

Individual replacement prosthesis: contemporaneous treatment of parry-Romberg syndrome. A case report

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Abstract

Introduction: Parry-Romberg syndrome or progressive hemi facial atrophy is a slow and localized disorder of facial tissues that appears in early and middle age characterized by degeneration of the subcutaneous tissues which affects the symmetry of the face.

Case report: 18-year-old male individual who presented facial asymmetry with evidence of alterations in both the intraoral and extraoral volume.

Conclusion: In order to get an adequate treatment of the parry-romberg syndrome an interdisciplinary approach is required after an adequate study of the case to execute a treatment supported by a good scientific base, achieved through the use of the design and manufacture of stereolithography.

Keywords: parry-romberg, prosthesis, scleroderma, hemifacial atrophy

Introduction

Parry-Romberg syndrome also known as progressive hemifacial atrophy (PHA) is a rare degenerative disorder that affects the tissues on one side of the face, including the soft tissues, muscular fibers, adipose tissues or connective tissues as well the hard tissues such as bone tissue or cartilage. This case was first described in 1825 by Caleb Hillier Parry and then by Moritz Heinrich Romberg in 1846.¹

This pathology is attributed to a variety of extrinsic and intrinsic factors that have not yet been proven, which is why this pathology is considered idiopathic, however, these factors can be broken down into prenatal and postnatal traumatic factors that can lead to the appearance of this chronic degenerative process, likewise the presence of vasculitis in the structures affected by this syndrome suggests a possible vascular nature that is part of the onset and development of PHA.^{2,3}

In parallel, it is considered that, due to the existence of different facial asymmetries related to the history of the individual's family history, the etiology of PHA is possibly related to genetic. Some studies have revealed the existence of antinuclear antibodies similar to those found in diseases such as lupus and rheumatoid arthritis, suggesting the autoimmune etiology as a justifiable reason for the onset of Parry-Romberg syndrome.⁴

Within the clinical manifestations is the slow and localized degeneration on one side of the face and in advanced cases affects the osseous and cartilaginous structures of the craniofacial sector giving a hypoplastic appearance to the face that is accentuated by the degradation of the subcutaneous tissues and the pigmentation of the

skin, the oral cavity is also affected, since the epithelial lining tissues, muscles and connective tissue suffer from the same affections that manifest the facial structures.⁴

Is common the occurrence of neuropathic processes in nerves associated with neuralgias of cranial nerves III, V, VI and VII, at the same, time the epileptic syndrome appears as convulsions and in some cases, atrophy affects the intracranial structures causing paresthesia, dysesthesia and hemiparesis.¹

The subcutaneous tissue is gradually reduced, including the connective tissue and adipose tissue, in a similar way, the scleroderma that can start in the forehead getting to the neck and causes hyperchromic or hypochromic skin pigmentation.⁵

Although the characteristics that the PHA presents are visible and easily evaluable in its beginnings and due to its slow evolution it is possible to confuse it with different pathologies of similar manifestation, among them the most common, the scleroderma (morphea) in coup de saber (ECDS) that exhibits a marked hemifacial scleroderma that is related in large measure to cases of PHA.⁵⁻⁸

Rasmussen's encephalitis is an inflammatory hemiencephalic condition characterized by the appearance of seizures in those who have it in a similar way as can be seen in many individuals with PHA and in the other hand the lipodystrophy that characterizes a Barrequer-Simons syndrome in its beginnings in a localized way in the face can also become confused with the PHA.⁹⁻¹²

PHA, appearing during the first two decades of life, commonly alters the correct development of facial bones, so it is possible to confuse

it with different facial asymmetries of various syndromes, such as Goldenhar's.¹³⁻¹⁵

Case presentation

18-year-old male individual who attended a dental and maxillofacial consultation in the company of a guardian for presenting a case of facial asymmetry of 5 years of evolution on the right side of his face. the guardian reported that the individual suffered antecedent of traumatism in craniofacial region at eight months of age, the guardian did not report any pathological or neurological manifestations, the individual was found lucid, oriented, without posture alterations, presenting a fluid and coordinated march, however, he referred a couple of episodes of seizures in previous years, for which he denies receiving treatment.

Physical examination revealed a hemifacial asymmetry in the middle and lower thirds of the face in the right side, also in the upper third no alterations were evidenced. The facial midline was deviated to the right side, where the nasal septum showed a lateral inclination towards the affected side with elevation of the right wing of the nose and in the same way an elevation of the right side lip commissure was observed along with a slight thinning of the upper lip. The right auricular pavilion was found without alterations in bilateral normal position without discrepancy between the contralateral sides. There were no alterations in the eyes or eyelids (Figure 1).



Figure 1 frontal and semi-profile view of the individual.

During palpation, a normotonic consistence and asymptomatic response was found as well as an absence of nodules in the preauricular and postauricular region, submandibular and cervical ganglion chains were found without masses and adenopathies, a lax consistency of the skin associated with the thinning of the skin in the right facial region accompanied by a significant reduction of the space of connective tissue and the volume of adipose tissue, in addition the mimetic muscles were found without a loss of function.

