

# The Late Demonstration of the Parry-Romberg Syndrome

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Received: April 04, 2016; Accepted: April 20, 2016; Published: May 09, 2016

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## Abstract

We report the case of a 46 years-old woman, who developed a linear lesion sclero-atrophic from the medial frontal region and which expanded in cranio-caudal sense, with subsequent skin and dermic atrophy.

A biopsy was performed and confirmed a dermal-epidermal sclerosis. These data, associated with the clinical characteristics, let us to diagnose a Parry-Romberg Syndrome.

Parry Romberg syndrome is a scleroatrophic syndrome, linear of the face which presents a localized partial hemiatrophy without systemic involvement.

In addition, the patient reported diffuse pain in the joints, previous autoimmune thyroiditis and reduced salivation and lacrimation bilaterally with the positive Schirmer test. The data confirmed the hypothesis of a possible S. Sjogren associated with Parry-Romberg.

This medical case is suggestive because the Parry-Romberg is a rare syndrome in adults, and usually not observed in other autoimmune disease, as S. Sjogren.

**Keywords:** Parry-Romberg Syndrome; Skin atrophy; Coupe de sabre; Sjogren Syndrome

## Introduction

Parry-Romberg syndrome, also known as progressive hemifacial atrophy, is a rare neurocutaneous syndrome, characterized by unilateral facial atrophy affecting the skin, subcutaneous tissue, muscles, and sometimes extending to the osteocartilaginous structures. Although the etiopathogenesis is still unknown, an autoimmune mechanism is suspected and the syndrome may be a variant of localized scleroderma. The syndrome has a higher prevalence in females and typically appears between 5 – 15 years of age [1,2].

The clinical manifestations are highly variable. Initially, the disease involves the region of the buccinator and temporal muscles, and gradually extends to the whole unilateral facial side. It causes atrophy of skin and adnexa, deepening to the subcutaneous tissue. The mouth and nose are typically deviated

towards the affected side of the face.

Initially, it begins as an area of sclerosis from the frontal region of the scalp, with hair loss. Subsequently, it appears a scar which extends along the median part of the face, called "coup de sabre" [7].

In 20% of cases, the affected area may be predisposed to altered pigmentation [4, 5].

Ocular involvement is relatively rare. [3]

Also, Parry-Romberg Syndrome is usually not observed in other autoimmune disease, including Sjogren Syndrome, which is a chronic inflammatory autoimmune disease characterized by destruction of exocrine glands, in particular lacrimal and salivary glands [3, 6, 8].

## Materials and Methods

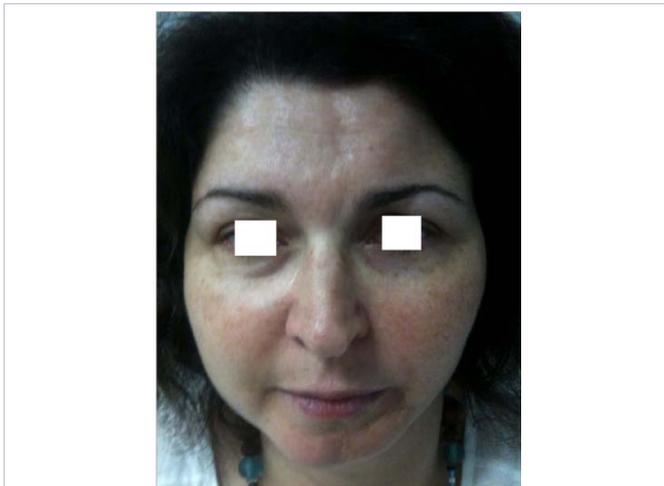
We report the case of 46 years-old woman who, during 3 months, developed a linear sclero-atrophic lesion in the medial frontal region, extending in cranio-caudal sense, with involvement of the glabellar region, nasolabial fold and ipsilateral chin angle, with subsequent skin and dermic atrophy (**Fig. I-II**).

We performed a cutaneous biopsy (**Fig. III**), which highlighted sclerosis of the dermal-epidermal junction and the reticular dermis in the context of which it was present a lymphomononucleated chronic inflammation with perivascular and periadnexal distribution. In sections, at the level of the hair follicle hairy, three structures were visible in complete atrophy.

According to the clinical and histological characteristics, it is done diagnosis of a saber scleroderma (S. Parry-Romberg).

The patient has performed the Magnetic Resonance Imaging (MRI) and electromyography, but in our case it did not highlight alterations worthy of note.

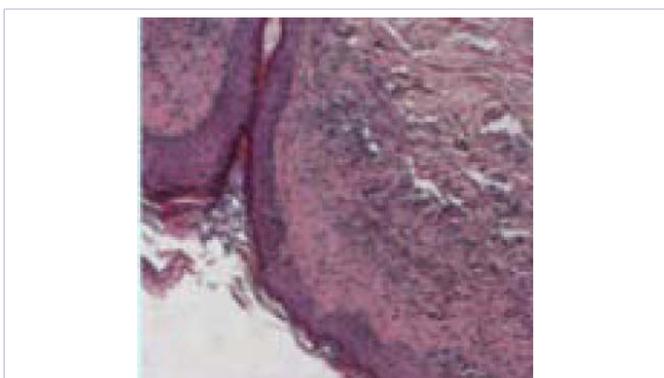
In addition, the patient reported diffuse pain in the joints, previous autoimmune thyroiditis and symptoms and signs suggestive of Sjogren Syndrome, as bilateral xerophthalmia, xerostomia with the positive Schirmer test.



**Figure I:** Patient with a linear sclero-atrophic lesion in the medial frontal region, with involvement of forehead, nose and chin (coup de sabre).



**Figure II:** Patient with a linear sclero-atrophic lesion in the medial frontal region, with involvement of forehead, nose and chin (coup de sabre).



**Figure III:** Interstitial lichenoid chronic infiltration forming a band in the reticular dermis surrounding a pilar follicle. H&E 100X.

Laboratory examinations showed negative antibody.

## Conclusion

This medical case is suggestive because the Parry-Romberg is a rare syndrome in adults, in fact it typically appears between 5 – 15 years of age, while in our case the disease was diagnosed

in a 46-years-old woman [9].

In addition, an other particular aspect is that Parry-Romberg Syndrome is usually not observed in other autoimmune disease, while in our case it is associated with Sjogren Syndrome. In fact the association between Parry-Romberg and the S. Sjogrenreopens the discussion about autoimmunity because, in our case, the patient is suffering simultaneously from a localized connectivitis, as Parry-Romberg, and a systemic connectivitis, as Sjogren Syndrome [7].

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