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CASE REPORT

Acquired Partial Lipodystrophy (Barraquer-Simons Syndrome): Early Cosmetic Intervention with Autologous Fat

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Barraquer-Simons syndrome is a rare acquired lipodystrophy characterized by gradually symmetric subcutaneous fat loss in a craniocaudal distribution, often associated with hypocomplementemia and nephropathies. Facial cosmetic treatment in this disorder has not been fully described in the literature. We present a patient with Barraquer-Simons syndrome with emphasis on early cosmetic intervention with autologous fat grafting and its long-term efficacy. At the follow-up 37 months after the last fat grafting, preservation of the grafted fat was noted while lipodystrophy progressed in the trunk regions. Autologous fat grafting is suggested for the correction of facial dysmorphism in this type of lipodystrophy.
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-Keywords-
Autografts, Lipectomy, Lipodystrophy, Subcutaneous fat

INTRODUCTION

Lipodystrophies comprise a group of abnormal or degenerative conditions affecting adipose tissue, which are characterized by selective fat loss, ranging from partial to generalize[1]. Barraquer-Simons syndrome (BSS), also known as acquired partial lipodystrophy, is a rare disorder characterized by a gradual loss of subcutaneous fat starting in the face and upper half of the body progressing downward toward the gluteal line[2]. Patients with BSS usually need facial cosmetic interventions because they develop progeroid looks which impair patient's quality of life[1,2]. Until now, long-term efficacy of autologous fat grafting has not been fully studied in this rare form of lipodystrophy.

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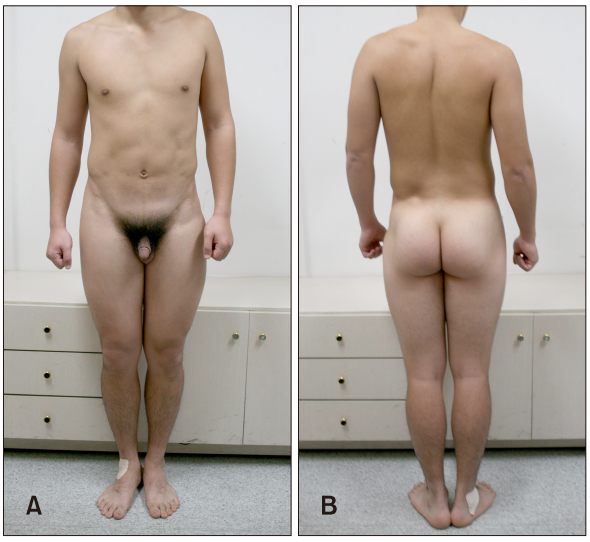

Fig. 1. (A) Markedly decreased subcutaneous fat in the abdomen, which accentuates the contour of muscles. (B) Fat depositions in the gluteal and thigh regions.

CASE REPORT

A 22-year old Korean male patient was referred to the dermatology department for restoration of facial fat loss. He noted that his subcutaneous fat volume gradually began to decrease in the cheeks and the abdomen since adolescence. Past medical history revealed no abnormality, and he had no history of medications that could cause lipodystrophy. There was no family history of the similar condition. We received the patient's consent form about publishing all photographic materials. A physical examination revealed multiple foci of lipoatrophy (Fig. 1, 2), with loss of facial and abdominal subcutaneous fat. The subcutaneous fat was preserved in other anatomic regions, particularly in the buttocks and the thighs, resulting in a disproportion between the upper and lower parts of the body. Thyroid was normal upon palpation. Hepatosplenomegaly and umbilical hernia were absent. Other signs suggestive of endocrinologic abnormalities, including acanthosis nigricans and acromegaly, were not found. Neurologic and ophthalmologic examinations revealed no abnormal findings. Laboratory data was all within normal limits except for a decreased level of C3 complement (32 mg/ml; normal: 88～252 mg/dl in males) and the presence of C3 nephritic factor. The patient presented no systemic manifestations at the time of the evaluation. Computed tomography images demonstrated near complete absence of subcutaneous fat along the chest and abdomen, whereas fat was well-preserved in the gluteal and lower extremity regions (Fig. 3). Intraabdominal fat was spared. Scan images of the face showed a significant reduction of both superficial and deep fat volumes in the midface, more predominant in the left side. Skin biopsy from the atrophic skin of the abdomen showed an absence of subdermal adipose tissue.

Autologous fat transplantation was performed for volume restoration of the patient's face. We infiltrated the skin with a tumescent Klein solution in the gluteal and posterior thigh regions and continued with fat extraction using a 3-mm blunt cannula connected to a 10-ml syringe. After centrifugation, concentrated fat was injected in the mid- and lower face using 1-ml syringes connected to an 18-gauge blunt cannula; 22 ml of the concentrated fat was injected in midfacial deep fat compartments and 16 ml was injected in superficial fat compartments (nasolabial, malar, and jowl fat pads). Two additional injection sessions were held with 3-month intervals. At the long-term follow-up 3.7 years after the treatment, the facial volume was well-preserved with no signs of significant graft absorption (Fig. 2) whereas lipodystrophy further progressed in the trunk region.

