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LIPODYSTROPHIC SYNDROME ASSOCIATED WITH DERMATOMYOSITIS.

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BACKGROUND

Dermatomyositis is an idiopathic inflammatory myopathy characterized by proximal muscle weakness and typical skin manifestations. Etiopathogenesis is multifactorial, with the participation of genetic and environmental factors. Cases of acquired forms of lipodystrophic syndrome in association with dermatomyositis has been reported.

CASE REPORT

40 years old woman, diagnosed with dermatomyositis in 2007, after presenting with proximal muscle weakness, calcinosis cutis, heliotrope and Gottron papules and interstitial lung disease (NSIP pattern) with associated pulmonary hypertension. She has been treated with corticosteroids, cyclophosphamide, azathioprine, zoledronic acid (for calcinosis cutis) and is currently using rituximab. She developed diabetes mellitus requiring insulin therapy. An endocrinology evaluation was requested on account of difficult glicemic control, which lead to the diagnosis of lipodistrorphic syndrome, based on the presence of diabetes mellitus, mixed hyperlipidemia, hepatic steatosis and loss of fatty tissue in the face, upper trunk and upper limbs.

CONCLUSION

Lipodystrophic syndromes comprise a heterogeneous group of congenital or acquired disorders characterized by complete or partial lack of fat tissue. The Barraquer-Simons syndrome, reported in approximately 250 people, also known as acquired partial lipodystrophy, is characterized by the loss of fat tissue of the face and upper trunk, sparing, or even increasing of adipose tissue in the rest of the body. In addition, metabolic abnormalities such as insulin resistance, hyperinsulinemia, hypertriglyceridemia and low concentrations of HDL are common. Autoimmune diseases such as dermatomyositis, hypothyroidism, pernicious anemia, rheumatoid arthritis, temporal arteritis and mesangiocapillary glomerulonephritis have already been described in association with lipodistrorphic syndrome.