

Autologous fat transplantation for the treatment of progressive hemifacial atrophy (Parry-Romberg syndrome: case report and review of medical literature)

Lipoenxertia autóloga no tratamento da atrofia hemifacial progressiva (síndrome de Parry-Romberg): relato de caso e revisão da literatura

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Abstract: Parry-Romberg syndrome, also known as progressive hemifacial atrophy, is a rare disorder characterized by slow and progressive hemifacial atrophy. The treatment offered for the syndrome generally aims at improving aesthetics. Fat grafts, silicone injections or acrylic prosthesis are alternatives suggested for correction of facial atrophy. Currently, the recommended technique for correction of facial atrophy is cosmetic dermatologic surgery with autologous fat grafting. This study reports a case of Parry-Romberg syndrome and demonstrates that dermatologic surgery can relieve serious damage to the patient's anatomy, starting from the discussion of the therapeutic aspects of the syndrome with emphasis on autologous fat grafting.

Keywords: Facial hemiatrophy; Lipodystrophy; Transplants; Homologous transplantation

Resumo: A Síndrome de Parry-Romberg, também conhecida como atrofia hemifacial progressiva, é uma doença rara caracterizada por lenta e progressiva atrofia de hemiface. O tratamento ofertado para a síndrome, geralmente, visa melhorar o aspecto estético. Os enxertos gordurosos, as injeções de silicone ou as próteses de acrílico são alternativas sugeridas para correção da atrofia facial. Atualmente, a técnica recomendada para correção da atrofia facial é cirurgia dermatológica cosmética com lipoenxertia autóloga. O objetivo deste estudo é relatar um caso de SPR e demonstrar que a cirurgia dermatológica pode aliviar danos sérios à anatomia do paciente, a partir da discussão dos aspectos terapêuticos da síndrome, com ênfase na lipoenxertia autóloga.

Palavras-chave: Hemiatrofia facial; Lipodistrofia; Transplantes; Transplante homólogo

INTRODUCTION

Progressive hemifacial atrophy was described by Parry, in 1825, and better studied by Romberg, in 1846.^{1,2} Also known as Parry-Romberg syndrome (PRS), this rare disorder is characterized by unilateral facial atrophy which affects the skin, subcutaneous, muscle and rarely, osteo-cartilagenous adjacent structures. The left hemiface is the most affected and it can spread to the ipsilateral hemi-body.

PRS involves dermatomes or multiple branches of the fifth cranial nerve.³ Its etiology is unknown suggesting as pathogenic factor autoimmunity, abnormality of the trigeminal nerve, cervical-facial trauma or infections.³⁻⁵

Both sexes are equally affected.⁶ It begins in adolescence, in general in the first or second decade of life and develops during a period of time which

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varies from two to twenty and then it becomes stationary.³

Treatment offered for the syndrome usually aims at improving aesthetics. There are several alternatives suggested to correct the defects of the face such as dermal graft, fat, cartilage or bone, neighborhood retail, inclusion of tantalum, acrylic and solid or liquid silicone.⁴ The choice of technique depends on the conditions of the patient, life expectancy and severity of the lesion.

The aesthetic correction for PRS is autologous fat grafting. This technique has the advantage of a better cost-benefit, better skin texture and more natural contours and facial expressions. However, unpredictability of results due to the possibility of absorption and consequent loss of desired volume are still limiting factors for fat grafting.

The objective of this study is to report a case of PRS and to demonstrate that cosmetic dermatologic surgery can relieve serious damages in the patient's anatomy, starting from the discussion of the therapeutic aspects of the syndrome, with emphasis on autologous fat grafting.

CASE REPORT

JCL, female, aged 38, mullato, single, from Bom Jardim (MA) and coming from Fortaleza (CE), housemaid, catholic.

Patient attended at the Plastic Surgery Service from the Walter Cantídio University Hospital in August, 2008, complaining of a "problem on the face". The patient, previously healthy, reports that when she was 15 years old she noticed the appearance of a white spot on the right hemiface, followed by progressive "sinking" of this hemiface. The symptoms progressed gradually and during the consultation she complained of worsening of the sinking of the right hemiface and ipsilateral temporomandibular joint arthralgia.

When examined, it was observed important atrophy of the right hemiface with preserved sensibility, strength and mobility (Figures 1 and 2).

The patient was treated with autologous fat grafting (Coleman type). Collection was made from the abdomen. The procedure was performed in two surgical times. The amount of fat to be sucked was estimated from the atrophy of the affected face.

The first surgical time occurred on 5th October, 2009. Under general anesthesia it was infiltrated a local anesthetic solution with lidocaine 1% and 1:100.000 of adrenalin in the abdomen and 100 mL of fat tissue was sucked with a 5 mm liposuction cannula. From this amount, a total volume of 50mL of fat was washed with saline and injected using 26 G needles, in angle from 30° to 40°, according to the tun-



FIGURE 1:
Patient's preoperative



FIGURE 2:
Patient's preoperative

nels technique, spaced approximately 0,5 cm, in the patient's atrophic hemiface.

