Juvenile dermatomyositis

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Abstract:

Juvenile dermatomyositis (JDM) is a rare multisystemic autoimmune disease of uncertain origin that results in chronic inflammation of striated, smooth muscles and skin in children less than 16 years of age. The hallmark of this disorder is the skin lesions. The case we will present was rare in our clinical practice and it is hoped that will heighten the index of suspicion among medical practitioners .

Key words:

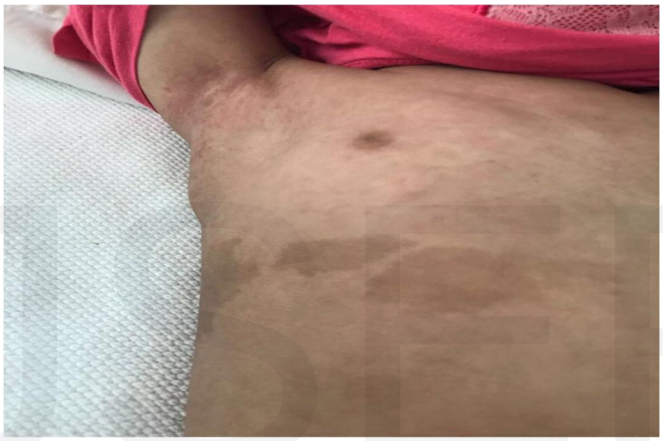
JuvenileDermatomyositis, Heliotroperash, erythmatous non-scaly patches.

Case report:

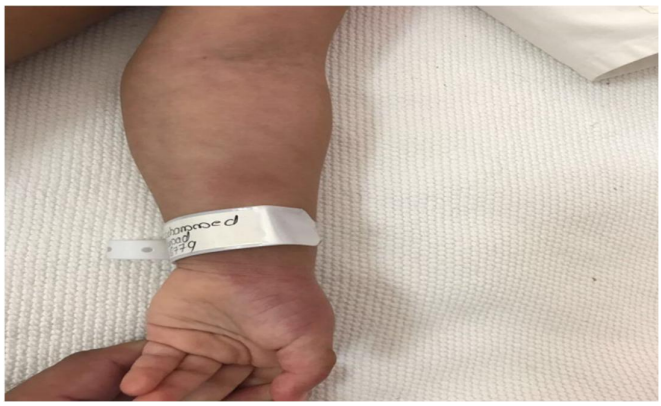
An 3-year-old Saudi girl who presented to the Emergency department of the Abha Maternity & Children Hospital, Asser, Saudi Arabia with a one and half month history of insidious onset of muscle weakness and pain principally affecting the proximal muscles of both arms and legs, mild to moderate in severity with no diurnal variation and is not preventing her from doing her usual activities. She also presented with 2-week history of fever, multiple joints swelling and skin rash. The fever was documented at 38C and was relieved by anti-pyretics, associated with sore throat, runny nose & cough. she diagnosed with URTI at a local hospital and they prescribed antibiotics and anti-pyretic. The multiple joint swelling involving both ankles, knees & wrists, associated with periorbital swelling. As for the skin rash, she developed asymptomatic red lesions, over face, both axilla and left wrist. The muscle pain increased in intensity to the extent that she can't do her usual and normal activity and she started walking with limping. There is a history of painful mouth ulcers, started simultaneously with the previous symptoms. And also there was decrease in urine output and it became more darker in color. No history of genital ulcers, photosensitivity, photophobia, bleeding from orifice, bloody vomiting, bruises, yellowish discoloration of skin or sclera. Also there wasn't history of urine frequency, urgency, dysuria,or suprapubic pain.

Physical examination she looked ill, not dehydrated or in respiratory distress. There was a heliotrope rash above both upper eyelids and her lips were swollen (Figure1). and in both axillae there was erythematous non-scaly scattered macules, there were birth marks (well-defined brownish interrupted patches) on her right side of abdomen (Figure2). There were two well-defined erythmatous non-scaly patches on left hand (Figure 3).

"Figure 1" Heliotrope rash and swollen lip.



"Figure 2" Erythematous non-scaly scattered macules in axillae with birth marks showing on right side of the abdomen.

