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Case Report

Progressive hemifacial atrophy or Parry Romberg syndrome case report and literature review

Andrade-Garduño J.[1]*, Lazcano-Blanco P.[2], Lara-Ralon L.[1], Rodríguez-Rojas E.[3], Hernández-Moreno A.[1], Arturo Hernández-Agallo R.[1]

[1]Department of Plastic and Reconstructive Surgery, Hospital General, Lic. Adolfo López Mateos, Ciudad de México, México
[2]Department of General Surgery, Hospital General, Dr. Fernando Quiroz Gutierrez, Ciudad de México, México
[3]Department of Plastic and Reconstructive Surgery Service, Hospital Regional, Adolfo López Mateos, Ciudad de México, México

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***Correspondence:**
Dr. Andrade-Garduño J.,
E-mail: javier.andrade.gar@gmail.com

ABSTRACT

Parry-Romberg syndrome is a condition characterized mainly by left-sided hemifacial atrophy that affects skin, subcutaneous tissue, muscle, and bone. The pathophysiology is not established and several theories have been proposed, among which lymphocytic neurovasculitis manifested as a chronic vascular lesion measured by incomplete endothelial regeneration stands out. The actual treatment consists of autologous fat transfer, making voluminous grafts to restore symmetry and improve the quality of the skin. An overcorrection is recommended to compensate for the reabsorption estimated of up to 80%. The treatment consists of repeated applications until correction is achieved, there is currently no consensus regarding the preparation of the graft. The objective of this work is to propose an alternative to conventional treatment by enriching fatty tissue with platelet-rich plasma, which favors the formation of extracellular matrix, collagen fibers, and angiogenesis in an accelerated manner. We conclude that the use of platelet-rich plasma significantly improves skin quality and fat graft integration.

Keywords: Parry-Romberg, Hemifacial atrophy, Lipofilling, Platelet-rich plasma

INTRODUCTION

Parry-Romberg syndrome, or progressive hemifacial atrophy, is a rare craniofacial disorder characterized by hemifacial atrophy of skin, subcutaneous tissue, fat and in severe cases, muscle and bone.[2] Its name is attributed to 2 great physicians. The clinical data of hemifacial atrophy were initially described by Dr. Caleb Hillier Parry, which became known in 1816 after he suffered a stroke that forced him to abandon the practice of clinical medicine. Thanks to the support of his daughter, he was able to continue his written observations, including the observations on primary hemifacial atrophy. After the Doctor's death, his son Charles Parry, in 1825, published these writings.[1] It was not until 1846 that Dr. Moritz Heinrich Romberg revolutionized the field of neurology in Europe by describing in more detail the clinical manifestations of hemifacial atrophy. Disease that appears in the first 2 decades of life, usually accompanied by other disorders such as: extracutaneous manifestations of the disease including neurological, ocular diseases. Being common to present at any stage of the disease.[2]

Parry Romberg syndrome is currently classified as mild, moderate/ severe to stratify disease and its management.

Mild disease: Atrophy of skin and subcutaneous tissue limited to a single sensory branch of the trigeminal nerve.

Moderate disease: Atrophy limited to 2 branches of trigeminal nerve.

Severe disease: Atrophy involving all 3 branches of the trigeminal nerve or any bonyinvolvement.[2]

The pathophysiology of hemifacial atrophy is basically unknown, in recent years theories have emerged about the etiology, one of the most accepted theories today is described by Mulliken who called this entity a "lymphocytic neurovasculitis" involving chronic cell-mediated vascular injury with incomplete endothelial regeneration with respect to the participation of the long branches of the trigeminal nerve.

CASE REPORT

Female patient, 30 years old, with important antecedents congenital: Parry Romberg syndrome, perinatal: septic arthritis in right hip at 1 month of birth meriting surgical treatment. Rasmussen syndrome (autoimmune epilepsy, diagnosed in 2017) in treatment with: levetiracetam, magnesium valproate and topiramate, lacosamide, in treatment every 6months with rituximab (last application in Dec. 2022). Surgical history: rhinoplasty 10 years ago, intake and application of fat graft in 2017 and 2018 (performed at hospital general Dr. Manuel Gea Gonzalez). hospitalizations: Last hospitalization for use of rituximab, in 2017 as study protocol.

She refers the onset of the current condition at approximately 14 years of age, being managed with a diagnosis of scleroderma, presenting a plaque at the level of the right zygomatic region, then facial asymmetry until 2016 when she had a seizure, for which reason it was decided to perform a diagnostic protocol with the result of Parry Romberg Syndrome, being treated by taking and applying a fat graft on two occasions in 2017 and 2019, without complications, continuing with facial asymmetry, so she was evaluated by our service and decided to undergo surgical treatment.

Physical examination showed alteration of the normal anatomy in the skull, with the presence of subsidence in the right temporal region, facial asymmetry, with hypotrophy in the right hemiface, lipodystrophy in the right hemiface, as well as thinning of the upper andlower lip with predominance of involvement of the right side. Rest of the examination, in the upper extremities with limited mobility of the left extremity, with mild edema in the hand and forearm, adequate capillary filling and present sensitivity.

Laboratory and cabinet studies-10.02.23: Hb 12, Ht 36.7, Leu 11, Neu 52% Plat 355, Glu78, Ac Ur 3.4, BUN 5.6, Urea 11.9, Cr 0.46, Na 134, k 4.35, CI 108, TGL 253, ColT 173, Alb 3.6, Prot T 6.3, T3 127.3, T4 12.6, TSH 3.9, TTPA 29, TP 10.5, INR 0.89.

X-rays of the skull (anteroposterior and lateral views) were performed, showing depressedright temporal region.

