ACTAS
Dermo-Sifiliográficas

Full English text available at
www.actasdermo.org

IMAGES IN DERMATOLOGY

Barraquer-Simmonds Syndrome

Síndrome de Barraquer-Simmonds

E. Rozas-Muñoz*, N. Ramos-Arancibia

Servicio de Dermatología, Hospital de San Pablo, Coquimbo, Chile

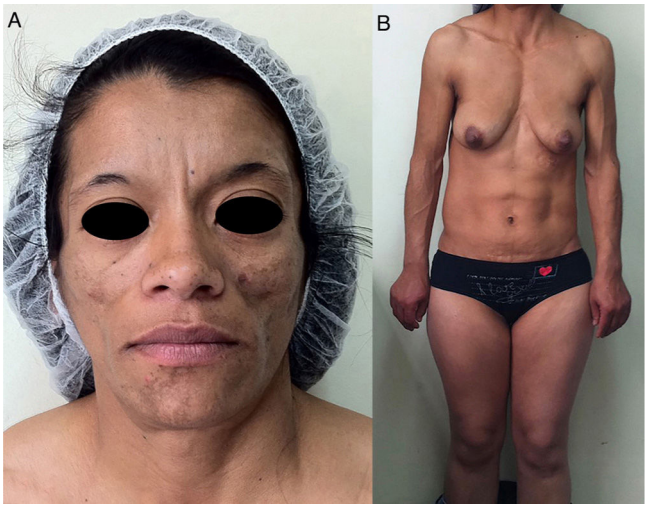

Figure 1

A 33-year-old woman presented with progressive cephalocaudal loss of subcutaneous fat that began at age eight. Physical examination revealed decreased subcutaneous adipose tissue limited to the face, with loss of temporal fat pads and prominent zygomatic arches, trunk and upper extremities, resulting in a muscular appearance (Fig. 1a and 1b). She was not taking any medications. Histopathological examination showed focal loss of subcutaneous tissue without inflammatory infiltrate. Testing for human immunodeficiency virus infection was negative, and the results of laboratory investigations including complete metabolic screen were unremarkable. A diagnosis of Barraquer-Simmonds syndrome was made.

Barraquer-Simmonds syndrome, is a rare, acquired partial lipodystrophy, that begins during childhood and most commonly affects women. Patients present with a distinctive clinical phenotype characterized by progressive loss of subcutaneous fat affecting the upper body, in a cephalocaudal distribution. In contrast, the lower body is spare or shows an increased fat deposition. Unlike other lipodystrophy syndromes, metabolic abnormalities are infrequent. The pathogenesis remains unclear. Although, the frequent detection of C3 nephritic factor (C3NeF), accelerated complement activation, and association with autoimmune disorders such as membranoproliferative glomerulonephritis suggest an autoimmune etiology. Recently the HLA DRB1:11:3 has been found in a significant number of patients suggesting a potential role.

* Corresponding author.
E-mail address: docrozas@yahoo.com (E. Rozas-Muñoz).

https://doi.org/10.1016/j.ad.2022.01.022
0001-7310/© 2022 Published by Elsevier España, S.L.U. on behalf of AEDV. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).