Juvenile Dermatomyositis Early Onset, Case Report

Dear Editor,

Juvenile dermatomyositis (JDM) is the most common idiopathic inflammatory myopathy of childhood. The peak incidence is from 5 to 10 years of age. The average period is 6 months between the onset of the first symptoms and the diagnosis.[1] Although it is the most common idiopathic inflammatory myopathy of childhood, it is rarely seen under 3 years of age. Herein, we present a rare case of early-onset JDM, diagnosed at the age of 27 months.

A 27-month-old male child patient was admitted with the complaints of not being able to walk for 2 weeks, decreased speech, and fever 4–5 times a month. He also had growth retardation and malnutrition. Dermatological examination revealed bright reddish-purple papules and plaques on the dorsal surfaces of the knuckles, reddish-purple rashes on the upper eyelids with periorbital edema and atrophic scars with a full-thickness skin loss under both axillae [Figure 1]. Muscle enzymes, electromyography (EMG) and muscle magnetic resonance imaging (MRI) were requested from the patient with a presumptive diagnosis of JDM. The EMG showed a low amplitude in the left perineal muscles. MRI of the hip showed T2A signal increases in the thigh muscles, iliac region muscles and lumbar region muscles, and muscle edema was observed. Echocardiography (ECHO) was reported as normal.

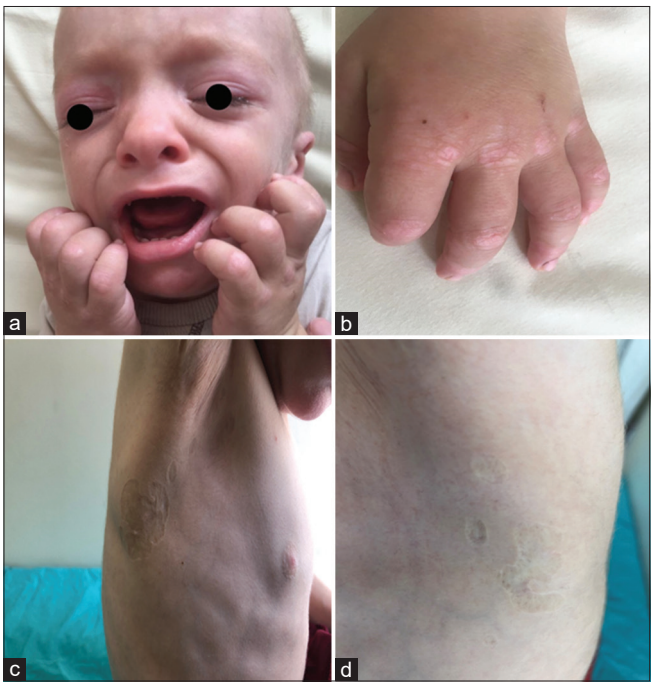

Figure 1: At the time of diagnosis, there were (a) reddish-purple rashes on the upper eyelids with periorbital edema; (b) bright reddish-purple papules and plaques on the dorsal surfaces of the knuckles; and (c and d) atrophic scars with a full-thickness skin loss under both axillae

Creatine phosphokinase (CPK) was 165 U/L (40–170 U/L), creatine kinase myocardial band (CK-MB) was 40 IU/L (0-25 IU/L) and lactate dehydrogenase (LDH) was 844 U/L (120–300 U/L). Antinuclear antibody (ANA) was 1/1000 granular pattern (+), anti-DsDNA was negative, anti-Jo1 was negative, anti-Mi2 was negative, complement C3 was 128 mg/dl (79–152 mg/dl) and complement C4 was 34 mg/dL (16–38 mg/dl). Muscle biopsy was performed with the pre-diagnosis of dermatomyositis, and the report showed "atrophic striated muscle showing perivascular T-cell infiltration, pan fascicular atrophy in the foreground and regeneration was not observed." Due to the presence of symmetric proximal muscle weakness, muscle biopsy findings, EMG findings, laboratory findings, and a typical skin rash, our patient met all Bohan and Peter classification and diagnostic criteria (5/5), and a definitive diagnosis of JDM was made.[2] 2 mg/kg/day of Methylprednisolone, 1 g/kg of intravenous immunoglobulin (IVIg), and 15 mg/week of methotrexate were started. As the patient was being investigated and diagnosed, new bright purplish erythematous plaques appeared on the bilateral knee and elbow extensors, and the plaques on the dorsum of the hand, finger, and around the eyes became prominent. The child had significant pruritus and was given IVIg therapy for 6 months. The patient, who received decreasing doses of Methylprednisolone and methotrexate for 1 year, achieved near complete improvement with respect to muscle strength. After 1 year of treatment, the patient could walk, run, climb stairs and keep his head upright. The pruritus resolved entirely. Despite effective treatment, the bright reddish-purple papules on the dorsal surfaces of the knuckles, the reddish-purple rash on the upper eyelids and the erythematous plaques on the bilateral knee and elbow extensors, persisted [Figures 2 and 3].