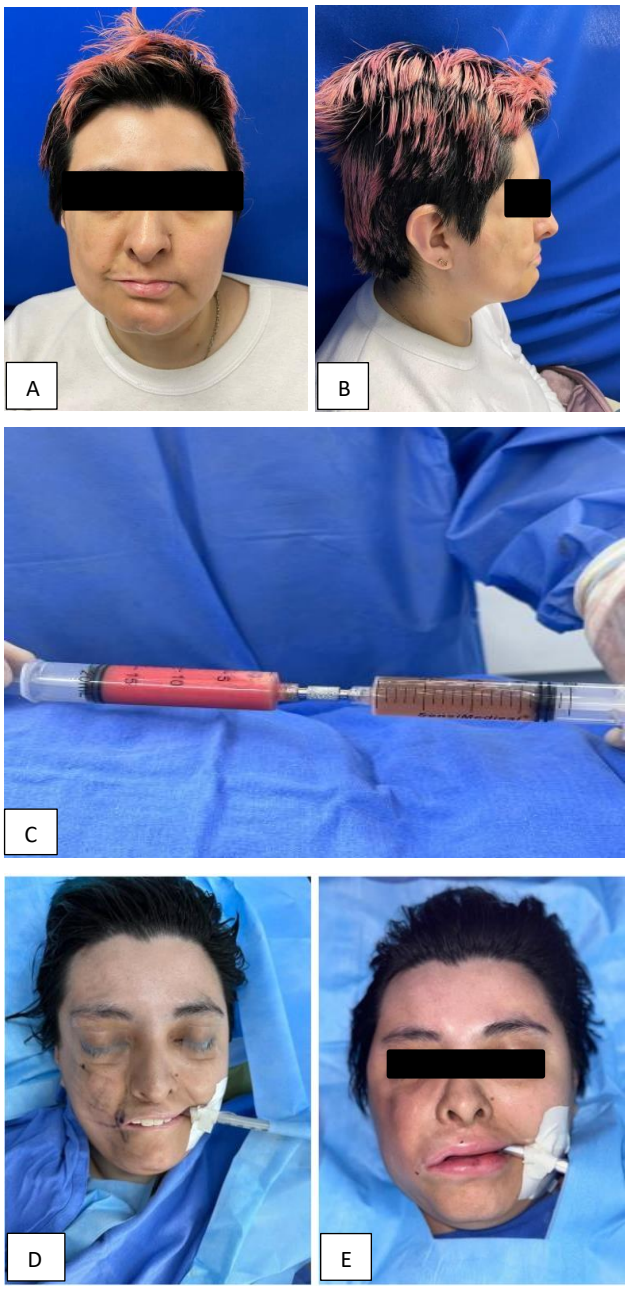

Figure 1 (A-E): Frontal view, marked atrophy of right hemiface, with ipsilateral chin deviation. Lateral view, area of skin hyperpigmentation, depression in right frontoparietal region and malar hypoplasia. Pre-surgical marking. Preparation of lipograft with platelet-rich plasma. Result after fat transfer in affected areas.

DISCUSSION

Progressive hemifacial atrophy is an uncommon and devastating entity that results in severe disfigurement and functional impairment after years of progressive hemifacial atrophy.[3] The atrophy will end when the disease goes into remission, leaving important sequelae

of destruction. Different methods have been proposed to treat these defects such as fillers and implants or free tissue transfers.[3]

Autologous fat transfer

Lipofilling (fat transfer) provides an option for voluminous flaps, improving skin quality. fat transfer. Autologous fat transfer has long been a preferred method for facial volume augmentation because of its ease of access, biocompatibility, and versatility.[3,4] The main limitation to this therapeutic option is the persistent inflammatory state to which these patients are subjected, the poor environment at the recipient site and chronic use of corticosteroids, which makes fat transfer more susceptible in these patients. Despite these limitations, fat transfer still represents an important treatment modality. Some surgeons have attempted to improve the unpredictable survival of fat by combining cell cultures including platelet-rich plasma, adipose-derived mesenchymal stem cells or bone marrow-derived mesenchymal stem cells.[1,3]

Among the challenges in using autologous fat grafting are the variable resorption rates of the injected aspirate. According to the literature some authors suggest an overcorrection ranging from 20-50% to compensate. The variability in fat resorption is due to a combination of factors such as: donor site, method of fat harvesting, graft preparation and recipient site. Some studies show more satisfactory results such as the malar area and lateral regions of the cheeks (medial region) followed by the chin, nasolabial fold. Although it has not been justified why, the most widespread theories in the literature are related to the degree of vascularization and mobility of the recipient site and the quality of the donor site.[1,3,4]

During the first sessions, deep compartments are grafted to establish.

The appropriate tissue projection and subsequent sessions focus on the superficial compartments to refine facial contours. In mild to moderate cases, serial injections alone may generally be sufficient, while patients with severe presentations generally undergo fat grafting in conjunction with other procedures. The fat graft undergoes some degree of post-procedure resorption and hypertrophy if postoperative weight gain occurs. It has been seen that patients who have undergone previous craniofacial bone surgery have worse graft retention than the general population, retaining only 40% of the original graft. Most of the volume resorption will be evident at the 3-month evaluation, where subsequent lipografting may be beneficial for correction of asymmetry in the initial procedures.[1,3]

CONCLUSION

Autologous fat transfer in combination with various cell cultures such as platelet- rich plasma is a viable and long-lasting alternative for a better integration of fat grafting in patients with this pathology. It is currently the treatment of choice for surgeons due to its efficiency, reproducibility and low cost, with remarkable patient satisfaction and few complications.

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