Barraquer-Simons Syndrome: An Unusual Form of Acquired Partial Lipodystrophy in a Child with Lupus Nephritis

Dear Editor,

The Barraquer–Simons syndrome is a rare type of partial symmetric lipodystrophy characterized by the loss of subcutaneous adipose tissue and is primarily limited to upper part of the body. It is often associated with autoimmune connective tissue disorders like systemic lupus erythematosus and dermatomyositis.[1] The condition has a female predisposition with a female to male ratio of 4:1.[2] The affected patients often have insulin resistance and other metabolic complications. Here, we describe a case of acquired partial lipodystrophy in a child with lupus nephritis.

A 11–year-old male child diagnosed with focal lupus nephritis class III presented to the skin out patient department (OPD) with muscle and joint pain and progressive swelling of face with loss of fat predominantly from the lower part of face, cheek, and temple area for last six months [Figures 1 and 2]. The child was on treatment with prednisolone 35 mg, hydroxychloroquine 200 mg, enalapril 5 mg, and monthly injection of cyclophosphamide at 500 mg/m^2 dose. On clinical examination, there was loss of subcutaneous fat pad from bilateral cheek and temples. The repartition of fat did not affect trunk and lower parts of the body. The child had developed some degree of cushingoid body habitus from long-standing oral steroid therapy for lupus nephritis [Figure 3]. He had joint tenderness involving both small and large joints and proximal myopathy. Examination of other systems was unremarkable. On biochemical evaluation, anti nuclear antibody test (ANA) was positive 4+ (1:160 titer) homogenous pattern and anti-ds DNA was positive. Echocardiography revealed chink of pericardial effusion with mild aortic regurgitation. Electromyography features were suggestive of myopathy. Magnetic resonance imaging of brain showed symmetrical confluent calcification involving bilateral lentiform nuclei, thalamus, and cerebral gyri. Serum urea and creatinine levels were marginally elevated. Lipid profile, liver function test, and blood glucose level were within normal limit. Serum C3 level was low. Serology for hepatitis B, C, and HIV was negative. The overall clinical presentation and biochemical