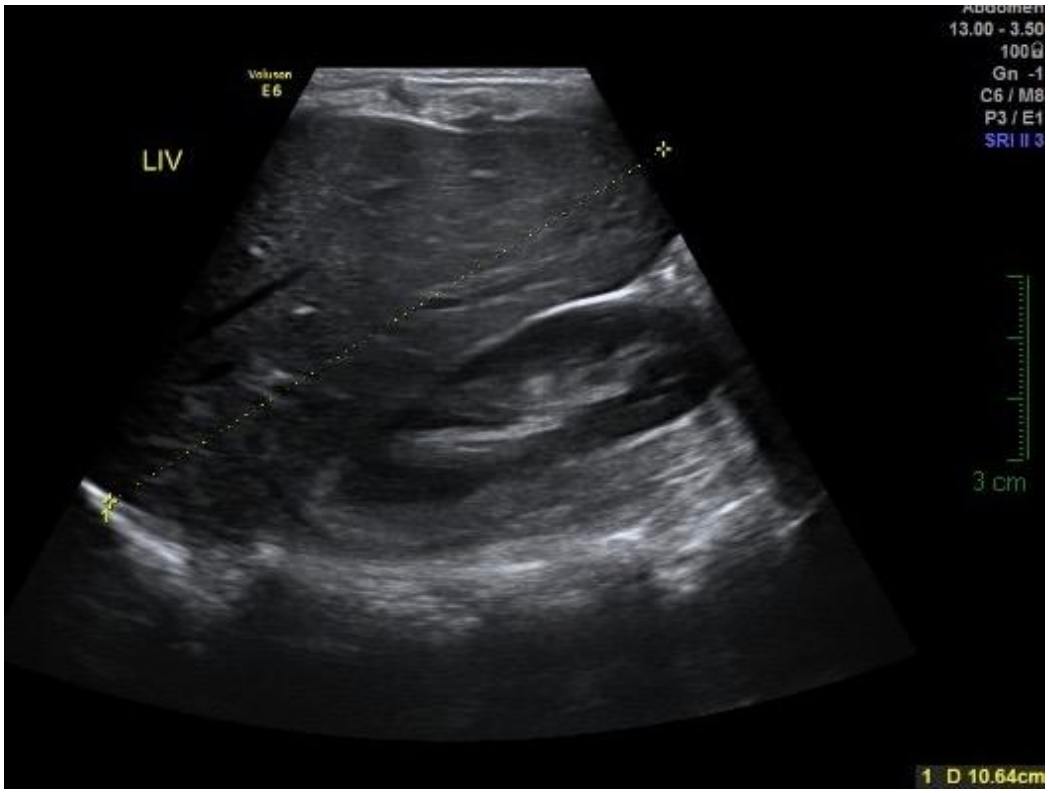


"Figure 3"well-defined erythmatous non-scaly patches.

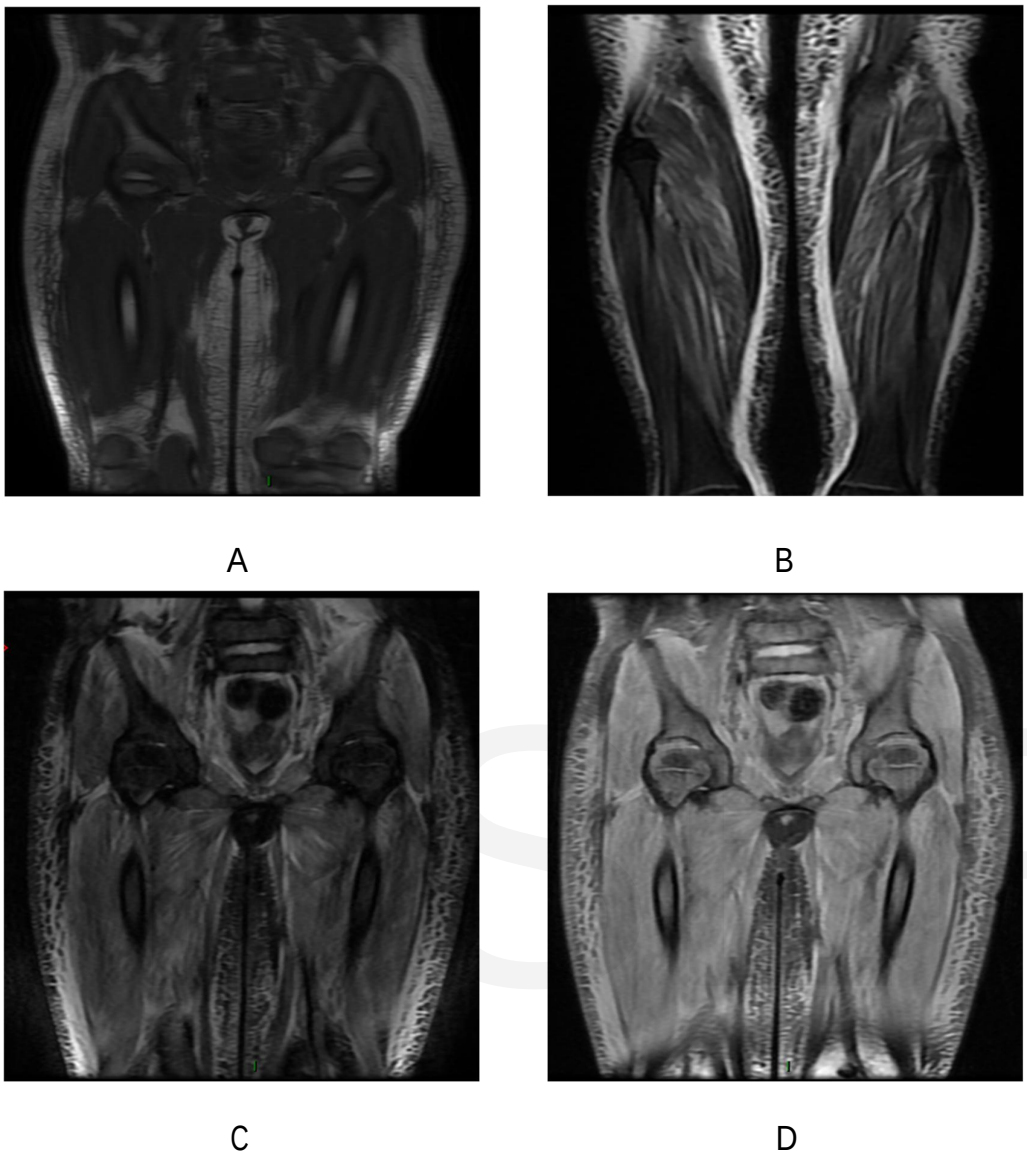
Musculoskeletal examination revealed minimal tenderness of the thigh and proximal arm muscles with marked difficulty in raising her arms above the head. Muscle power in the proximal muscles was grade 2–3 while the distal muscles power was grade 5 . There was no joint tenderness. All other systems were essentially normal.

