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Treatment of “*En Coupe De Sabre*” Linear Scleroderma. Soft Tissue Augmentation and Regeneration by Autologous Fat Transfer

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ABSTRACT

Aim: Frontal linear scleroderma, also known as “en coup de sabre,” is a congenital deformity characterized by atrophy and furrowing of the skin of the front parietal area above the level of the eyebrows. In most cases it occurs as a single paramedian line that may be associated with hypoplasia of underlying structures and facial hemiatrophy. In case of a wide lesion many reconstructive strategies have been proposed. The modern approach is aimed at augmentation of the tissue deficiency by using liposuction and tissue regeneration. Autologous fat transfer (AFT) seems to be an effective strategy to restore the normal volume and contour of the face while providing a source of adipose-derived stem cells (ADSCs) with a multilineage differentiation potential. **Methods:** In this report, we present a rare case of linear scleroderma en coupe de sabre which was successfully managed with three stages of autologous fat grafting. **Conclusion:** The proposed uses for ADSCs in tissue repair and regeneration are quite impressive. Recent works on ADSCs would suggest that adult cells may prove to be an equally powerful regenerative tool in treating congenital and acquired maxillofacial disorders. More importantly, physicians, researchers and international associations need to cooperate in informing clinicians about what practices are based on evidence and to encourage support of additional research. There is increasing interest in a possible therapeutic effect of ADSCs from processed lipoaspirate for a wide spectrum of clinical applications in the facial and craniofacial area. AFT can be used in any facial area where soft tissue is lacking or where there is scarring, producing natural and long-lasting results. Mesenchymal stem cells represent a great tool in regenerative medicine. However, more definitive studies are needed to answer specific questions regarding the best technique to be used and the role of ADSCs. Autologous fat grafting provides a safe and easy approach for the treatment of linear scleroderma en coup de sabre, long-term clinically satisfactory results can be obtained.

Keywords: Autologous fat transfer, frontal linear scleroderma, congenital craniofacial malformations, reconstructive surgery, rare autoimmune diseases, tissue regeneration, linear scleroderma, en coup de sabre

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INTRODUCTION

Scleroderma is a chronic inflammatory disease of the connective tissue distinguished by functional and structural alterations in the small blood vessels and by changes related to the accumulation of collagen in the tissues [1,2]. Based on several clinical features, scleroderma may be classified into the following subtypes: localized scleroderma, systemic sclerosis, overlap syndrome, and similar forms of scleroderma induced by toxic agents or pharmaceutical drugs. Localized scleroderma is characterized by skin fibrosis in the form of plaques or macules. Lesions are rather common on the arms or legs. The prevalence is estimated to be approximately 1 to 9 per 100,000 and is more common in women (female-male ratio about 4:1), and occurs more frequently in children than in adults. Localized scleroderma has an unknown cause. It is widely thought to be an autoimmune reaction that induces increased collagen production. Other origins of the disease have been suggested, including genetic and infective factors. Diagnosis is based on disease aspects and on clinical examination. Localized scleroderma is classified in different subtypes, which include morphea, generalized morphea, and linear scleroderma. However, more than one form can coexist in the same patient [3,4,5]. Linear scleroderma may develop into severe asymmetry of the face or limbs, leading to contracture in flexion and disability. Research in the field of reconstructive surgery and regenerative medicine has a common goal of finding the ideal technique to correct this acquired or congenital deformity of the soft tissues by means of autologous grafting material (autologous fat transfer). Therefore, transplanted adipose tissue should be considered not only as a filling, but also as a means to improve quality of the recipient site. More recently, the development of a new concept has led to the possibility of a suitable technique with established indications and predictable results. Indeed, due to the research and experience provided by Coleman, [6,7,8], who