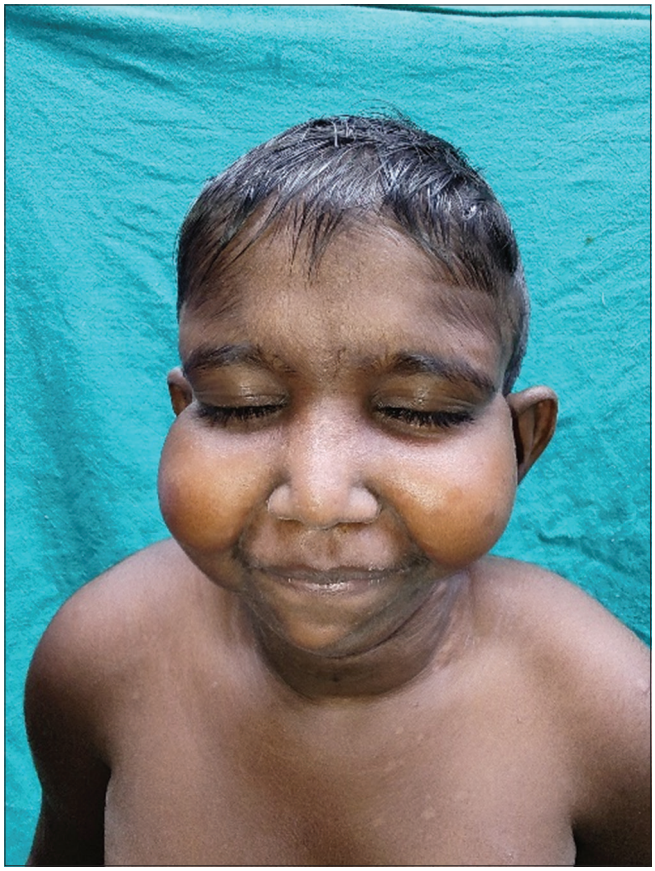

Figure 1: Loss of fat from check and temple area

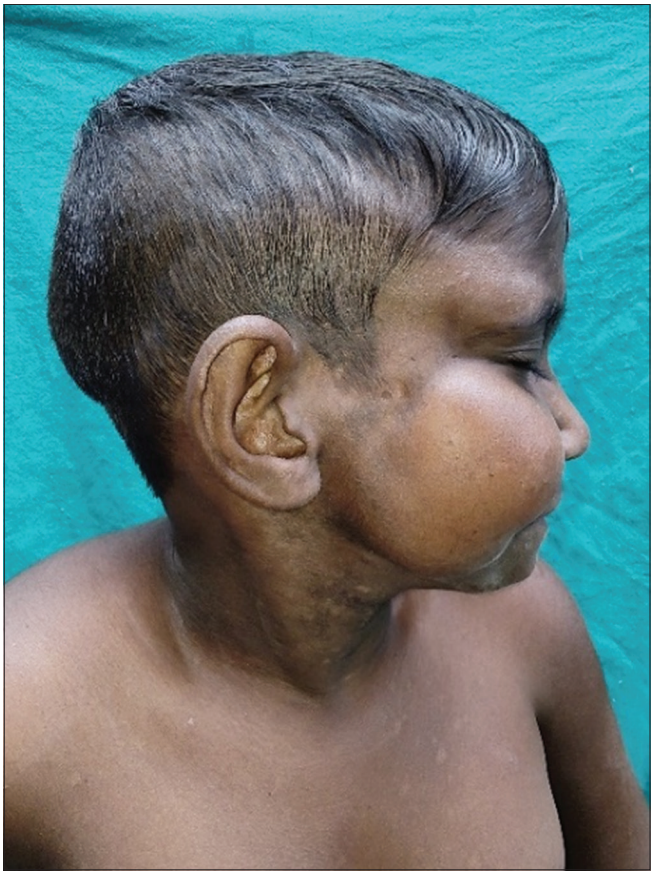

Figure 2: Lateral view showing the characteristic partial lipodystrophy

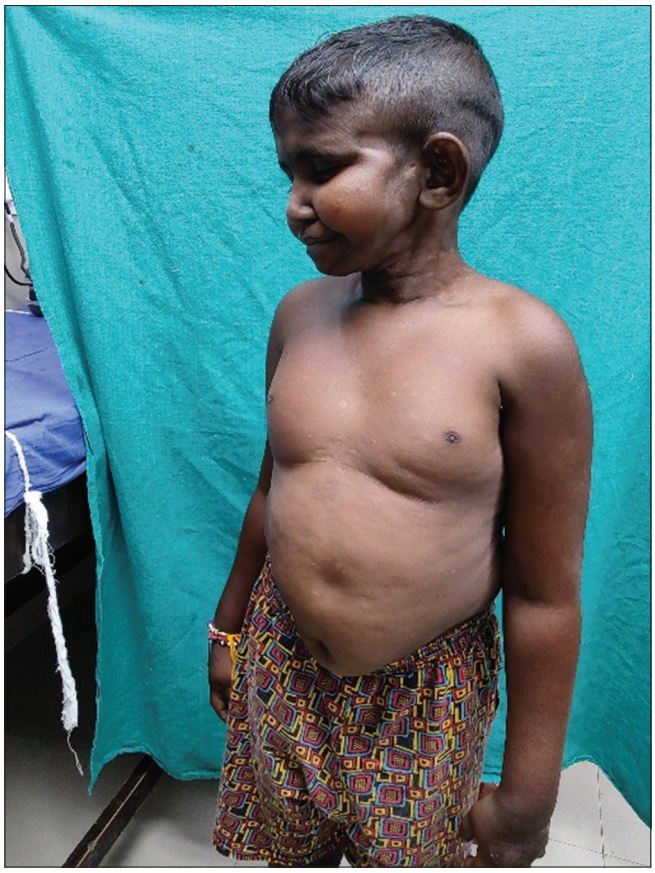

Figure 3: Cushingoid body habitus due to prolonged steroid therapy, no lipodystrophy seen in other body parts

parameters led us to consider Barraquer-Simons syndrome as the main diagnosis.

The lipodystrophy syndromes constitute a rare group of disorders characterized by a selective deficiency of adipose tissue without any other explainable cause like inflammation or nutritional deficiency.[3,4] Barraquer-Simons syndrome or acquired partial lipodystrophy primarily occurs among children and young adults. The exact pathogenesis of selective fat loss in patients with acquired partial lipodystrophy is largely unknown. However, alternate complement pathway activation and C3 hypocomplementemia with lysis of adipocytes induced by C3NeF have been implicated.[5] The condition can be associated with metabolic syndrome, acanthosis nigricans, hirsutism, and autoimmune connective tissue disorders like systemic lupus erythematosus, dermatomyositis, and localized scleroderma. Currently, highly active antiretroviral therapies are the most common cause of lipodystrophy worldwide and other causes are much rarer. In our case, childhood onset, involvement of face and sparing of other body parts, associated lupus nephritis, low C3 level, and negative history of antiretroviral therapy led us to the clinical diagnosis of acquired partial lipodystrophy. The presence of low C3 helps to differentiate Barraquer-Simons syndrome from other forms of lipodystrophy. The diagnosis is often difficult owing to the rarity of the condition and its insidious course. There is no effective treatment for Barraquer-Simons syndrome currently. Therapeutic approach is primarily aimed at plastic surgery to improve aesthetics and early diagnosis and management of systemic complications. Recently, metreleptin, a recombinant analogue of human leptin, has been approved for the treatment of metabolic derangements of lipodystrophy. Our patient was prescribed hypolipidemic diet and regular exercise along with continuation of medications for lupus nephritis. Renal involvement is a major prognostic factor in Barraquer-Simons syndrome and patients may require renal transplantation for end-stage renal disease due to glomerulonephritis.

Declaration of patient consent

The guardian of the patient consented to publish data regarding his medical condition and has been assured that his child's identity will not be disclosed anywhere in the process.

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Conflicts of interest

There are no conflicts of interest.

Nirjhar Mondal, Sudip Mandal, Pramit Nandy, Biswanath Naskar

Department of Dermatology, IPGME and R, SSKM Hospital, Kolkata, West Bengal, India

Address for correspondence:
Dr. Nirjhar Mondal,
Department of Dermatology, IPGME and R, SSKM Hospital, 17/2/1 Ishwar Ganguly Street, Kolkata - 700 026, West Bengal, India.
E-mail: nirjharmondal1@gmail.com

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