

Case Report

Juvenile dermatomyositis associated with partial lipodystrophy

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Abstract Dermatomyositis with its cutaneous, myopathic and other systemic manifestations stand a high chance of malignancy in adults, but in children it has a rather benign and prolonged course and may have a number of late sequelae. We present here a case of juvenile dermatomyositis with partial lipodystrophy as a late sequel.

Key words

Juvenile dermatomyositis, partial lipodystrophy.

Introduction

Like the disease in adults, the juvenile variant of dermatomyositis also affects the skin, muscles and blood vessels. However it may differ in its severity, course, and sequelae. The disease is severe and generally has prolonged course taking several years to burn out. Late recurrences are uncommon. It is rarely associated with malignancy. There is a higher incidence of soft tissue calcification. Contractures may develop in 5% of cases. There are sparse reports of partial lipodystrophy among childhood cases. The following case is one of the rare associations of juvenile dermatomyositis and partial lipodystrophy.

Case report

A 10-year-old child reported with six years history of persistent erythema around his eyes, shoulders and back of trunk. It was

associated with intermittent blue discoloration of his fingers and pain involving small joints of hands, elbows and shoulders, more so during winter season. After one year of the disease the parents noticed gradual change in his facial features. The boy started losing weight. His cheeks sunk and he developed a cadaver look. The limbs became thin and his buttocks lost their rounded contours. These complaints were initially associated with generalized muscle aches and pains with difficulty in climbing stairs but settled with oral steroid treatment which he received from local general practitioners.

For the last three months his periorbital and shoulder erythema had recurred and he also developed painful purpuric rash over the tips of his fingers along with erythematous papules over the dorsum of his hands. There was no associated muscle pain but felt difficulty in rising from squatting position. The loss of facial fat and the rounded contours of the buttocks persisted. There was no history of shortness of breath, changing voice or difficulty in swallowing food.

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Figure 1 Heliotrope rash with sunken cheeks.



Figure 4 Gottron's papules over dorsa of hands.



Figure 2 Thin limbs.



Figure 5 Vasculitic rash over finger tips of both hands



Figure 3 Shawl sign.



Figure 6 A hard nodular lesion over left elbow.

On examination he was an active looking child with sunken cheeks (**Figure 1**), thin limbs (**Figure 2**) and flattened contours of his buttocks. Cutaneous examination

revealed heliotrope rash (**Figure 1**), shawl sign (**Figure 3**), Gottron's papules over

dorsa of hands (**Figure 4**) and vasculitic rash over finger tips of both hands (**Figure 5**). In addition he had a hard nodular lesion over left elbow (**Figure 6**). On examination of his limb girdles he had difficulty in standing from squatting position. He however, did not have any muscle tenderness.

Skin biopsy was performed from the atrophic area over his left buttock, which revealed marked epidermal atrophy with basal cell liquefaction degeneration and sparse perivascular and periadenexal lymphocytic infiltrate. Subcutaneous fat was scanty and there was no infiltrate. The skin findings were consistent with lipodystrophy. His laboratory parameters include erythrocyte sedimentation rate (ESR) - 44 mm fall after 1 hour with normal total and differential leukocyte counts. Creatinine phosphokinase was normal and aldolase was 14.3 U/L (upper limit of normal). Serological examination was inconclusive. X-rays of his left elbow showed soft tissue calcification (**Figure 7**).

The patient was placed on tablet prednisolone 5 mg; 8 tablets daily along with topical steroids for facial rash. Steroids were continued in this dose for 2 months and after that gradually reduced to 10 mg daily, which the patient is still using after 5 months. There is marked improvement in the skin rashes and ability to rise from squatting position. The lipodystrophy, however, is still persistent.

Discussion

Juvenile dermatomyositis is a rare inflammatory myopathy with characteristic



Figure 7 X- rays of his left elbow showed soft tissue calcification.

cutaneous eruptions. It affects children of either sex between 2-18 years of age. The clinical course may vary from mild weakness of limbs to severe physically incapacitating illness. Likewise cutaneous manifestations may range from mild violaceous erythema involving extremities to extensive cutaneous injury to the extent of subcutaneous fat atrophy. Most patients exhibit periorbital erythema (heliotropic rash) and vasculitic rash over finger tips and shoulders (shawl sign). Flat violaceous papules (Gottron's papules) are commonly observed over metatarsophalangeal and interphalangeal joints along with periungual redness and telangiectasias.

Calcification occurs more often in children than in adults. Widespread vasculitis affecting small arteries, capillaries and veins of skin, muscles, gastrointestinal tract and subcutaneous tissue is a prominent feature of so called Banker's type 1). It may take years for the disease to burn out. Occasionally a child may recover without any residual disability or suffer from a late recurrence. Psychological difficulties are common.²

Subcutaneous involvement is extremely rare. These may precede, occur simultaneously, or appear as a late sequel to the muscle changes.³ Only 10 cases of lipodystrophy associated with dermatomyositis have been reported since 1924,^{3,4,5} out of which only three cases have been reported in children.^{6,7,8} First case was reported in 1991.⁷ The lesions appear as painful subcutaneous nodules commonly over arms, buttocks, thighs and abdomen. Other noticeable associations include periungual infarcts, calcinosis cutis, flexion contractures and cutaneous and mucosal ulcers.

On initial musculoskeletal examination, individual or group muscular weakness may be noted. Many patients also exhibit signs of synovitis. On electron microscopic examination of muscle tubuloreticular inclusions are visible within the endothelial and perithelial cells.

Abnormal laboratory findings may be non-specific in the initial as well as late stages of the disease. Elevations of erythrocyte sedimentation rate, lactic dehydrogenase and creatinine phosphokinase are present in most cases with active disease but these parameters may be deceptive at times and serial recordings are recommended.⁹ Likewise serial muscle biopsies and electromyographic recordings are suggested for correct interpretation of clinical data.

Rest is essential in acute phase of the disease. Corticosteroids are the mainstay of treatment. Dose is 1-2 mg/kg body weight in children, which should be gradually reduced and maintenance dose of 5-10 mg/day may be required for many months. Children with

minimum myopathy may remit with indomethacin without steroids and antimalarials may help the rash of dermatomyositis. After an acute phase lasting some months most cases settle slowly and burn out in time. Therapy with steroids may be required for this period and reduced very gradually with monitoring the muscle enzymes.

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