perfected a procedure using adipose tissue in 1997, the technique of structural fat grafting has been reevaluated. The benefit of adipose tissue is its biocompatibility - which easily adapts to the growth of the face over time - and its lower morbidity compared with many other alloplastic materials that frequently lead to the typical reactions caused by foreign bodies [9,10,11,12,23]. Today AFT is ideal for the correction of localized tissue atrophy, radiodermatitis loss of post-traumatic sequelae, post-tumor resection, craniofacial deformities, effects of radiotherapy, and burns¹⁴. Recent studies have shown that adipose tissue with its angiogenic and antiapoptotic features contains the highest percentage of stem cells compared with other body tissues^[15,16,17]. The percentage of stem cells is distinctly higher than of those in bone marrow. The use of these cells does not create such debatable ethical issues as do embryonic stem cells. Therefore, transplanted adipose tissue should be considered not only as a means of filling, but also as a means to enhance the quality of implanted tissue whether it is for reasons of aging, scarring, loss of substance, radiotherapy, or any other injurious event sequelae. Grafting of adipose tissue is an excellent tool for craniomaxillofacial surgery as it accomplishes natural results that stand the test of time. AFT is not simply a filler in that it regenerates, creating new vascularization and real structural alterations. It is still unclear how the adipose tissue brings about these alterations, but research in this continues developing the fat grafting technique. This technique should become the building block of every maxillofacial surgeon's armamentarium in the field of reconstructive surgery.

MATERIALS AND METHODS

The case of a 34-year-old woman affected by "en coup de sabre" of the right fronto-orbital region is reported. Facial examination revealed deep furrowing of the right fronto-orbital region and marked asymmetry of the arch of the eyebrow from both the frontal aspect and from a sub-menton vertex view. The patient underwent

facial reconstruction in three different stages. In the first stage, surgical step fat grafting involved harvesting of 60 mL of adipose tissue, and after centrifugation, 39 mL of concentrated adipocytes were injected. In the second and third surgical steps, 30 and 40 mL of concentrated autologous adipocytes, obtained from

centrifuged fat, were injected into the right fronto-orbital zygomatic region, respectively. Two years later the patient was satisfied with noticeable improvement in the fronto-orbital symmetry and morphology, as well as enhancement of soft tissue thickness and texture (Figure 1 A,B,C,D,E,F).