It is thought that genetic susceptibility, immune system mechanisms and infections play a role in JDM pathogenesis. T cell invasion, epitope distribution, ANA

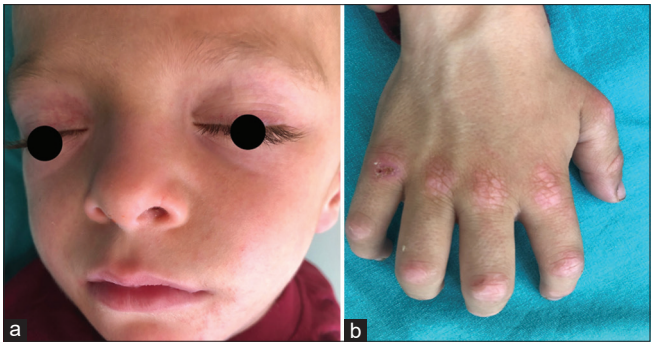

Figure 2: After the treatment, there were (a) reddish-purple rashes on the upper eyelids and (b) bright reddish-purple papules on the dorsal surfaces of the knuckles

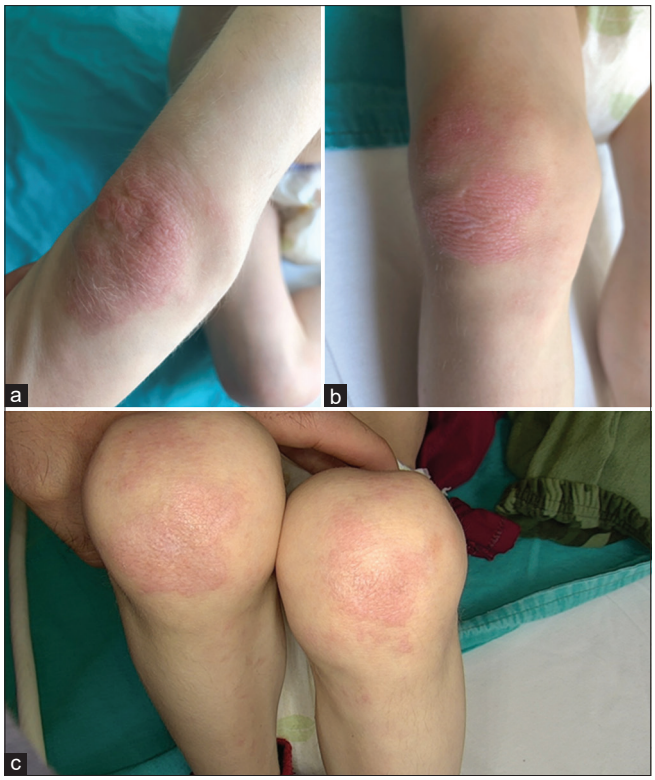

Figure 3: After the treatment, there were (a and b) erythematous plaques on the bilateral elbow extensors and (c) bilateral knees

pattern and myositis-specific autoantibodies (MSAs) suggest immunological processes. JDM may develop as an unusual response to infection in a genetically susceptible host.[3]

Evaluation of nail bed capillary changes is important in the early diagnosis of JDM. The most commonly observed nail bed capillary changes are capillary dilatation, tortuosity and dropout (loss of capillary loops).[3] In our case, nail bed capillary changes were not evaluated before treatment. Myositis-specific antibodies may be helpful in the diagnosis of JDM, but their sensitivity is low. They are especially useful in patients who do not have the typical skin eruptions and who also show features of the diseases included in the differential diagnosis.[3] In our patient, anti-Mi2 was negative, but he was diagnosed with JDM because of the typical findings of the disease.

Early-onset JDM is rare compared to classical JDM. In early-onset JDM, compared to classical JDM, the mean time to diagnosis is longer, calcinosis cutis is more common and muscle biopsy findings are more severe. Even though the rates of remission are the same, the rates of relapse are much higher in early-onset JDM. Advanced step treatments such as IVIg, cyclophosphamide and biologic agents are more required in early-onset JDM.[4] Early-onset JDM is harder to diagnose and has a worse prognosis. Clinicians should keep in mind that JDM can also manifest in children under the age of three.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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