Case Report

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Bone Resorption of the Jaws and Occlusal Disturbances in Parry-Romberg Syndrome

Mendez LA¹, Martinez R², Briones AM³ and Sosa LR⁴

¹Basic Sciences, University of Montemorelos, School of Dentistry, Montemorelos, NL, Mexico

²Oral Pathology, University of Montemorelos, School of Dentistry, Montemorelos, NL, Mexico

³Dentistry National Social Service, University of Montemorelos, School of Dentistry, Montemorelos, NL, Mexico

⁴Advance Dentistry, University of Montemorelos, School of Dentistry, Montemorelos, NL, Mexico

***Correspondence:** Luis A Mendez, Basic Sciences, University of Montemorelos, School of Dentistry, Montemorelos, Avenida Libertad 1300 Poniente, Matamoros, NL 67530, Mexico, E-mail: luismendez@um.edu.mx

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Abstract

Parry-Romberg Syndrome is a very rare condition with a very low prevalence. Therefore, it is important to report particular cases that will continue helping the medical community with the identification of such condition. We present a case of a young woman worried because she has observed a decrease in size of one side of her face as well as other symptomatology.

Keywords: Parry romberg syndrome, Occlusion, Osseous resorption

Introduction

Background

Parry – Romberg Syndrome or progressive hemifacial atrophy was first described in 1825 by Caleb Parry and posteriorly in 1846 by Moritz Romberg and Eduard Hensch. It is a degenerative rare disease characterized by a unilateral slow progressive atrophy of one side of the face being the left side the most involved. Subcutaneous cellular tissue, fatty tissue, cartilage and underlying bone structures are affected causing an asymmetrical appearance of the face. The prevalence is at least 1/700 000. The incidence is higher in women than men, in a ratio of approximately 3:2 [1],[2]. The disease initiates within the first and second decade of life. The atrophy might be preceded by hyper or hypopigmentation of the affected side. It also presents neurologic conditions such as migraine, trigeminal neuralgia, epilepsy, among others, accompanied by ocular lesions and conditions affecting hair, sebaceous and sweat glands. In severe forms of the

disease, some other anatomical structures can be involved, for instance trunk, extremities and even structures of the contralateral side [3],[4]. Soft and hard tissues of the oral cavity are affected. Atrophy of the lips, tongue, soft palate and mastication muscles of the affected side is noted, as well as a decrease in the size of the mandible and maxilla leading to malocclusion disturbances and even fractures. Partial anodontia, delayed eruption and morphological root disturbances including root resorption are the most common teeth conditions found in this syndrome. There is no genetic component able to explain this disease [5],[6].

Case Presentation

The patient is a 23-year-old female with no relevant medical history. She came to our university clinic for a routine dental examination and prophylaxis. The patient referred to us that she had noted some asymmetric changes on the left side of her face since she was 13-years-old especially an important decrease in the size of the nose ala accompanied with dental mobility. Also, visual defects and

alopecia on the contralateral side have been present since she started noticing these changes. Clinical examination revealed that soft tissues of the left side of the face tend to invaginate towards the center between the nostril and the left angle of the mouth (Figure 1). During oral examination, we observed an intrusion of both dental arches on the affected side leading to a lack of centric occlusion as well as a deviation of the midline to the affected side (Figures 2 and 3). Diagnosis of Parry-Romberg Syndrome was made based on clinical examination and symptomatology described by the patient. The patient was referred to an institution that supports people with rare diseases for proper treatment and follow-up. CT was performed revealing a marked osseous resorption in quadrants two and three in molar and bicuspid areas being more severe in the upper quadrant (Figure 4).

Figure 1: Atrophy of the left side of the face, invagination of tissues and small ala.

Figure 2,3: Open bite of the affected side due to the atrophy and deviation of the midline to the left side.

Discussion

Parry-Romberg Syndrome is a rare disease that encompasses aesthetics and functionality of the involved tissues. Considered differential diagnosis for Parry-Romberg Syndrome are Rasmussen Syndrome, circumscribed scleroderma and hemifacial dystrophy [6],[7]. It is a self-limiting condition and there is no developed cure for such disease [8]. Treatment in Parry-Romberg Syndrome is multidisciplinary. A wide range of treatments exists in the re-establishment of normal function and aesthetics: silicone implants, bovine collagen implants, hyaluronic acid injections, autogenous fat grafts, cartilage grafts, and others.

Figure 4: Alveolar bone resorption being more severe in the upper maxilla, shown by CT.

The aim of these treatments is primarily the reposition of fat tissue lost due to the atrophy [9],[10],[11]. Regarding the orthodontic treatment during the atrophy, Grippaudo et al. established that it will limit the osseous deformity stimulating the mandibular growth, thus optimizing the symmetrical function of the mandible since the craniofacial development is mainly determined by muscular function [12]. Each case of Parry-Romberg Syndrome will vary in presentation and severity. Therefore, when approaching the case, treatment management will be different but with the aim of improving the quality of life.

Conflict of Interest Disclosure

The authors declare that there is no conflict of interest regarding the publication of this paper.

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