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Autologous Fat Transfer to Improve Aesthetic Appearance in Facial Asymmetry from Parry-Romberg Syndrome: A Case Report

ABSTRACT

Objective: To present a case of progressive hemifacial atrophy in a young woman with Parry-Romberg Syndrome and the role of autologous fat transfer to improve her aesthetic appearance and lessen facial asymmetry.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Result: A 20-year-old woman consulted because of drooping of the right eyelid and gradual thinning of right cheek muscles since age 16. On examination, the right facial muscles were hypoplastic with prominent facial bony ridges. An MRI scan showed atrophy of the right medial pterygoid and masseter. She underwent autologous fat transfer on the right side of the face to augment the cheek, improve cosmetic appearance and lessen facial asymmetry.

Conclusion: Our patient is satisfied and happy with the outcome and cosmetic appearance of her autologous fat transfer and is ready to undergo the same procedure if the need arises in the future. Although no definite cure exists for Parry-Romberg Syndrome, our report illustrates the role of autologous fat transfer as an inexpensive, easily harvested and biocompatible material to improve facial asymmetry. The procedure yielded encouraging results although long-term benefits remain uncertain.

Keywords: *Parry-Romberg syndrome; progressive hemifacial atrophy; autologous transplantation*

Facial asymmetry or hemifacial atrophy may result from a number of craniofacial syndromes, or develop as a result of trauma, pathology, or abnormal growth.¹ Parry-Romberg Syndrome is a rare, progressive but self-limiting degenerative disorder characterized by atrophy of the skin, subcutaneous tissue, and occasionally muscles, cartilage and bone, mainly on one half of the face.²

The exact etiology of this syndrome remains unclear but it may affect personality, psychosocial and aesthetic make up. The usual surgical approaches involve such complicated tissue transfers as a latissimus dorsi free flap.³ However, such procedures require resources that are unavailable and impractical in many low- and middle-income country settings such as ours and it becomes imperative to consider low-cost, minimally invasive alternatives.

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We present a case of progressive hemifacial atrophy in a young woman with Parry-Romberg Syndrome and discuss the role of autologous fat transfer to improve aesthetic appearance and lessen facial asymmetry.

CASE REPORT

A 20-year-old woman was admitted due to drooping of the right eyelid. Beginning at age 16, she noted gradual drooping of her right eyelid with no other symptoms such as headache or body weakness. Two years later, there was progressive thinning of muscles on the right cheek which eventually prompted consultation and subsequent admission. The maternal and family history were unremarkable with no history of previous illness and trauma.

Ophthalmologic evaluation showed normal visual acuity and pupillary size without diplopia. There was a significant difference in the palpebral apertures at 6mm O.D. and 10mm O.S. and levator function at 7mm O.D. and 14 mm O.S. with intact extraocular muscle function. She was assessed to have ptosis of the right eye and referred to our service.

Head and neck examination showed right-sided facial and forehead asymmetry from subcutaneous fatty tissue loss and prominent bony ridges on the right forehead, maxillary and mandibular regions. (Figure 1) There was slight deviation of the nasal septum to the right. Aside from a shallow right nasolabial fold from cheek muscle atrophy and right lip deviation, the oral cavity was normal with no trismus or tongue involvement. Her eyebrows and lips were not on the same horizontal plane. However, she could close her eyes against resistance and blow her cheeks symmetrically. There were no sensory deficits on both sides of the face and motor strength on manual muscle testing was intact. No other cranial nerve deficits were observed.

Cranial magnetic resonance imaging (MRI) revealed atrophy of right facial muscles (particularly the medial pterygoid and masseter) and incidental polysinusitis. No hemorrhage, mass or lesion was seen. (Figure 2) With an impression of Parry-Romberg Syndrome, the patient was observed for progression of atrophy over three months. There was no significant change in or progression of atrophy noted in baseline and three-month photographs. (Figure 3) She consented to autologous fat transfer to augment the cheek.

