Case Report

Juvenile dermatomyositis associated with lipodystrophy

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ABSTRACT

Lipodystrophy and associated metabolic abnormalities are being increasingly recognized as complications of juvenile dermatomyositis. We report one such case.

KEY WORDS: Juvenile dermatomyositis, Lipodystrophy

INTRODUCTION

Dermatomyositis is a disease in which characteristic patterns of autoimmune inflammatory injury occur in striated muscle and skin.[1] Dermatomyositis also occurs in childhood and has certain characteristic presentations. Juvenile dermatomyositis is frequently associated with calcinosis cutis.[2] Lipodystrophy and associated metabolic abnormalities are being increasingly recognized as complications of juvenile dermatomyositis.[3] We report a case of juvenile dermatomyositis associated with lipodystrophy and hypertriglyceridemia.

CASE REPORT

A 10-year-old girl presented with a history of fever, joint pains and pigmentation over the eyelids (Figure 1) of nine months' duration. On examination, she was thinly built with a wasted appearance over the chest and limbs but with a protuberant abdomen. The muscle outlines and veins were clearly visible over the limbs, indicating a loss of subcutaneous fat (Figure 2). Both the eyelids and the ears were erythematous and hyperpigmented. Scaling and hyperpigmentation was present over the scalp. The skin could be easily pinched. Skin colored papules were present over the metacarpal and interphalangeal joints over the dorsum of both hands. Multiple subcutaneous nodules were present around the elbow and knee joints (Figure 3). They were mobile and tender and extruded a whitish pasty material. The hair, mucosa, nails and palms and soles were normal. Investigations showed a normal hemogram with normal levels of serum calcium and serum uric acid. Muscle enzymes such as CPK, LDH and aldolase were within normal limits. ANA was negative. Lipid profile revealed a raised triglyceride level of 250 mg/dl as compared to the normal value of below 200 mg/dl.

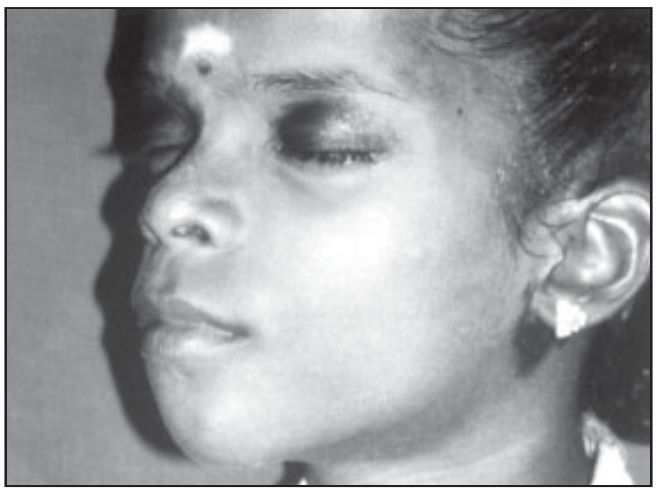

Figure 1: Heliotrope erythema

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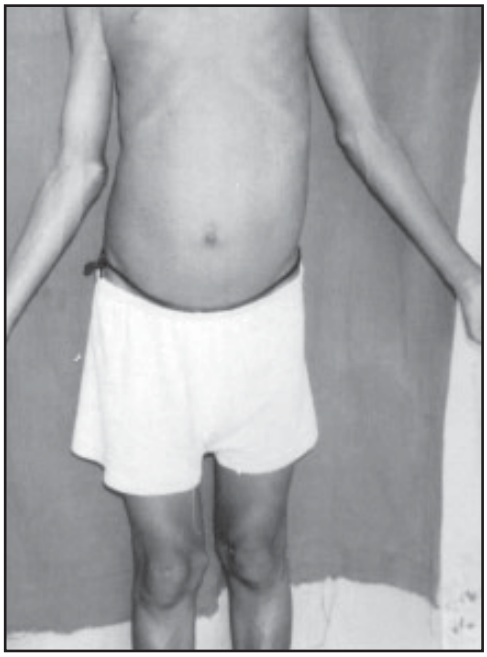

Figure 2: Lipodystrophy

X-rays of the elbow and knee joints showed extra-articular calcifications. X-ray of the chest and sonogram of the abdomen were normal.

Histopathological examination of a subcutaneous nodule showed calcium deposits in the dermis. Histopathology of the deltoid muscle showed a loss of transverse muscle fibres with hyalinization of the sarcoplasm and sparse lymphocytic infiltrate. In one area there was no subcutaneous fat, with muscle fibres being arranged directly below the dermis.

DISCUSSION

Calcinosis cutis occurs more frequently in childhood or juvenile dermatomyositis than in the adult type. Clinically our patient had hyperpigmentation over the eyelids, Gottron's papules and calcinosis cutis. These features were consistent with juvenile dermatomyositis. Though her muscle enzyme studies were normal, she had muscle weakness and pain, and histopathology also showed involvement of muscle. Lipodystrophy was clinically evident, with loss of subcutaneous fat over the limbs and chest.

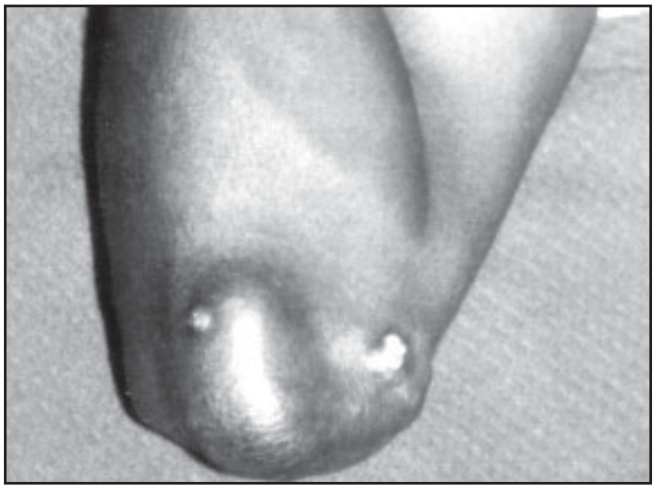

Figure 3: Calcinosis cutis over the elbow

Histopathology showed loss of subcutaneous fat in some areas. Metabolic abnormalities were in the form of raised triglyceride levels and LDL cholesterol.

In a study of twenty patients with juvenile dermatomyositis, Huemer et al found that there was an increased incidence of lipodystrophy and metabolic abnormalities. Twenty five percent of the children studied showed lipodystrophy and 50% had hypertriglyceridemia.[3] Juvenile dermatomyositis has also been reported with partial lipodystrophy.[4] Our patient is yet another evidence of lipodystrophy with triglyceridemia which is being increasingly reported in children with dermatomyositis.

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