Regenerative Approach to Scleroderma with Fat Grafting

Guy Magalon, MD[a,b,*], Aurélie Daumas, MD[c], Nolwenn Sautereau, MD[c], Jérémy Magalon, PharmD[b], Florence Sabatier, PharmD, PhD[b,d], Brigitte Granel, MD[c,d]

KEYWORDS

• Systemic sclerosis • Cell therapy • Microfat injection • Stromal vascular fraction • Fat grafting • Autologous fat graft • Adipose tissue

KEY POINTS

- Systemic sclerosis (SSc) is a rare autoimmune disease characterized by skin fibrosis, microvascular damage, and organ dysfunction.
- Facial manifestations in SSc are disfiguring and lead to social disability with psychological distress.
- Hand involvement in SSc can lead to a severe disability, with no effective therapy.
- Adipose-tissue-derived stem cell therapy has emerged as a therapeutic alternative for regeneration and repair of damaged tissues.
- Patients with SSc can benefit from fat grafting: microfat injection in the face to improve skin pliability and quality with esthetic benefit, and injection of the autologous adipose-tissue-derived stromal vascular fraction (ADSVF) in fingers for a trophic effect.

Three surgical technique videos accompany this article showing the authors approach to the treatment of the face and of the hands in systemic sclerosis patients, and the inside of 2-mm, 14-gauge cannula harvesting and microfat injection with 0.8-mm, 21-gauge cannula at http://www.plasticsurgery.theclinics.com/

INTRODUCTION

SSc (scleroderma) is a chronic systemic autoimmune disease characterized by microvascular abnormalities and progressive skin and internal organ fibrosis.[1] Life-threatening organ lesions leading to pulmonary arterial hypertension, pulmonary fibrosis, and scleroderma renal crisis only affect a minority of patients. By contrast, lesions of the hands and face are almost always present. Although not life-threatening, these manifestations are very obvious, hard to conceal, and lead to disability and worsening quality of life.[2–4] Facial symptoms are associated with cosmetic

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[a] Plastic Surgery Department, Assistance Publique Hôpitaux de Marseilles (AP-HM), Aix-Marseilles University, 13005 Marseilles, France; [b] Culture and Cell Therapy Laboratory, INSERM CBT-1409, Assistance Publique Hôpitaux de Marseilles (AP-HM), Aix-Marseilles University, 13005 Marseilles, France; [c] Internal Medicine Department, Assistance Publique Hôpitaux de Marseilles (AP-HM), Aix-Marseilles University, 13010 Marseilles, France; [d] Vascular Research Center Marseilles, INSERM UMRS-1076, Aix-Marseilles University, 13005 Marseilles, France
* Corresponding author. Conception Hospital, Assistance Publique Hôpitaux de Marseilles (AP-HM), 13005 Marseilles, France.
E-mail address: secretariat.magalon@gmail.com

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disfigurement and limited expression with mask-like stiffness of the face. Lesions in the hand lead to substantial difficulty in performing everyday tasks (such as dressing, eating, and applying makeup) as well as an increased risk of chronic digital ulcers (DUs). Therapeutic interventions in this disease are mainly based on the use of vasodilators. No antifibrotic treatment has proven effective. Unlike other autoimmune diseases, immunosuppressive drugs have a limited clinical interest.[5–10] Thus, functional improvement of hand motion and face appearance represent a real challenge for physicians and a priority for patients who often feel that this aspect of their disease is neglected.

Use of adipose tissue as filling product in plastic and esthetic surgery is an ancient technique. Significant renewal of interest in this approach for the restoration of all volume defects was observed after the description of the LipoStructure® technique by Coleman.[11–13] Recently, identification and characterization of the ADSVF, a population that includes mesenchymal-like stem cells, endothelial progenitor cells, and hematopoietic cells, have revolutionized the science showing that adipose tissue is a valuable source of cells with multipotency as well as angiogenic and immunomodulatory properties that facilitate tissue repair. The ease of harvest by liposuction and the abundance of these cells (by comparison to bone marrow) avoid the need for ex vivo expansion before clinical use. Because of these practical factors and the stromal vascular fraction's ability to differentiate and secrete immunomodulatory, angiogenic, antiapoptotic, and hematopoietic factors, use of adipose tissue is becoming more attractive and is expanding in regenerative medicine.[14–19]

In this article, the authors present their clinical approach using adipose tissue in the treatment of the face and hands of patients with SSc.

PATHOLOGY OF SCLERODERMA

Face

Involvement of the face with associated oral complications, esthetic changes, and impairment of the patient's self-image is found in over 90% of patients with SSc.[3,20,21]

Fig. 1 and **Table 1** illustrate the main orofacial findings in patients with SSc.

Several validated tools have been developed for assessing the involvement of the face. Skin involvement is usually assessed by the Rodnan skin score. This semiquantitative score rates the severity of skin sclerosis from 0 (normal) to 3 (most severe). Xerostomia can be easily measured by sugar test (time to melt a sugar on the tongue, without crunching it) and with the xerostomia inventory index. Mouth opening is assessed in centimeters by measuring the distance between the tips of upper and lower incisive teeth. Elastosonography and three-dimensional photographs can also be used. Mouth-related disability can be assessed by the Mouth Handicap in Systemic Sclerosis (MHISS) scale, which is the first mouth-specific disability outcome measure designed for patients with SSc.[3] This scale evaluates 3 factors: reduced mouth opening, sicca syndrome, and esthetic concerns. Although mouth disability seems to have less weight than hand disability in total disability, the MHISS score explained up to 36% of the variance of the Health Assessment Questionnaire score. This fact highlights the need to specifically assess disability involving the mouth in patients with SSc. Rehabilitation and management of the face is mainly based on physiotherapy with mimic exercises, massage, and self-administered home-based exercises. Mouth and dental care are not specific.

Some case reports have shown the efficacy of autologous fat grafting in the treatment of linear scleroderma.[22,23] Besides the volumizing effect of mechanical lipofilling, autologous fat grafting also seems to produce trophic and angiogenic effects. The use of autologous grafting of adipose tissue seems to have substantial potential to correct signs of face involvement in SSc.

Hands

Involvement of the hand is common in patients with SSc and represents a large burden in work and daily activities. Hand disability has a multifactorial origin with microvascular lesions, skin sclerosis, tendon retraction, bone and articular involvement, and subcutaneous calcinosis.[4,24–31] Each of these lesions causes pain, functional impairment, esthetic issues, and psychological distress.

Vascular involvement

Vascular dysfunction including Raynaud's phenomenon (paroxysmal vasospasm) (**Fig. 2**), acrocyanosis (permanent ischemia), and subsequently DUs with their potential complications (infections, digital necrosis, autoamputation) are the main manifestations. Raynaud's phenomenon occurs in almost all (95%) patients with SSc. DUs, defined as necrotic lesions that occur either at the pulp of the digits (ischemic DUs) or over bony prominences (mechanical DUs), occur in up to 50% of patients with limited or diffuse SSc. DUs typically occur early in the course of SSc. A study assessing functional limitations owing to