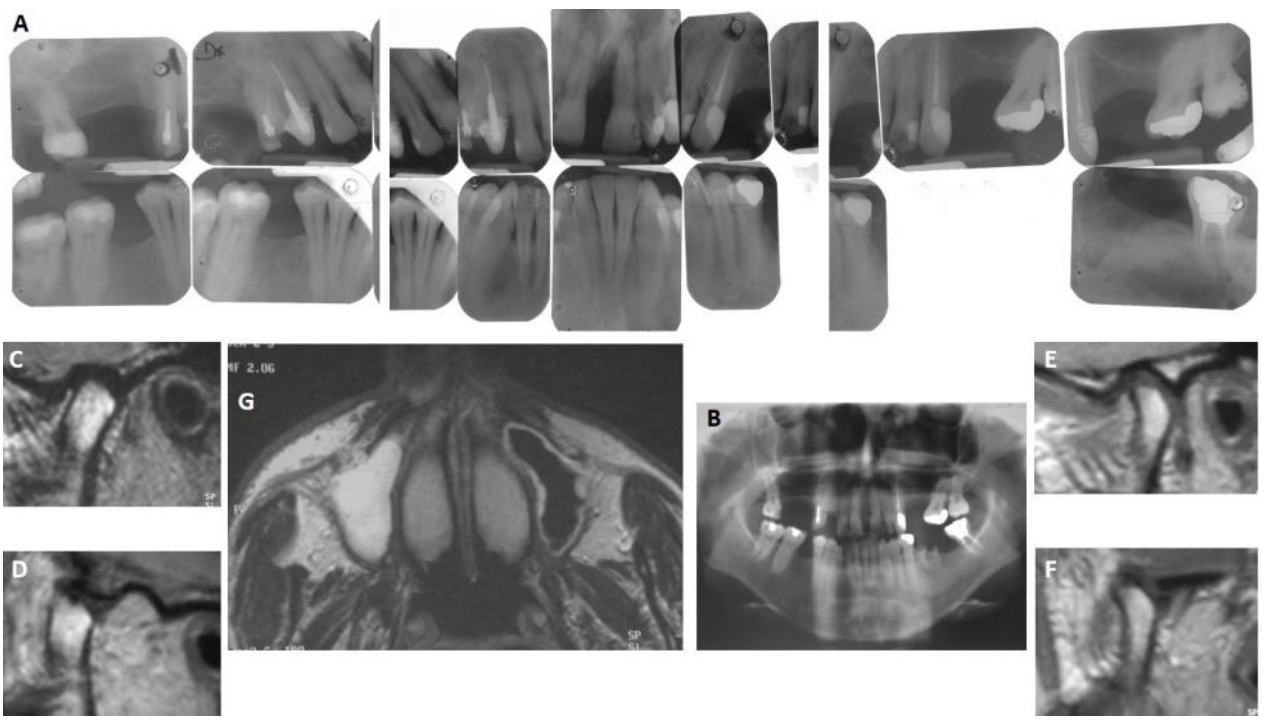


**Fig. 2.** ***A):*** *Intraoral radiographs;* ***B):*** *orthopantomography;* ***C):*** *MRI of right TMJ at closed mouth position;* ***D):*** *open mouth position;* ***E):*** *MRI of left TMJ at closed mouth position;* ***F):*** *opened mouth position;* ***G):*** *face MRI scan on axial plane.*

*Authors Contributions*

DM: acquisition of clinical and imaging data and interpretation of data; FC: drafting the manuscript; MDG revising the manuscript; LB given final approval of the version to be published.

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