Juvenile Dermatomyositis

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Abstract

Juvenile dermatomyositis (JDM) is an autoimmune vasculopathy affecting children and adolescent under the age of 18 years. In this report, we describe a 9 years old boy who had myopathy and typical skin rash. Upon treatment with oral prednisolone and topical corticosteroid the patient condition considerably improved. Our case report illustrates that JDM requires comprehensive evaluation and multidisciplinary management.

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Introduction

Juvenile dermatomyositis (JDM) is a rare autoimmune vasculopathy of childhood that preferentially affects dermal and muscular vessels. By definition, the onset of JDM is prior to the age of 18, whereas the average onset is in the 7th to 8th year of life, with a slight preference for the female gender[1]. the etiology of JDM is not yet clear, but there is a disproportional association with certain HLA alleles, such as B8[2], DRB1*0301, DQA1*0501[3] and DQA1*0301[4] has been reported, suggesting genetic susceptibility.Polymorphisms of both the TNF-α promoter[5] and the interleukin-1 receptor antagonist[6] have been identified as additional risk factors. The incidence of DM is 9.63 cases per million population[7]. Muscle weakness may occur concurrently or after weeks to years[8]. Dermatomyositis may be associated with systemic manifestations like malaise, arthralgia, dyspnoea, etc. However, subcutaneous calcifications are especially common in children[9].Early diagnosis and aggressive treatment can lead to remission and prevention of severe complication[10].

Case Report

A 9 years old boy presented to our department with malar and periorbital heliotrope erythema in figure I below:

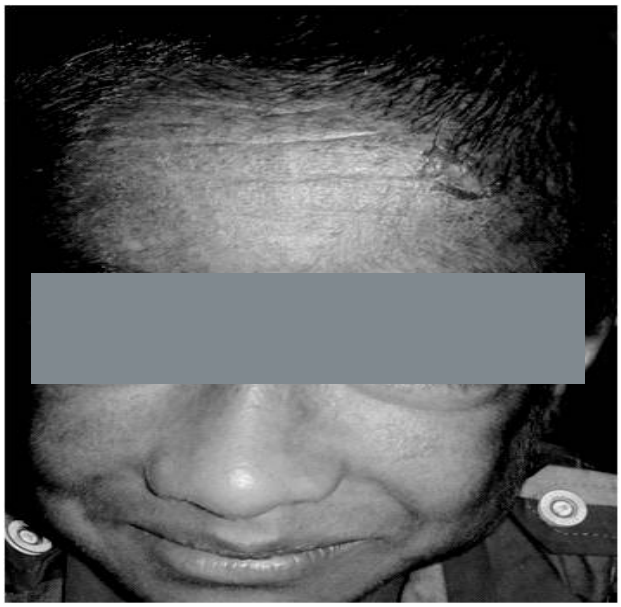

Fig- I: Periorbital heliotrope erythema

The rash also involve forehead, chest and different parts of the body in figure II below:

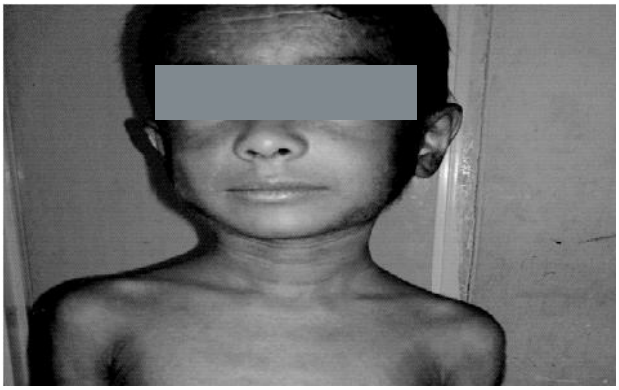

Fig- II: Rash involving chest, forehead and face.

He had severe proximal muscle weakness and arthritis of the different joints. All symptoms persist for more than one year, physical examination revealed erythematous scaly plaques (Gottron papules) over metacarpophalangeal and proximal interphalangeal joints in figure III below:

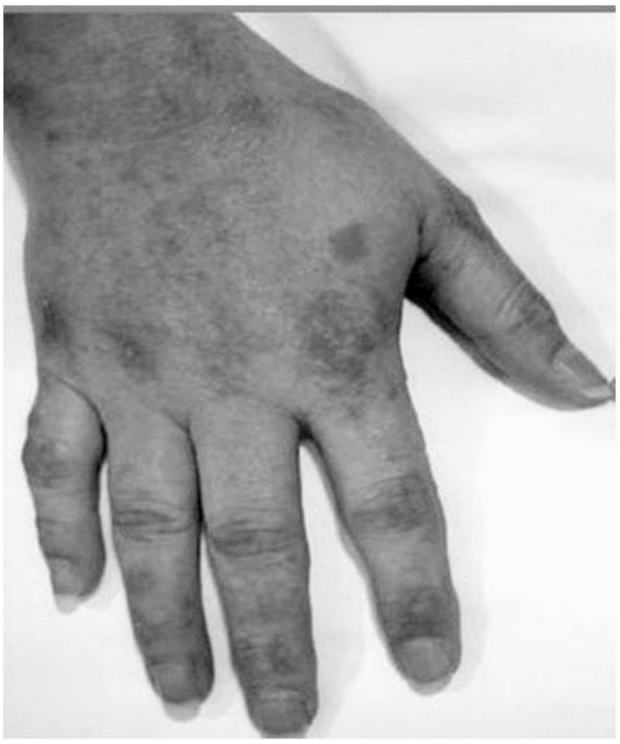

Fig- III : Gottron papules.

and calcinosis cutis, a tender dystrophic calcification over palmer surface of the finger in figure IV below:

Fig- IV: Calcinosis cutis over palmer surface of finger.

