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Research Paper

Case report of Barraquer-Simons Syndrome

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Abstract
Barraquer-Simons Syndrome (BSS), also known as Progressive partial lipodystrophy, is a rare acquired condition characterized by the progressive loss of subcutaneous fat from the upper body while sparing or even hypertrophying the lower body fat. Here, we present a case of a 27-year-old woman, diagnosed with BSS, highlighting the clinical presentation, diagnostic process, and treatment approaches.

Keywords: *Barraquer-Simons Syndrome, Progressive Partial Lipodystrophy, Subcutaneous Fat Loss, Autoimmune Diseases, Facial Reconstruction*

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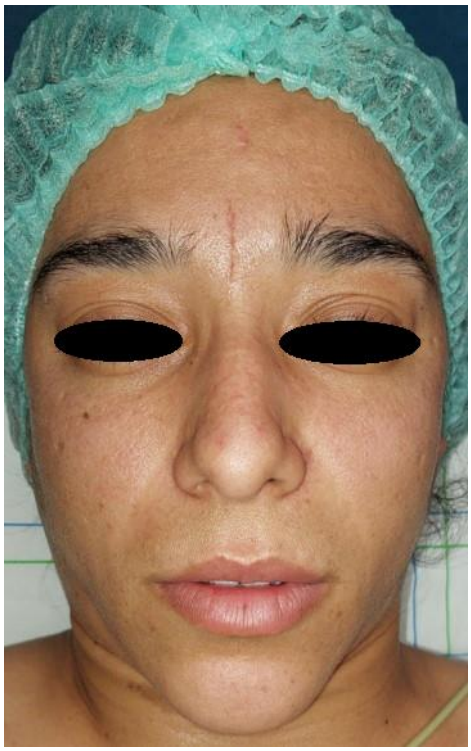
I. Introduction

Barraquer-Simons Syndrome (BSS) is an uncommon disorder first described by Barraquer in 1906. It predominantly affects females and typically manifests in childhood or early adolescence. The condition is characterized by the selective loss of subcutaneous fat in the face, neck, upper trunk, and arms, contrasted by normal or increased fat deposition in the lower body. The exact etiology remains unknown, but immune system dysregulation plays a significant role. [1]

II. Case Presentation

A 27 year-old female presented to our department with a noticeable asymmetry in fat distribution. The patient's medical history was unremarkable until she noted progressive thinning of her face over the past two years, without significant weight loss or other systemic symptoms. **FIGURE 1**



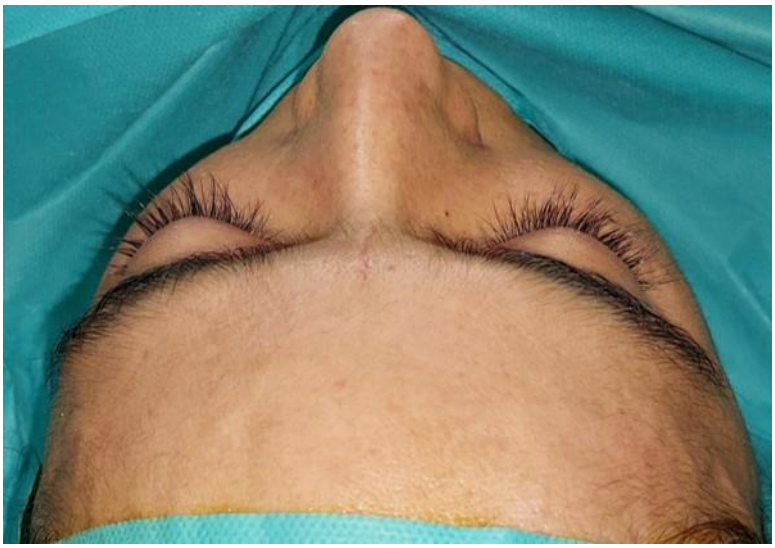


FIGURE 1,2 : preoperative photo of the patient

1. Clinical Examination

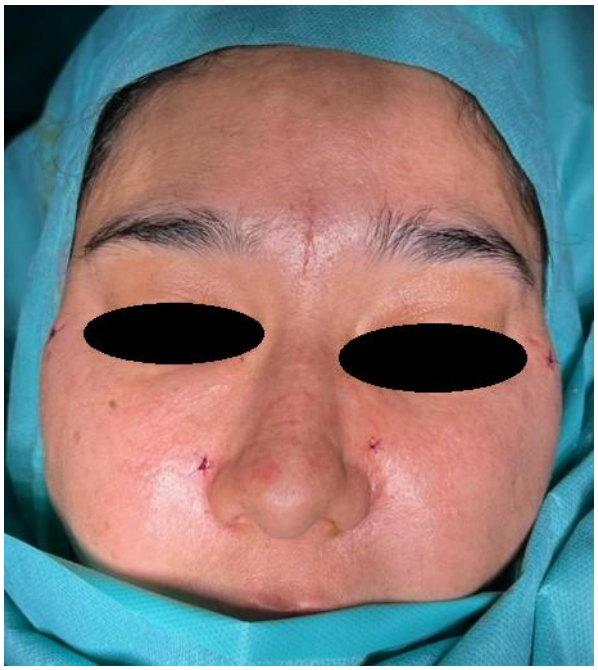
Physical examination revealed marked lipoatrophy in the face. The lower half of her body, particularly the hips, buttocks, and thighs, appeared mildly hypertrophic. No other abnormalities were noted on systemic examination.

