A New Minimally Invasive Aesthetic Procedure for Correction of Frontal Coup de Sabre Deformity in Romberg's Syndrome

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Introduction

Parry–Romberg syndrome (PRS according to OMIM %141300) was first described by Parry (1825) and Romberg (1846) as a trophoneurosis facialis [1–3]. It is an uncommon poorly understood condition, characterised by slowly progressive atrophy of skin, subcutaneous tissue, fat and muscle. Involvement of bone and cartilage takes place only if it occurs prior to second decade of life, when the facial structures cranium are not fully developed [2]. The distribution is most commonly hemifacial, although bilateral cases have been reported. The sine qua non of the syndrome, the “Coup de Sabre,” describes a distinct line of soft-tissue thickness discrepancy, most commonly occurring along the supraorbital and infraorbital distributions of the trigeminal nerve [4]. Overlying skin may exhibit thinning and signs of hyper- or hypopigmentation [5]. Its onset is between 5 and 15 years of age. The progression of atrophy often lasts between 2 and 10 years, at which point the process tends to stabilize [6, 7].

Etiology is unknown. A cerebral disturbance in fat metabolism has been proposed as the primary cause [8, 9]. Trauma, infection, heredity, immunologic abnormality, trigeminal neuritis and scleroderma are the other suggested

causes [10]. The treatment is systematic and directed at reconstruction of the deficient tissue to improve the contour and symmetry of face. Described treatments include free tissue transfer, pedicled flaps, autologous tissue (fat, bone, cartilage) and biomaterials (silicone, alloderm, polyethylene) [11]. Among these techniques, fat grafting is the most commonly performed procedures, especially for mild to moderate asymmetry [7, 12–16].

Case Report

In 2012, a 19 year old female reported to the Department of Plastic and Reconstructive Surgery, KEM Hospital, with a chief complaint of facial asymmetry. Patient was apparently alright up to 6 years of age after which patient's mother noted slowly progressive facial asymmetry with no change in past 2 years. Physical examination revealed right hemifacial atrophy with mild deviation of mouth and nose to the affected site, slight enophthalmos and depression at the frontal region in midline with skin hyperpigmentation resulting in the classical ‘Coup de Sabre’ deformity (Fig. 1). Three dimensional CT of face revealed midline bony defect at the frontal region (Fig. 2). A clinical diagnosis of type 3 PRS was made based on history and physical findings [17].

Autologous fat transplant was planned and the areas to be infiltrated were marked preoperatively. Harvest was done by Coleman's technique from bilateral thighs and the amount injected over face is as shown in the figure (Fig. 3). Patient was followed up at regular intervals and serial photographs were taken (Fig. 4). At 6 months postoperation, a decision for second procedure was taken in view of reappearance of mild asymmetry, more pronounced at the frontal region. Fifth costal cartilage was harvested and

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carved to approximate size of the defect. With 1 cm incision placed within the hairline (Fig. 5), subperiosteal pouch was developed into which the graft was placed and adequately secured using pull through 4–0 prolene sutures.

Layered closure was achieved. At the remaining areas of mild discrepancy autologous fat grafting was done as described previously. Injection sites are illustrated in Fig. 6. Patient is on regular follow up with serial

Fig. 1 Initial appearance of the patient, front view

Fig. 3 Injection sites, stage 1

Fig. 2 Frontal defect on 3DCT scan

Fig. 4 Comparison of initial appearance with results after first procedure. *PREOP* Preoperative, *POD* postoperative day

Fig. 5 Placement of costal cartilage into subperiosteal tunnel

photographs at each visit (Fig. 7). At the time of writing, the patient had been in postoperative follow-up for 15 months after the second surgical procedure, with stable and satisfactory result (Fig. 8).

Discussion

The treatment of Romberg disease is both complex and challenging. In all cases, the aim of treatment is to restore facial symmetry and contours and reconstruct the atrophic bones and soft tissues. Most surgeons suggest that surgical intervention should be performed after cessation of

Fig. 6 Injection sites, stage 2

ongoing atrophic process, usually after a period of 1 year. Before, the surgical interventions are performed, it is crucial to evaluate the degree of atrophy. Two classifications have been proposed for the same [17, 18] (Tables 1, 2).