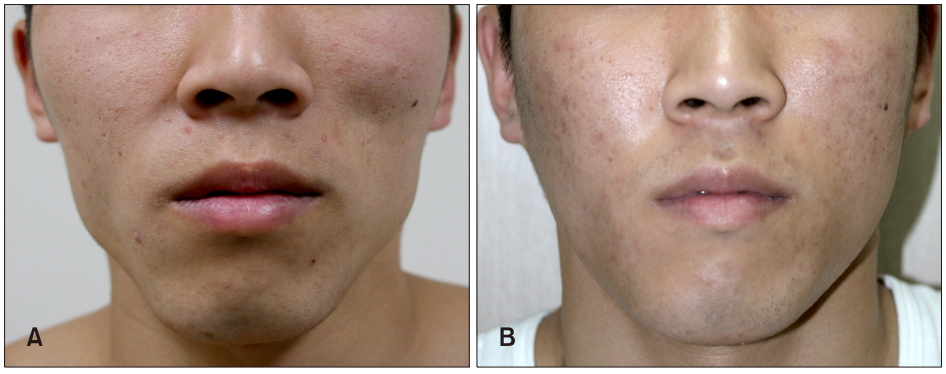

Fig. 2. (A) A preoperative photograph of the face demonstrating a significant fat loss mainly on cheeks, creating an old and wasted look. (B) A follow-up photograph 3.7 years after three fat grafting sessions showing stable results in the mid- and lower face injections.

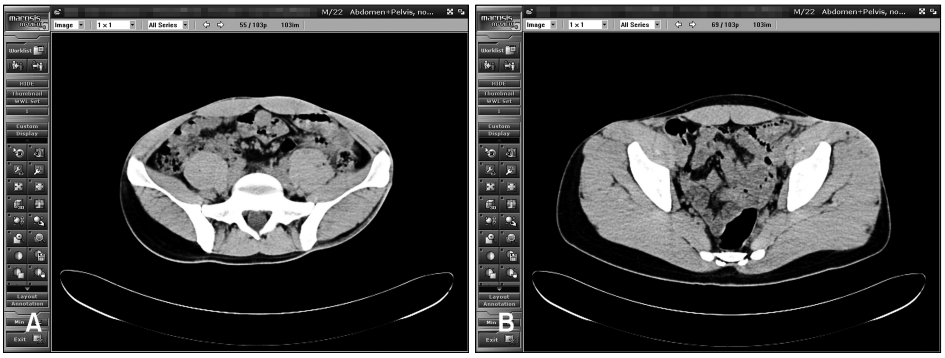

Fig. 3. Computed tomography images of the abdomen and pelvis demonstrate a fat distribution characteristic of acquired partial lipodystrophy (Barraquer-Simons syndrome). Near complete absence of subcutaneous fat in the abdomen (A), in contrast to the prominence of gluteal fat at the level of the pelvis (B).

Table 1. Clinical differential diagnosis of lipodystrophy syndromes

Type	Congenital generalized lipodystrophy	Familial partial lipodystrophy	Acquired generalized lipodystrophy	Acquired partial lipodystrophy
Age range of onset	Infancy to early childhood	Childhood through adulthood	Childhood through adulthood	Childhood through adulthood
Male:Female ratio	1:1～2	1:1～2	1:3	1:4
Sites of involvement				
Face and neck	+		+	+
Chest/trunk	+	+/-	+	+
Upper extremities	+	+	+	+
Lower extremities	+	+	+	
Intra-abdominal	+		+/-	
Sites of fat accumulation/sparing				
Face and neck		+/-		
Hips and buttocks				+/-
Lower extremities				+/-
Intra-abdominal		+/-	+/-	+/-

+: present, +/-: absent.

DISCUSSION

Classification of lipodystrophies includes three main groups: generalized, partial (extensive, but not generalized), and localized (limited to isolated areas)[3]. Acquired lipodystrophies are more common than inherited varieties[2]. Acquired partial lipodystrophy is characterized by the gradual onset of bilaterally symmetrical loss of subcutaneous fat starting from the face, spreading towards the trunk[2]. In contrast, the adipose stores of the gluteal regions and lower extremities tend to be either preserved or are increased[1]. The exact pathogenesis of BSS remains unclear. Lipodystrophy is often associated with glomerulonephritis, low C3 serum complement levels, and the presence of a C3 nephritic factor. It has been hypothesized that C3 nephritic factor induces the lysis of adipocytes that express factor D (a serine protease enzyme also referred to as adipsin), and differential expression of factor D by various tissues in the body dictates the cephalocaudal pattern of fat loss characteristic of BSS[3].

Our patient met the following diagnostic criteria of BSS: gradual onset of subcutaneous fat loss starting from the face to the abdomen but sparing the lower extremities (essential criterion); onset of subcutaneous fat loss during childhood and adolescence; absence of family history of lipodystrophy; characteristic body fat distribution documented by imaging studies; low serum levels of C3 with or without related nephritis; presence of autoimmune diseases (supportive clinical criteria)[2]. The key clinical findings in BSS and other major lipodystrophy subtypes are summarized in Table 1.

Therapeutic approaches for BSS consist of improving esthetic appearance and the management of additional systemic disorders[1]. It is noteworthy that in the present case, grafted fat harvested from the gluteal and posterior thigh regions remained stable in the injected areas of the face. While it is hard to explain why fat grafting was effective in spite of the remaining low serum level of C3, the present case raises the possibility that the grafted adipocytes could retain properties of the original donor site which is resistant to the disease progression. When considering timing of fat augmentation in lipodystrophy patients, evidence shows that patient age effects the survival of fat grafts, i.e., unfavorable growth kinetics and differentiation capacity of adipose-derived stem cells are related with increasing donor age[4]. Based on our observation, we suggest an early facial cosmetic intervention by autologous fat grafting in patients with BSS. We also suggest further investigations on adjuvant use of platelet-rich plasma or adipose-derived stem cells and their stromal vascular fractions to improve the efficacy and stability of fat grafting in this group of patients.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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