



Case Report

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Adult onset dermatomyositis associated with lipodystrophy and underlying calcinosis: Case report

Suad Hannawi*¹, Issa Al Salmi¹

¹ Associate Professor, Ministry of Health, Dubai, P.O. Box 65522, Unite Arab Emirates

Abstract

Dermatomyositis is an idiopathic inflammatory myopathy, with skin manifestations. A less frequently recognized but clinically important complication of dermatomyositis is lipodystrophy (LD), and associated metabolic abnormalities that increasingly recognized as complications of juvenile dermatomyositis. Lipodystrophy is less frequently reported in adult onset dermatomyositis. In this paper we are presenting a case of adult onset dermatomyositis associated with lipodystrophy and underlying localized calcinosis; at the exact distribution of lipodystrophy.

Keywords: Dermatomyositis, Lipodystrophy, Calcinosis.

INTRODUCTION

Dermatomyositis is a chronic systemic autoimmune disease characterized by proximal muscle weakness and characteristic skin rashes^[1,2]. Dermatomyositis occurs in childhood; Juvenile dermatomyositis has certain characteristic presentations such as calcinosis cutis, that has been frequently reported^[3]. More, Lipodystrophy and associated metabolic abnormalities are being increasingly recognized as complications of juvenile dermatomyositis^[4].

Although, adult onset dermatomyositis have almost all of the juvenile dermatomyositis features, lipodystrophy with underlying calcinosis (distributed among the same body regions) has not been reported previously.

In this case report we are presenting an adult onset dermatomyositis with lipodystrophy and underlying localized calcinosis; at the exact distribution of lipodystrophy.

CASE REPORT

We report a case of 38 year old lady, who developed adult onset dermatomyositis, initially she presented with classic skin findings, fever, tiredness, non destructive polyarthralgia, and proximal myalgia and myopathy of both upper and lower limbs. All the symptoms were of two months duration.

Physical examination revealed violaceous eruption (heliotope rash) on the upper eyelids, accompanied by eyelid swelling, periungual erythema, and mechanic's hands. At the time of initial examination, she had unremarkable vital signs. No lipodystrophy or soft tissue calcifications detected at this stage. Apart from the above mentioned signs the rest of the systemic examination was normal.

Laboratory investigations showed high inflammatory markers. Other results including Creatinine Kinase level, troponins, metabolic panel, and complete hemogram were within normal limits. Serological examination was inconclusive.

Patient diagnosed as a case of adult onset dermatomyositis and started on prednisolone and Methotrexate.

A year after regular treatment and disease control, with disappearance of all the dermatomyositis related clinical signs, she developed multiple areas of regional lipodystrophy. That was mainly located over the lateral aspect of her thighs (figure 1), buttock and lower back (figure 2).

*Corresponding author:

Dr. Suad Hannawi

Associate Professor, Ministry of Health, Dubai, P.O. Box 65522, Unite Arab Emirates

Histopathological examination of a skin biopsy taken from the atrophic area (over the right buttock) revealed marked epidermal atrophy with basal cell liquefaction degeneration. There was perivascular chronic inflammatory reaction with periadenexal lymphocytic infiltrate. Subcutaneous fat was scanty. The skin findings were consistent with lipodystrophy.

X-rays (obtained after development of lipodystrophy) showed muscle calcifications distributed around the affected area of lipo-atrophy (figure 3).



Figure 1: Lipodystrophy at the upper lateral aspect of the thigh



Figure 2: Lipodystrophy at the lower part of the back



Figure 3: Pelvis x ray with soft tissue calcification distributed over the body region of lipodystrophy

DISCUSSION

A less frequently recognized but clinically important complication of adult onset dermatomyositis is lipodystrophy^[1], in which patients lose subcutaneous fat in a localized or generalized distribution, frequently with resultant metabolic abnormalities such as insulin resistance and hyperlipidemia^[4]. These patients have a loss of mature, functional adipocytes, as opposed to an absence of lipid in otherwise normal adipocytes^[5].

Bringham et al reported that although lipodystrophy is associated with Juvenil Dermatomyositis, It has been infrequently reported in adult onset dermatomyositis. The reason for this lack of association with adult myositis are unclear^[1].

Clinically, our patient had typical skin changes of adult onset dermatomyositis and proximal muscles weakness and tenderness. There were hyperpigmentation over the eyelids, mechanic hands and perungual erythema at time of diagnosis. Though her muscle enzyme studies were normal, she had proximal muscle weakness and pain. These features were consistent with adult dermatomyositis, which has been confirmed by histopathological findings.

A year after diagnosis, lipodystrophy occurred, with loss of subcutaneous fat over the buttocks and upper thighs. Histopathology of the atrophied areas showed loss of subcutaneous fat in the affected area. Despite a report of increased incidence of metabolic abnormalities in association with lipodystrophy^[4], our patients showed no such findings of dyslipidemia or dysglycemia.

Although it has been reported that prevalence rates for acquired lipodystrophy associated with Juvenile Dermatomyositis vary from 12% to 40%^[4, 6-8], this manifestation rarely reported in adult onset dermatomyositis. Interestingly, among Juvenile Dermatomyositis, it has been reported that lipodystrophy is associated with more severe chronic disease and sequelae such as calcinosis^[1]. Same finding has been observed in our patient; Lipodystrophy with underlying calcinosis distributed over the same affected areas.

CONCLUSION

The presented case of adult onset dermatomyositis showed lipodystrophy and underlying calcinosis after one year of clinical diagnosis. Although it has been reported that lipodystrophy is a late complication of Juvenile Dermatomyositis, and it's associated with more severe chronic disease, and sequelae such as calcinosis, few

reported it presence in adult onset dermatomyositis. However, our case did not have any metabolic abnormalities. The presence of lipodystrophy and calcinosis might be associated with severe form of adult onset dermatomyositis. Hence, a proper management strategy for such cases may be required.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

HS wrote the manuscript and compiled the figures. AI edited the manuscript. All authors read and approved the final manuscript.

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We thank the patient for giving us consent for the publication of the case report.

Consent

An informed consent was obtained from the patient for publication of this case report and accompanying images in a medical journal. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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