Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report DOI: 10.6003/jtad1262c3

A Case of Juvenile Dermatomyositis Associated with PDA Presenting with Heart Failure and Pulmonary Hypertension

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Published:
J Turk Acad Dermatol 2012; **6 (2)**: 1262c3
This article is available from: http://www.jtad.org/2012/2/jtad1262c3.pdf
Key Words: Juvenile dermatomyositis, PDA, heart failure

Abstract

Observations: Juvenile dermatomyositis (DM) is an uncommon disease of childhood and one of the idiopathic inflammatory myopathies. Heterogeneous clinical picture seen in the disease that may also resulting from severe systemic involvement. Here we report a 8-year-old case of juvenile DM associated with patent ductus arteriosus (PDA) presenting with heart failure and pulmonary hypertension because of unusual presentation. The patient was a dramatic response by methylprednisolone therapy and agressive management results in improved outcome and decreased severe complications of the disease.

Introduction

Juvenile dermatomyositis (DM) is a rare disease of childhood and one of the idiopathic inflammatory myopathies. It is clinically characterized by progressive symmetrical proximal muscle weakness and a specific skin manifestation [1]. Heterogeneous clinical picture seen in the disease that may also resulting from severe systemic involvement [2].

Extramuscular manifestations include joint contractures, dysphagia, pulmonary symptoms, and subcutaneous calcifications cardiac disturbance [2].

Majority of cardiac lesions found in the course of DM are silent, and clinical manifestation has seen in only 10% to 15% of cases [2]. The cardiac effects of DM has found frequently at autopsy [3]. Myositis, cardiomyopathy, pericarditis, valvular defects, and rhythm or conduction disturbances were reported in DM [2].

Here we report a 8-year-old case of juvenile DM associated with patent ductus arteriosus (PDA) presenting with heart failure and pulmonary hypertension because of unusual presentation. The best of our knowledge the association of juvenile DM and PDA has not been previously reported in the literature.

Case Report

A 8-year-old girl was referred to outpatient clinic of pediatrics with skin rash, edema on limbs, weakness, cough, dyspnea, inability to walk. Skin eruptions with pruritus and erythema firstly appeared on face of the patient 6 months before ad-

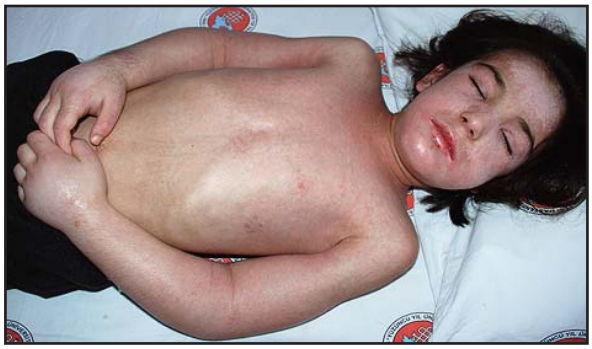

Figure 1. The patients pretreatment appearence: Anasarca eudema and poikilodermatous reticulate erythematous lesions were seen.

mission to hospital. The patient had cutaneous sun-sensitivity. Edema, weakness, fatigue and pain was noted two months ago. These complaints were more prominent on the arms and legs. She could not climb stairs, and raise her head in the supine position for 20 days. Because she also had proximal weakness in the limbs, she was unable to stand or sit up unaided. She was also unable to get out of bed unassisted. In addition, the patient noted weight loss, cough and difficulty of breathing and swallowing.

On physical examination decreased breath sounds and bibasilar crepitant rales, tachycardia, gallop rhythm and a grade 4/6 systolic murmur, hepatomegaly and splenomegaly. Generalized anasarca edema was noted. Muscle power on upper and lower extremities was 0/5. She had generalized hypotonia and deep tendon reflexes were absent on all extremities.

Dermatological examination revealed erythematous scaly plaques (*Gottron* papules) over knee joints. Poikilodermatous reticulate erythematous lesions were apparent at the upper thoracic, V-sign location, postinflammatory hipopigmentary macular lesions, desquamation on face and malar rash (**Figure 1**).