Under intravenous (I.V.) sedation using midazolam 5mg/ml, surgical landmarks were outlined on the patient’s face and donor site. (Figure 3 A,B) Fat graft harvesting was performed with local anesthesia infiltrated in the umbilical area (6 o’clock position) and a blade 11 incision to create the port site. (Figure 4A) Tumescent Solution (1L saline solution with 50cc lidocaine 1% + 1 ampule of epinephrine 1:1,000,000) was infiltrated into the donor site and allowed to set for 30 minutes before a 20-gauge infusion, 4mm cannula was inserted in the port site with

manual negative pressure using a large-bore needle attached to 50-mL syringe to harvest fat.

After harvesting, the aspirate was left to stand upright for 15 to 20 minutes to allow gravity to separate the liquid and solid portions into an upper oily layer of fatty acids and adipocyte cell mass and a lower mass of packed red blood cells. (Figure 4B) Excess plasma was drained.

A gauge-18 needle was used to make stab incisions in the recipient areas (right cheek and right infraauricular area) and fat was infiltrated with blunt tip 21-gauge cannulas into the subcutaneous layer. (Figure 5 A, B) The infiltrated amount per area was estimated based on the unaffected side, with emphasis on smooth convex facial contours to determine whether volume augmentation was sufficient, with over-correction to account for possible resorption. (Figure 6) The stab incisions were closed with single interrupted nylon 5-0 sutures. An abdominal binder was maintained for 7 days. The immediate post-operative and comparative pre- and three-day post-operative photos are shown in Figures 7 A-D and 8 A, B. She has remained well with minimal noticeable resorption of the fat graft at 3 months follow-up. (Figure 8 C)

DISCUSSION

Parry-Romberg Syndrome (PRS) is a rare disorder characterized by atrophy of skin, subcutaneous tissue and sometimes bone on the one side of the face.² The condition slowly progresses over 2 to 20 years



Figure 1. Preoperative photographs before autologous fat transfer procedure. Note the asymmetry in eyes, cheek and lip, **A**, frontal view; **B**, worm’s eye view revealing pronounced differences in cheek bulk; **C**, left lateral view showing the normal side, and **D**, right lateral view emphasizing bony prominences. Photos published in full with permission.

before stabilizing. It is typically restricted to one half of the face but occasionally involves the arm, trunk and leg. Neurological complications such as trigeminal neuralgia, migraine and seizures may be present² as well as cranial nerve dysfunction, fixed focal neurologic defects, hemiparesis and cognitive impairment.⁵ Cutaneous manifestations such as skin discoloration (hyperpigmentation or depigmentation) and cicatricial alopecia may be observed as well as mandibular and teeth involvement.² Our patient had no focal neurologic complaint, skin lesions or alopecia.

Ophthalmologic manifestations linked to PRS include enophthalmos, uveitis, retinal vasculitis, ipsilateral and contralateral third nerve paresis, glaucoma and eyelid atrophy.⁶ Eye manifestations

of our patient were limited to right ptosis and slight atrophy of the eyelid.

Parry-Romberg Syndrome was first reported by Parry in 1825 and later described as a syndrome by Romberg in 1846.⁵ Although there are many hypotheses for its pathogenesis, the etiology of PRS varies and remains unclear. Some proposed etiologies include heredity, autoimmune disorders, trauma, hypo- or hyperactivity of the sympathetic nervous system, disorders of the trigeminal nerve and infectious diseases from slow viruses such as herpes and Lyme disease.² Other associated infectious causes are otitis, dental infections, diphtheria, syphilis, rubella and tuberculosis.² In our case, hemifacial atrophy appeared spontaneously and developed gradually over four years with no apparent cause or precipitating event.

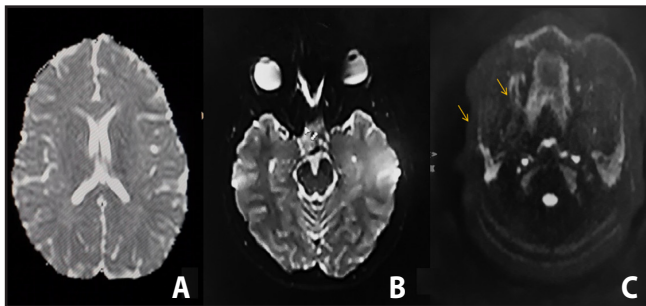


Figure 2. MRI showing **A.** normal brain; **B.** intact orbits and **C.** atrophy of right facial muscles, particularly the medial pterygoid and masseter (arrows), and incidental polysinusitis.