Seeking to correct the asymmetry presented by the individual, different treatments were taken into account that would provide a high probability of success and acceptance by the individual, to finally perform an individual replacement prosthesis that is known as a procedure where the facial volume is returned with a Titanium plate designed by stereolithography design and fixed with titanium screws in the malar and zygomatic bone.

The procedure was performed under general anesthesia, the replacement prosthesis was prepared in advance using 3D designs and

stereolithographic printing for proper adaptation to the individual's malar bone (Figure 2).



Figure 2 Individual replacement prosthesis fixed in the malar and zygomatic bone.

Finally, it was possible to restore the volume in the zygomatic region (Figure 3) (Figure 4), providing the individual the necessary conditions to continue the rehabilitation process in conjunction with the area of plastic surgery and orthodontics.



Figure 3 before and after semi-profile view.



Figure 4 Before and after frontal view, the black dotted lines correspond to facial midline and the division of the three facial thirds in proportional longitudes, the yellow dotted line represent the level of the wing of nose on the left side of the face and the red dotted line represent the level of the wing of nose on the right side of the face, with a 7mm of discrepancy. The blue dotted line represent the level of the labial commissure on the left side of the mouth and the green dotted line represent the level of the labial commissure on the right side of the mouth with a 6mm of discrepancy.

Discussion

Parry-Romberg syndrome is known as a rare unilateral facial atrophy that progressively degenerates the adipose and connective tissues, skin, muscles fibers and some cranial nerves responsible for eyeball mobility (III, IV, VI) along with the sensitive and motornerves (V,VII). In the same way, vascular irrigation is affected and, in more advanced cases, an osseocartilaginous condition occurs. All of these compromise the three thirds of the face in only one side, causing a progressive underdevelopment of the facial bones which in turn generate an asymmetry together with intracranial alterations that can trigger episodes of seizures and headaches.¹⁶

In the middle and lower thirds, a significant reduction in facial volume revealing a sunken aspect of the face associated with the degradation of the subcutaneous tissue and thinning of the skin with or without the skin pigmentation in a hypochromic or hyperchromatic way. In the oral cavity the manifestations are evidenced in the same way with a reduction of connective tissue and the atrophy of the lining epithelium of the tongue, cheeks and the gingiva.³

The etiology of this condition is unknown but associated with different factors such as autoimmune factors, genetic factors, vascular factors and traumatic factors. The great number of characteristics of this pathology allow a precise diagnosis, but in mild cases it resembles other pathologies with similar characteristics that can vary from scleroderma, vasculitis, lipotrophy, convulsions, and hypoplasia.^{2,3}

The aforementioned case mentions a mild Parry-Romberg syndrome in an individual within the second decade of life with an appreciable degree of manifestation on the right side of his face for which an implant was chosen with a titanium maxillomalar replacement prosthesis anchored with fixing screws to return the volume to the face correcting the symmetry and adding to the interdisciplinary treatments that the individual requires.¹⁷

Reconstructions and modifications in the facial architecture are one of the great challenges for the surgeons who commit to perform the best possible procedures, hence the use of advanced technologies that can improve the quality of treatments has become a clear need in all cases in which it can be carried out. The maxilla is one of the most important structures in the anatomy of the stomatognathic system which has functions of phonation, mastication and deglutition, besides giving form and volume to the facial region, reason why its modification in cases of being necessary should be carried with great precision in order to avoid affecting facial symmetry or to correct defects and imperfections, which leads to the use of design technology, modeling and 3-D impressions for the development of cutting guides for osteotomies used in autologous grafts and prosthetic additives with intimate relationship with the surfaces to which they will be fixed thus returning the form and the function to the craniofacial structures.¹⁸

In order to make the right decision to use an individual replacement prosthesis, different treatments, such as demo-fat grafts or porous polyethylene, are commonly used to correct facial asymmetries. The aforementioned were taken into account as a way to aesthetically model the facial volume, correctly fixed on the bone surface could return the volume required to correct the facial asymmetry generated by the Parry-Romberg syndrome, however with the purpose of avoiding the possible resorption of skins grafts and a better acceptance of the titanium prosthetic implant by the individual, the final choice converged on the individual titanium replacement prosthesis designed with the support of 3-D stereolithographic models.^{19,20}

Conclusion

This singular clinical case known as progressive hemifacial atrophy or Parry-Romberg syndrome is a degenerative condition with a slow and delimited course on hemifacial tissues with idiopathic etiology, capable of causing a notorious facial asymmetry and easily misdiagnosed with different pathologies that present similar characteristics.

Its treatment must be carried out by an interdisciplinary group in order to provide the individual with a functional and aesthetic rehabilitation using all the necessary safety measures and taking advantage of the different avant-garde procedures supported by the available scientific knowledge that will help offer a quality service.

All of the above makes it clear that cases of bone regeneration can be performed more effectively and safely using stereo lithography technologies, which provide the most accurate parameters for the design and manufacturing of individual titanium prostheses with perfect adaptation and acceptance by the individual, avoiding failure and inconclusive treatments.

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Conflict of interest

The author declares no conflict of interest.

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