Laboratory examination revealed elevated levels of lactate dehydrogenase (LDH) (1905 U/l), creatinine kinase (4356 U/l), alanine transaminase (ALT) (70 U/l) and aspartate aminotransferase (AST) (365 U/L). Hemoglobine was 8.3 g/dL and white blood cell count was 13000 /mm^3. Circulating *von Willebrand* factor antigen, a marker of vasculopathy, was negative. Antinuclear antibody (ANA) (1/320) and anti skin antibody (1/10) were positive. Anti ds DNA, C3, C4, rheumatoid factor, lupus anticoagulant, VDRL-RPR, anti smooth muscle Ab (ASMA), anti-mitochondrial antibody (AMA), anti-extractable nuclear antigen (ENA) antibodies, liver

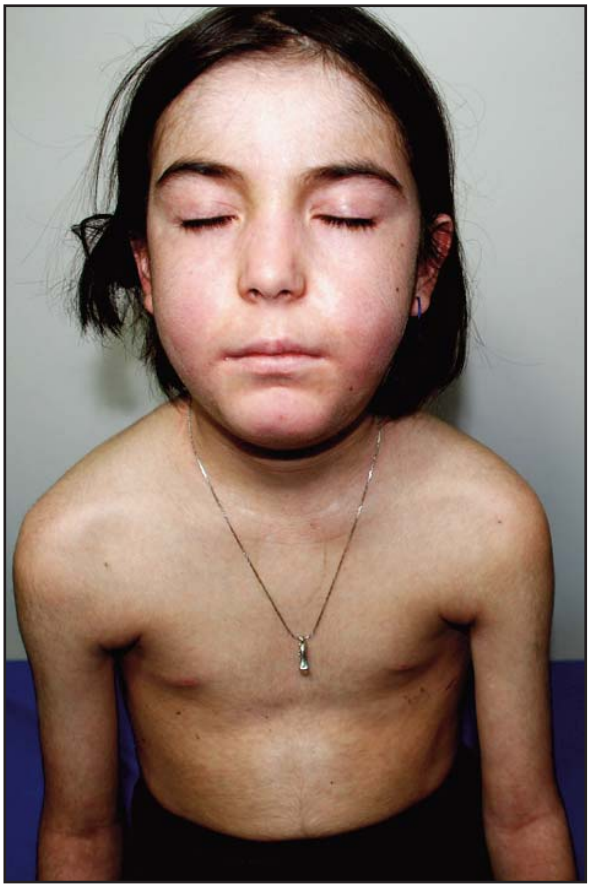

Figure 2. The patients post treatment appearence.

kidney microsomal antibody (IFA), mono test, treponema pallidum hemaglutination (TPHA), and anti-neutrophil cytoplasmic antibodies (ANCA) were negative. Chest radiograph showed bilateral perihilar pneumonic infiltration. Abdominal ultrasonography revealed 14 cm hepatomegaly. Echocardiography showed atrial septal defect, mild tricuspid insufficiency and PDA. Electromyogram (EMG) showed findings of myogenic involvement. Histopathologic examination of muscle revealed inflammatory myopathy.

The patient was hospitalized with the diagnosis of juvenile DM, PDA, congestive heart failure and bronchopneumonia. On physical examination, his general condition was moderate, vital signs were stable. Her weight was 25 kg (25-50 p), her length 122 cm (10-25 p). The patient's oral intake was not allowed. 1200 cc/m^2 intravenously fluid was started. 1 mg/kg was made from single-dose furosemide. To the patient whose insufficiency diagnosis do not regressed has began captopril and digitalized. For the bronchopneumoniae of this patient ampiciline-sulbactam and seftriaxon has given. High dose MTP was given to patient with juvenile DM was administered in a dose of 30 mg/kg/day

for 3 days. After 2 mg/kg/day maintenance of steroid therapy was continued. Responded dramatically to steroid therapy. She began to walk on the second week of corticosteroid therapy. She was discharged from hospital on the 25 day of admission. Edema and proximal weakness completely resolved 1 month. After initiation of therapy creatine kinase and lactate dehydrogenase gradually improved. She had no any significant symptom. Her abnormal skin signs markedly decreased and muscle enzymes were within normal ranges. The patient was on the 4th month of follow-up, and she continued use prednisolone. PDA was disclosed by using catheter-angiography, on the 40 day of hospitalization (**Figure 2**).