After two months, some areas needed surgical repair with new injections of fat. It was performed the second surgical time on 2nd December, 2009, also under general anesthesia. It was infiltrated serum anesthetic solution and xylocaine 2% more adrenaline 1:200.000 in the abdomen and a total volume of 50mL of fat was sucked with a 5 mm liposuction cannula and injected using 25G needles, in angle from 30° to 40°, also in accordance with the tunnels technique, spaced approximately 0,5 cm, in the patient's right hemiface.

The surgical result was assessed through analy-



FIGURE 3: Patient's post-operative after six months following the second surgery

sis of preoperative photographs and return 6 months after the second surgery and also through analysis of the patient herself. (Figure 3).

DISCUSSION

Unilateral atrophy of the face, as described in the case above, is a clinical symptom that occurs in different diseases. Diagnosis can be difficult specially at the beginning of the condition. Progressive hemifacial atrophy, also known as Parry-Romberg syndrome is characterized by progressive unilateral atrophy of the facial tissues.⁷ Etiology of PRS is not established yet and probable causes are neuritis of the trigeminal nerve or changes of the sympathetic nervous system.⁴ Other causes of unilateral facial atrophy include the Goldenhar syndrome and facial trauma. Unilateral facial lipodystrophy is also observed after nerve injury due to ischemia or mechanical trauma.⁸ Differential diagnoses to our patient are initially possible but the recommended treatments for the pathologies previously cited are identical. However, it is important to point out that the early diagnosis of facial atrophy is extremely important for an early correction of the symptoms.

In general, the treatment offered for PRS patients aims at improving their physical appearance, in this specific case, reconstructive surgical treatments should be assessed after clinical inactivity of the disease. Clinical treatment of the disease with immunosuppressive drugs or other medications also used in scleroderma such as chloroquine and calcipotriol is discussed by many authors but still there is no scientific evidence for the use of such medication to treat PRS.

The treatment often offered to the syndrome is based on the replacement of tissue that was lost due to atrophy.⁹ There is preference for autografts due to a lower risk of tissue rejection and, consequently, smaller local and systemic inflammatory reaction that would allow graft loss or other co-morbidities to the patient. Autologous fat tissue grafts are the most used. Fat tissue is malleable, has good availability and easy access. In the reported case we opted for the removal of abdominal fat with liposuction as it is a widespread technique. In the 1980s, Fournier and Illouz were the first to develop a method to transfer fat from the abdomen.¹⁰ This technique presents small intraoperative trauma, few postoperative complaints and the possibility to repeat the technique in patients who have already been operated according to the need. The main disadvantage is the unpredictable extent of fat tissue reabsorbed in the area which reduces the clinical efficacy of the procedure and frequently requires late repair.¹¹ An unquestionable fact: the result of the surgery depends on atraumatic manipulation of the operated area, therefore, it depends directly on the surgeon's experience. An expected reaction includes swelling of the donor and recipient areas. More rarely, hematoma or seroma can appear in the abdominal wall.¹² There have been reports of unilateral loss of vision and cerebral infarction due to fat embolism after fat-grafting as extremely rare complications.

Alternative methods used for treating PRS are application of dermal fillers agents such as bovine collagen implants, silicone implants and injections or filling with hyaluronic acid. However, these products lead to a greater local inflammatory reaction, loss of skin texture and location during the evolution besides absorption in the body producing inflammatory reactions in other organs. It was tried, for bone correction, bone autograft, acrylic prosthesis, hydroxyapatite and alloplastic implants.⁹ However, these materials can lead to infection or extrusion. Besides that, these techniques, many times, need more than one surgical time and can cause additional scars¹³

It was recently documented the good aesthetic result of RPS treatment with injections of poly-L-lactic acid (PLLA) after 18 months of follow-up of patients. PLLA is an alternative technique for patients that refuse surgical treatment and it is indicated only for the restoration of small tissue deficits. There was good restoration of facial volume, symmetrization of hemifaces, reduction of hyperpigmentation of the areas affected by atrophy and greater skin elasticity in the areas previously affected. Besides that, applications of PLLA do not require the use of anesthesia.¹³

Indeed, autologous fat transplant proved to be more effective and of lower cost. The long-term out-

come of patients, described in the medical literature, that underwent the use of autogenous tissue to correct deformities of the face is preferable to synthetic implant as they presented more natural texture, contour and facial expressions.¹⁴ It is important to highlight that autologous fat graft is not a definite treatment as the whole structure designed in the cosmetic surgery is lost with time due to gravity or by tissue reabsorption and the patient frequently needs a new intervention. According to the medical literature, absorption rates are variable, suggesting that fat survival depends not only of one factor but of several factors.¹⁵ Among them the site of collection and, mainly, the handling of fat tissue to be used as graft. In 1987, the American Society of Plastic and Reconstructive

Surgery reported its consensus about the theme, concluding that only 30% of the transferred adipose tissue remained viable after one year. They recommended overcorrection and stressed that unpredictable results were inherent to this procedure. Therefore, the follow up of a patient with PRS should be done indefinitely. It is also indicated treatment of psychological support for functional and neurological problems in patients with PRS.

It was concluded that the recent post-operative and assessment in 6 months showed good efficiency and high aesthetic satisfaction, measured by the patient's own judgement. Moreover, it is demonstrated that the cosmetic dermatological surgery can relieve serious damages to the anatomy of these patients affected by the Parry-Romberg syndrome. □

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