2. Management and Treatment

Given the aesthetic and psychological impact of the condition, the patient opted for facial reconstruction. The chosen method was autologous fat grafting using Coleman's technique under general anesthesia, which involves harvesting fat from the patient's lower body and injecting it into the areas of lipoatrophy.

The patient underwent two sessions of fat grafting with a total of 200 mL of fat transferred at the level of the nasolabial fold, temporal, lower jugal, malar and lower temporal with a fat injection quantity of 200 ml at the last session . These injections are subcutaneous, intramuscular and supraperiosteal .

Post-procedure, she experienced significant improvement in facial contour and symmetry. Follow-up at six months showed stable results with minimal resorption of the grafted fat. ***FIGURE 4-5***



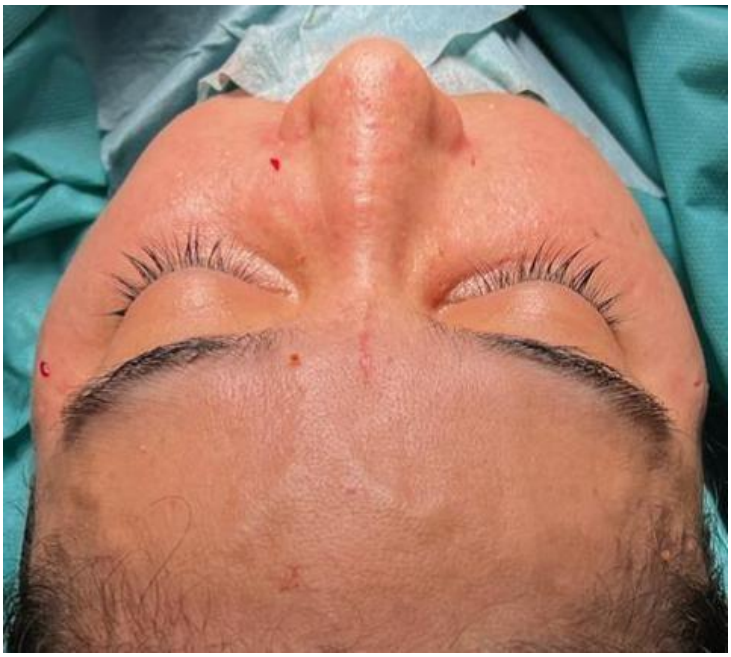


FIGURE 3,4 : intraoperative photo after injection of autologous fat

III. Discussion

Barraquer–Simons syndrome was reported initially by Mitchell [1885] and later by Barraquer [1907] and Simons [1911]. **[2]** The pathogenesis and genetic background is obscure .The onset of Barraquer–Simons syndrome in most patients is before the age of 15 years. Fat loss usually occurs over 18 months, however, it can be periodic during several years.The diagnosis of BSS was based on the clinical presentation of selective fat loss and the presence of C3NEF **[1].** Our patient did not exhibit symptoms of associated autoimmune diseases.The underlying pathophysiology of BSS involves immune system dysregulation, particularly the alternative complement pathway, which leads to the selective destruction of adipose tissue in the upper body. This patient's presentation and laboratory findings align with the typical features of BSS. [3]

There is no specific treatment. Therapeutic approaches consist of improving esthetic appearance with plastic surgery and the management of additional systemic disorders.[4] The main goal of cosmetic surgical procedures is to minimize the psychological discomfort that impairs the patient’s quality of life.More invasive

techniques have been proposed such as free TRAM type flaps , or flaps thigh [5]. Injections of heterologous products such that L-polylactic acid seems to be a good alternative to lipostructure (rapid technique under anesthesia local). The high cost, instability over time and the heterologous nature of the product limits their use . Treatment is symptomatic based on facial filling techniques [6] including Coleman adipocyte autograft technique , . This technique is simple, effective but requires multiple sessions [7]

BSS can be associated with several autoimmune diseases, , including systemic lupus erythematosus and juvenile dermatomyositis and diabetes... These possible associations must be researched by a systematic general examination.[8]Barraquer–Simons syndrome is a slowly progressive disease and if there is no associated renal impairment the prognosis is excellent [10]

IV. Conclusion

Barraquer-Simons Syndrome is a rare disorder with significant aesthetic and psychological implications. Early diagnosis and a multidisciplinary approach, including immunological monitoring and reconstructive options, are crucial for managing the condition effectively. In this case, autologous fat grafting proved to be a successful intervention, improving the patient's quality of life and self-esteem. Further research is needed to understand the underlying mechanisms and develop targeted therapies for BSS.

Declaration of Interests

The authors declare no conflicts of interest related to this case report.

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[10]. Syndrome de Barraquer-Simons Barraquer-Simons syndrome N. Mansouri Hattab, *, S. Lahmiti , S. Aimadeddine , S. Fawzi , M. El Bouihi , T. Fikry [6]