Laboratory examination revealed elevated Creatin kinase and Aldolase, negative rheumatoid factor, Blood count showed Hb(9.9 gm /dl),ESR 60 mm in 1st hour Total count 15200/cmm, X-ray chest and abdominal ultrasonogram showed no abnormality. X-ray of calcinosis cutis showed calcification.

Based on clinical appearances, laboratory and imaging data the diagnosis of JDM was made.

Patient was administered oral prednisolone 2mg/kg/day for 6 weeks then tapered. Calcium and Diltiazem 30 mg once daily was given. In addition topical steroid clobetason proprionate was given for skin lesion for 3 months. The skin lesion and muscle weaknes gradually resolved after 6 weeks of treatment (fig IV). After 3 months skin manifestation had disappeared and muscle enzymes were within normal range. Continuation of the treatment is planned for at least 12 months.

Discussion

Diagnostic criteria for JDM are currently still based on those established by Bohan and Peter[11], which include a characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes. The presence of 3 of these criteria characterizes definite JDM, whereas the prevalence of 2 criteria makes the diagnosis probable. The key to a favourable outcome in cases of JDM is early diagnosis and aggressive pharmacologic corticosteroid treatment[12]. However early diagnosis is often hampered by the nonspecific nature of the the initial signs of JDM, such as fatigue, fever, weight loss, irritability, myalgia and arthalgia. Identification of characteristic skin lesion may help to establish an early diagnosis. Typical cutaneous lesions include a characteristic periorbital heliotrope rash (present in more than two-thirds of patients), facial malar rash, Gottron papules (livid scaly plaques on the extensor surface of joints), and nailfold changes that may present as periungual infarcts. Nailfold capillaroscopy shows reduced capillary density, capillary dropout, branching, and dilatation[13]. In addition, nonspecific eruptions on the extremities and mouth, skin ulcers, lipodystrophy[14], psoriasiform scalp dermatitis[15], and limb edema have been described. Myopathy, mostly affecting the proximal muscles, is present in about 95 percent of dermatomyositis cases ; the existence of amyopathic dermatomyositis is controversial. Myalgia may precede skin rashes, thereby posing a diagnostic challenge[16]. However our patient had typical skin rash, gottron papules and severe proximal myopathy which help us to make diagnosis easy. Muscle enzymes are usually elevated in JDM. In our patient CPK and Aldolase was raised. Histopathologic findings characteristic of dermatomyositis include a moderate perivascular lymphocytic infiltrate around the superficial and deep vascular plexus of the dermis with some lymphocytes scattered at the dermoepidermal junction.but histopathology was not done in our cases. Systemic glucocorticosteroids are the mainstay of therapy.They are administered oraly (up to 2 mg/kg/day of prednisolone) and therapy is continued until there is improvement of clinical and laboratory parameter[17].

Our patient has tolerated oral prednisolone well in figure V below:

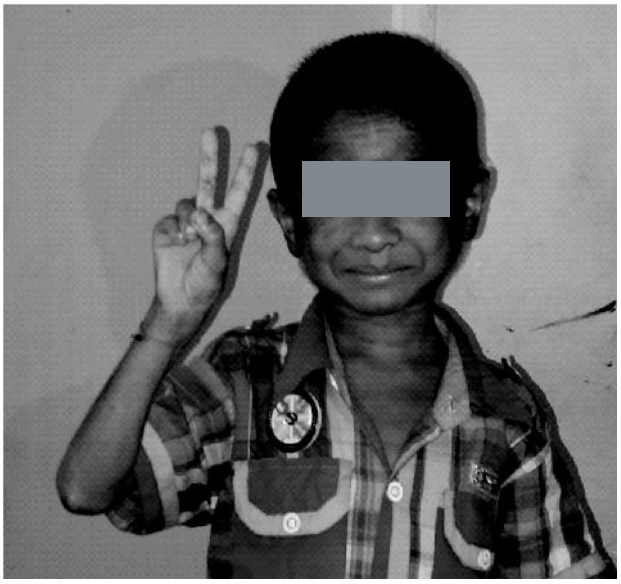

Fig- V: 3 months after treatment.

Juvenile Dermatomyositis is a rare disease which causes chronic disability in children.Early diagnosis and effective management with proper pharmacotherapy can prevent morbity and mortality.

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