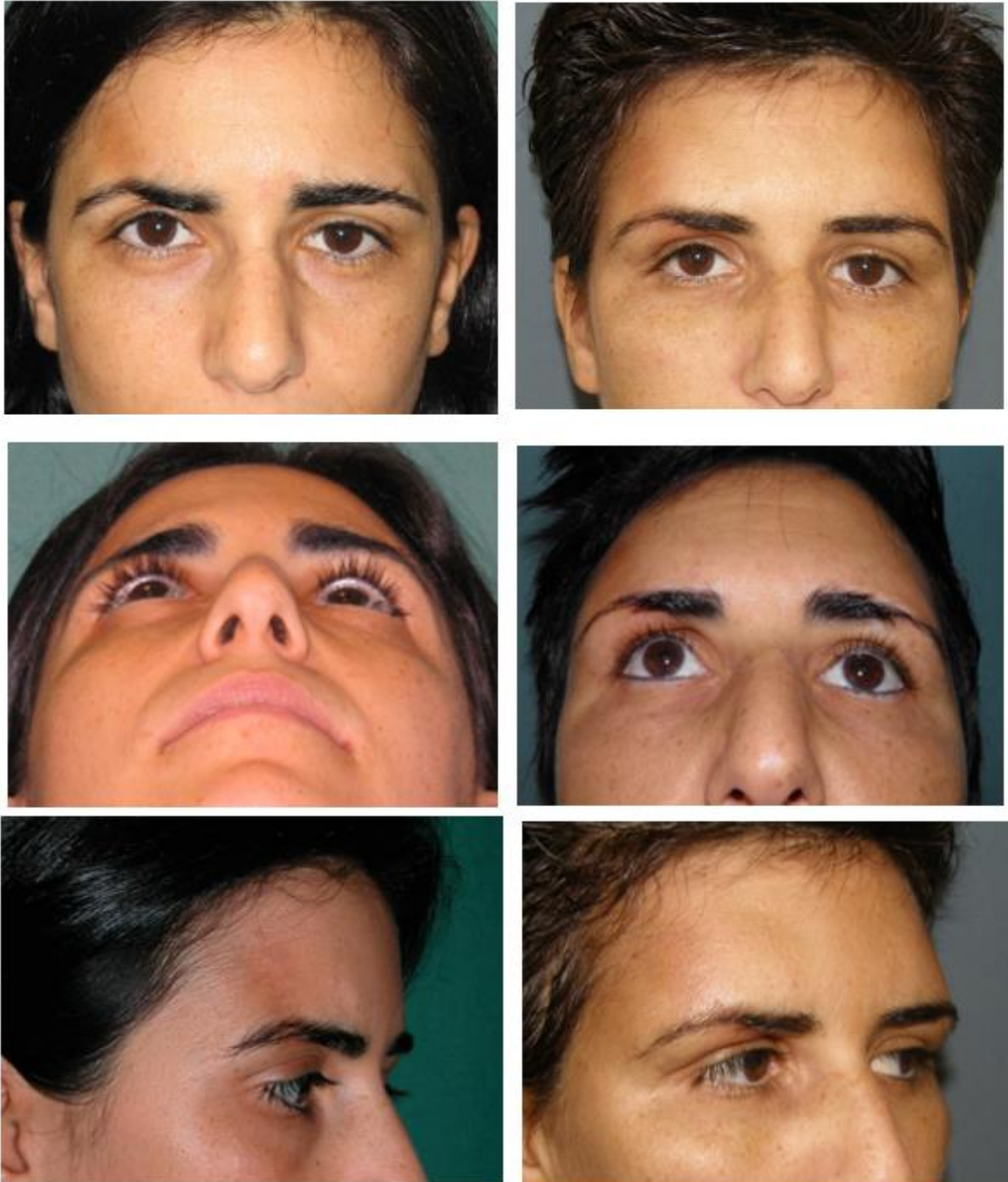


FIGURE 1 A,B,C,D,E,F (A)Preoperative frontal view of a right linear scleroderma .(B)Reconstruction with autologous fat transfer:result after two years. AFT improved contour reshaping, volumes and tissue texture(B)(C)Preoperative Submental view (D) Postoperative submental view after three surgeries (E)Lateral view revealing the right facial asymmetry . (F) Postoperatively:there is visible improvement in contour along the central forehead after fat grafting

RESULTS AND DISCUSSION

AFT is biocompatible and easily adapts to the growth of the face over time, with lower morbidity compared with many other alloplastic materials, which may frequently trigger adverse reactions to foreign bodies. Fat is the ideal autologous tissue for the correction of localized atrophic tissue, radiodermatitis loss of post-traumatic sequelae, post-tumor resection, craniofacial deformities, radiotherapy, and burn complications. Recent studies have shown that human adipose tissue is a rich source of stem cells, exhibiting multilineage potential and featuring secretion of angiogenic and antiapoptotic factors in animals. The studies have demonstrated the therapeutic efficacy of adipose stem cells for many pathologies and for the treatment of radiation-induced damage through a process of replacing damaged injured tissue. In conclusion, AFT introduced as a way of improving facial esthetics, has evolved in recent years into more complex reconstructive procedures. Today, tissue engineering together with regenerative medicine is a multidisciplinary science that is evolving along with biotechnological advances. The proposed uses for ADSCs in tissue repair/regeneration are quite impressive. Recent research on ADSCs would suggest that this adult stem cells may prove to be an equally powerful tool in treating congenital and acquired disorders. However, the availability and the processes for obtaining stem cells are still a challenge for both surgeon and scientist pursuing regenerative medicine and tissue engineering^[18,19,20]. Fat grafting in scleroderma patients likely improves skin manifestations by recreating fullness and correction of contour deformities. The injected fat provides a mixture of cells that influences the recipient site, resulting in improved outcomes. However, the availability and processes for obtaining stem cells still remain a challenge for both the surgeon and the scientist pursuing regenerative medicine. Further research from the bench to the bedside is needed.

DECLARATIONS

Authors' Contributions: Luigi Clauser and Antonio Lucchi conceived and designed the clinical data and research; Luigi Clauser and Antonio Lucchi performed surgical treatments and samples collection; Andrea Edoardo Bianchi, Carolina Sannino and Luigi Clauser prepared the article.

Availability of data and materials

Not applicable

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Conflicts of interest

All Authors declared that there are no conflicts of interest

Ethical approval and consent to participate

Not applicable

Consent for publication

Written informed consent was obtained for all patient images.

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