Partial Lipodystrophy

Sir,

Partial lipodystrophy is a rare condition characterized by progressive loss of subcutaneous fat from the face, neck, shoulder, forearms and trunk. It is much more common in females, having its onset in childhood. It is usually associated with C3 hypocomplementemia; and sometimes, with glomerulonephritis, impaired glucose intolerance, and mesangiocapillary glomerulonephritis etc.¹ There is no definite treatment available for this disease so far. However, plastic surgery has played some role in recovery.

A 10-year-old girl presented with loss of fat in upper half of body for the last three and a half years. The process started gradually first with loss of fat in the buccal region, later on involving both arms; sparing forearms and hands. Since the last few months, there was vulvar atrophy noticed by the mother. There was no family history of such kind of disorder. Clinical examination revealed symmetrical atrophy of fat over face, both arms and vulvar region, sparing the rest of the body (Figure 1).

The laboratory evaluation revealed normal blood counts, serum biochemistry, renal parameters, thyroid function test, and stool composition. ANA and RA factors were also negative. The lipid profile was also normal with serum lipid level. However, the plasma C3 level was reduced to 22 mg/dl (normal: 50–120 mg/dl) and the C4 level was normal. On that basis, a diagnosis of partial lipodystrophy was made and the child was referred to the plastic surgeon for further management.

Partial Lipodystrophy (PL) is one of the rare forms of lipodystrophy. There are many names for this syndrome like progressive lipodystrophy, lipodystrophia progressiva, etc. Two main syndromes are included: partial (cephalothoracic) lipodystrophy (Barraquer Simon disease) and partial face spacing lipodystrophy (Dunningon Kobberling syndrome).²

This syndrome has no exact known etiology and genetic basis. Several mechanisms have been proposed. These include an autoimmune process and complement anomalies. Different complement protein have a role in adipocyte lysis.³ Several other autoimmune disorders are associated with this syndrome like mesangiocapillary glomerulonephritis, SLE, dermatomyositis, etc. This disorder usually starts around 8-10 years of age with a predominance in females. The onset usually follow an acute febrile viral illness. The cephalothoracic partial lipodystrophy is characterized by the slow symmetrical disappearance of subcutaneous fat in the facial region and upper half of the body (the Weir Mitchell type). The whole process usually lasts for 1 to 6 years.

Many systemic abnormalities are associated with partial lipodystrophy. The most common one is C3 hypocomplementemia found in near by 70% of the patients. A large number of cases (25-90%) may have renal involvement with the biopsy showing mesangiocapillary glomerulonephritis (dense deposit type), which usually follows the dermatological findings and hypocomplementemia. So partial lipodystrophy is not a simply cosmetic disability, but may also be a generalized and potentially serious disorder.

As far as the treatment is concerned, no specific therapy is available. Treatments tried with limited success include facial contour restoration with dermal fat grafts, fat injection, microvascular free-flap grafts and even injections of liquid silicone. Associated problems also require close follow-up and their prompt treatment as well.

REFERENCES


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