Of the various options available for restoration of facial volume in PRS, the use of autologous fat graft has dramatically increased with advances in fat grafting techniques. Face is an ideal recipient site with rich blood

Fig. 7 a–c Initial appearance of patient. d–f 1-Month follow-up after second surgery. g–i 8-Month follow-up after the second procedure. j–l 15 Months follow up after second procedure

supply which acts as an optimal scaffold, yielding a high percentage take of transplanted adipose cells [11]. An article by Mojallal et al. [19] examined the effects of human fat transfer in mice from a clinical and histologic level. The authors found that the transplanted fat stimulated a “regenerative effect” on the overlying skin resulting in increased pore diameter and overall elasticity. As subcutaneous fat is the tissue most commonly affected in hemifacial atrophy, it should ideally be replaced. The various methods of fat transfer include free fat grafting, dermal fat graft and structural fat grafting as described by Coleman. Of these, fat transfer by Coleman’s technique yields the most stable results.

The present case, classified as type 3 disease was, thus managed with autofatgrafting using Coleman’s technique. However, recurrence occurred gradually over a period of 5 months mainly at the region of frontal bone deficiency. So, the need for a more stable procedure using costal cartilage graft at the frontal defect was explained to the patient. In this

technique, a subperiosteal tunnel is developed by a small incision in the hairline. Costal cartilage carved to the defect size is placed in the tunnel and secured using 4–0 prolene sutures.

Other options for correction of bony deformity in PRS include alloplastic materials and free flaps. Alloplastic materials including liquid silicone have been tried with great enthusiasm [20–24]. The large volumes of silicone often required, however, have led to late complications which include chronic relapsing inflammation, skin ulceration, extrusion and sinus tracts [18, 25].

The emergence of microsurgical procedures has changed the treatment of a number of disorders, offering innovative techniques for facial reconstruction, with good long-term results as well as the improvement of morphologic and functional aspects [26]. However, their use can be restricted to severe type 3 and type 4 disease as it requires special microvascular armamentarium and surgical expertise.

Fig. 8 Postoperative CT scan (@15 months) **Table 1** Inigo et al. [18] classification of PRS

Inigo et al. [18]	
Mild	Atrophy of skin and subcutaneous tissue in the region of 1 branch of the trigeminal nerve, with no bone involvement
Moderate	Involvement of the skin and subcutaneous tissue in two branches of the trigeminal nerve, with no involvement of the orbital bone region or maxillomandibular growth
Severe	Atrophy of the skin in the three branches of the trigeminal nerve, with bone involvement

Plastic surgical literature has clearly demonstrated the success of cartilage grafts in rhinoplasty and otoplasty. Its use for restoration of PRS defect has also been reported in the literature [27]. Avelar et al. [28] used auricular cartilage in a similar fashion for chin deformity in a patient with

Table 2 Guerrerosantos et al. [17] classification of PRS

Guerrerosantos et al. [17]	
Type 1	Very mild depression, occurring in acute phase of Parry–Romberg syndrome
Type 2	Reduced thickness of the soft tissue, with no bone or cartilage involvement
Type 3	Soft tissue thinner than in type 2; initial bone and cartilage involvement
Type 4	The most severe type of facial depression; in many cases the skin is quite close to the bone and there is bone involvement

PRS. In the present case, we have used costal cartilage for the frontal defect by a technique which is never so elaborately explained in the past. The salient points of the technique described by us are as follows:

1. Use of costal cartilage graft as it is structurally better suited for a frontal bony defect where it can provide a good contour.
2. Hairline small incision and minimal subperiosteal dissection so that the graft could be immobilised without any sort of fixation.
3. Subperiosteal placement of the graft resulted in bone formation at the bone cartilage interphase as appreciated in the 15 month postoperation CT scan.

Conclusion

The use of costal cartilage graft for frontal Coup de Sabre defect in PRS is a minimally invasive, simple, reproducible technique with easy execution resulting in a minimal hidden scar in the hairline, giving a good forehead contour and a long term stable result in addition to autologous fat grafting.

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