Discussion

Juvenile DM is a systemic autoimmune disease that is characterized by symmetric proximal weakness and distinctive skin rashes [**4**]. The incidence of juvenile DM is 2-3 cases per one million per year [**1**]. The etiology of juvenile DM remains unknown, but recent evidence suggests that combinations of genetic and environmental risk factors are involved [**4**].

Skin rash often precedes the onset of weakness by weeks to months. Early in its course, rash and muscle enzyme elevations may be the sole manifestations of DM [**1**]. In our case the skin findings began four months before beginning of systemic symptoms. The skin rashes of juvenile DM are numerous and varied. Classical juvenile DM skin rashes are well recognized: the heliotrope rash (66–100%) (purple-pink in color) over the eyelids, periorbital edema and *Gottron*'s papules (57–100%) (skin thickening over the extensor surfaces of joints). These cutaneous abnormalities are apparent in approximately three-quarters of patients presenting with juvenile DM. Other cutaneous findings are nail fold capillary changes (91%), malar or facial rash (42–73%), oral ulcers (35%), skin ulcers (23–30%), limb edema (11–32%) and calcinosis (6–30%) [**5**]. Generalized edema has been reported rarely. Our patient had *Gottron*'s papules, malar rash, reticulate erythematous macules and plaques on the face and trunk, generalized anasarca edema.

Heterogeneous clinical picture seen in the course of DM results from generalized changes within the tissues and the organs and is caused by vasculitis and fibrous degeneration. Clinical observations indicate that inflammatory lesions may be found not only in skeletal muscles but also in myocardium and pulmonary interstitial tissue [**2**].

The cardiac effects of DM are found frequently at autopsy, even though the clinical manifestations are rare and typically subclinical [**3,5**]. The mechanism of muscle destruction in skeletal muscles and in myocardium in DM; that process is caused by muscle infiltration with inflammatory cells, focal myocyte necrosis, and fibrosis [**2**].

The most prevalent electrocardiographic (ECG) changes seen in patients with DM include nonspecific ST-segment changes (71%-80% of cases) and T-wave changes (58% of cases). The other frequently reported changes include conduction disturbances, pathological Q waves, ventricular and supraventricular arrhythmias, and features of the sick sinus syndrome [**2**]. In our case no abnormality was noted on ECG.

Serious cardiac involvement is rare in juvenile DM, but mainly includes heart murmurs and cardiomegaly. Pericarditis has also been described; although more severe defects such as acute myocarditis, AV blocks and ventricular or supraventricular tachyarrhythmia conductive defects and first-degree heart block occur less frequently, they can be associated with significant sequelae [**5,6**]. Postulated mechanisms include: 1) Formation of re-entry circuits, 2) Myocardial fibrosis due to recurrent inflammation, and 3) Active inflammatory myocarditis [**6**]. She had congestive heart failure, which was most probably secondary to bronchopneumonia and PDA, that is a congenital abnormality, we think that PDA diagnosed in our case is a co-incidental finding.

The outcomes of juvenile DM are better than those of adult DM, and more aggressive therapy for juvenile DM should be advocated [**1**]. In our patient high-dose methyl prednisolone was initiated and then commenced oral corticosteroid therapy. Clinical signs and symptoms of the patient improved dramatically after the therapy.

Juvenile DM is a rarerly disease characterized by inflammation of various tissue and organs Systemic involvement seen at in juvenile

DM. The clinical diagnosis of dermatomyositis is confirmed by three laboratory examinations: serum muscle enzyme concentrations, electromyography, and muscle biopsy. Aggressive management results in improved outcome and decreased severe complication and mortality of the disease.

At this patient we have seen high muscle enzyme, skin findings, muscle and cardiac involvement which were harmoniously with juvenile DM. We think that PDA is a co-incidental diagnosis at this patient. We see that, by methylprednisolone therapy muscle enzymes and muscle spasm diagnosis has ended. This was a dramatic response for us and also this disease clinically is so severe but rare, aggressive management results in improved outcome and decreased severe complications and mortality of the disease.

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