Figure 3. Surgical landmarks traced **A.** on the patient's face (recipient area); and **B.** the abdomen (donor site). Photos published in full with permission.

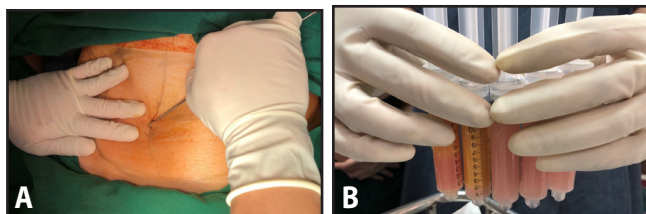


Figure 4A. Incision using blade 11 in the umbilical area to create a port site. (Photo published in full, with permission). and **B.** The aspirate was left upright for 15 to 20 minutes to allow gravity to separate the upper oily layer of fatty acids and adipocyte cell mass from the lower mass of packed red blood cells.



Figure 5. A, B. Recipient areas (right cheek and right infraauricular area) are infiltrated with fat in the subcutaneous layer using blunt tip cannulas through stab incisions. (Photos published in full, with permission).

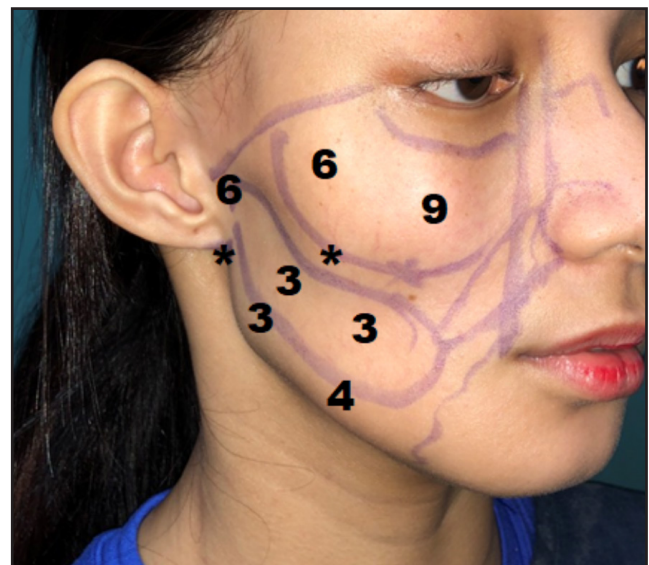


Figure 6. Distribution of fat injected per region in ml (*-injection site). Photos published in full, with permission.



Figure 7. A-D. The patient's appearance immediately after the autologous fat transfer procedure; compare with Figure 1 A-D. (Photos published in full, with permission).



Figure 8. Comparative pre-operative **A.** 3rd post-operative day **B.** and 3rd month follow-up **C.** photos (published in full, with permission).

Its incidence ranges from 0.3 to 2.5 cases per 100,000 population per year, most probably less than 3/100,000, and is more common in women, who represent more than 3 of 4 of the patients.⁵ Two cases of progressive hemifacial atrophy were reported in 1997 in the *Philippine Journal of Neurology*.⁷ Both involved women with a wide age gap and with different degrees of tissue and areas of involvement. Examinations and clinical investigations of both cases revealed no definite etiology.⁷

Parry-Romberg Syndrome is diagnosed based on clinical manifestations and MRI findings. It commonly affects the left side of the face in contrast to our present case which showed involvement of the opposite side.² In a study of brain MRIs of patients with Parry-

Romberg Syndrome, half were unremarkable while the remainder exhibited ipsilateral abnormalities including focal occipital and parietal region atrophy, and ipsilateral parietal and bilateral frontal white matter hyperintensities.⁸ Microhemorrhages, malformations, stenoses, and aneurysms were some of the vascular abnormalities reported in association with the syndrome.⁸ Progressive reduced contractility in the contralateral oculomotor nerve-innervated extraocular muscles was apparent in a Parry-Romberg Syndrome patient presented by Tama *et al.*, although there was no evidence of gross central nervous system disease.⁹ A small subarachnoid lesion of the oculomotor nerve may be detected using higher resolution, heavily T2-weighted technique.⁹ The MRI of the brain in our patient was unremarkable.

Parry-Romberg Syndrome deformities may be classified into 3 types based on the severity of soft tissue atrophy and bony involvement: mild, moderate and severe.¹⁰ Our patient may be classified as moderate, involving large areas of soft tissue atrophy and affecting the nasal ala and upper lip with deviation of the oral commissure and mild bony deficiency.¹⁰

No standard treatment algorithm currently exists for PRS. Topical and systemic corticosteroids, immunomodulators and plasmapheresis have been used with varying levels of success.⁵ Variable responses were demonstrated with antimalarials, antibiotics, vitamin D3 analogues and penicillamine.²

Surgery to restore a harmonious and symmetrical facial appearance after progression stabilizes within 2 to 20 years is the prevailing therapy.¹¹ For mild and moderate type of PRS, treatment focuses on soft tissue reconstruction (with autologous fat grafts, dermis grafts, dermal-fat flaps), injection of biomaterials (such as silicone or collagen), and free tissue transfers (omentum, rectus abdominis, latissimus dorsi).¹⁰ For severe types, microvascular-free flaps and autologous bone augmentation are needed.¹⁰ In a case series of 7 patients, a combination of autologous mandibular outer cortex (MOC) grafting with fat grafting was done in mild to moderate cases of PRS, and computer-assisted techniques was applied to improve surgical outcome with precision and accuracy.¹²

In clinical practice, autologous fat grafting is preferred and has been successfully performed with less morbidity.¹³ It has many of the characteristics of an ideal filler ("autologous, completely biocompatible, readily available in sufficient quantities, naturally integrated into host tissues, and removable if necessary")¹³ and has numerous applications.

Human adipose tissue also has the highest percentage of adult stem cells (ASC) of any tissue in the body, with as many as 5000 ASC per gram of fat compared with 100 to 1000 stem cells per milliliter of bone marrow.¹⁴ Rehman and colleagues reported that the regenerative potential of autologous adipose tissue was related to the presence



of multipotent mesenchymal stem cells which secrete multiple potentially synergistic proangiogenic growth factors: the adipokines.¹⁴ Further understanding and the ability to influence production of adipokines may lead to increased graft survival in hypovascular areas.¹⁴ To obtain a precise volume of soft-tissue augmentation and good aesthetic outcome, the lipofilling procedure may be repeated over multiple sessions¹⁵ as the resorption rate ranges from 30-70% within 1 year.¹⁶ A retrospective study of 13 patients with PRS twice treated with fat grafts revealed that the second fat graft could produce better cosmetic outcomes and volume retention without any sophisticated procedures.¹⁷ Additional procedures such as addition of autologous platelet rich plasma (PRP) have been proposed to address resorption. Platelet rich plasma (PRP) is a known natural reservoir of growth factors stimulating tissue repair and regeneration.¹⁸

Another option to improve our patient's appearance is correction of ptosis. The degree of ptosis and levator function are usually assessed to determine the appropriate surgical technique. Levator function may be classified as excellent (13 to 15 mm), good (8 to 12 mm), fair (5 to 7 mm), and poor (4 or less).¹⁹ The patient's levator function was fair (7 mm on the affected side) based on the classification. In patients with fair levator function, levator aponeurotic advancement is a possible technique that shortens the levator complex and produces excellent results.²⁰ We plan this as a future procedure in our patient.

Meanwhile, our patient is satisfied and happy with the outcome and cosmetic appearance of her autologous fat transfer and is ready to undergo the same procedure if the need arises in the future. Although no definite cure exists for Parry-Romberg Syndrome, our report illustrates the role of autologous fat transfer as an inexpensive, easily harvested and biocompatible material to improve facial asymmetry. The procedure yielded encouraging results, although long-term benefits remain uncertain.

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