Laboratory investigations showed elevation of muscle enzymes: CK Total 7477 µ/L , ALT:112 , AST: 396 , aldolase wasn't available. erythrocyte sedimentation rate (ESR) was 35 mm/h. CRP and ASO showed non-reactive results. Serology for Anti-Jo-1, C3, C4, antinuclear antibody and dsDNA antibody were all negative. Serology for HIV negative, tuberculin skin test negative.

The chest radiograph was normal. Echocardiology showed minimal pericardial effusion.
Ultrasound of abdomen showed mild hepatomegaly about 10.5 cm in size (Figure 4). MRI for lower limb showed diffuse inflammation of muscles (Figure 5). EMG and muscle biopsy wasn't done.



"Figure 4" Mild hepatomegaly about 10.5 cm in size.



"Figure 5"

(A) T1 delineated muscles from the surrounding bones and subcutaneous fat.

(B) T2 (C) T2 STIR, (D) T2 FS demonstrate diffuse hypersignal intensity throughout the visualized muscles associated with subcutaneous edema.

The diagnosis of definite juvenile dermatomyositis was made based on the Bohan and Peter diagnostic criteria namely insidious onset of symmetrical proximal muscle weakness, heliotrope rash, raised muscle enzymes and MRI for muscles showing radiological features of myositis.

Treatment was initiated with pulse intravenous methylprednisolone 0.5 mg/kg (320mg) daily for 5 days followed by ceforoxime 700mg IV q8hr, oral prednisone 25 mg daily in divided doses and methotrexate 5 mg once weekly. She also received Acyclovir ointment, vitamin D3,folic acid and calcium supplements.

Physiotherapy was also initiated. There was resolution of fever and muscle pain within few days with gradual improvement in muscle strength within the next few weeks. Patient continues to feel well and she is presently on prednisolone tablets 10 mg daily while she continues physiotherapy.

Outcome and follow-up:

The patient got better and was discharged. She is still being followed up by the consultant in his clinic.

Discussion:

Unverritcht had in 1887 previously clarified the muscular and cutaneous manifestation of dermatomyositis as a form of idiopathic inflammatory myopathy(9). Other forms include polymyositis and inclusion body myositis(7). However, there is an over-representation of juvenile dermatomyositis compared with adults. The diagnosis is usually made using the Bohan and Peter criteria developed in 1975(1-8). This is normally used in adults; however, this has been adapted for children as well under the name of juvenile dermatomyositis. Diagnosis of juvenile dermatomyositis was made based on the presence of skin manifestation (heliotrope rash) and four other criteria namely proximal muscle weakness, raised muscle enzymes, and MRI report of myositis, for EMG changes and muscle biopsy they weren't done(2-4). The other skin signs such as Gottron's sign, shawl and Holster's signs(10) are usually also influenced by the presence of antisynthetase antibodies, which are uncommon in children and may therefore explain why these signs were not florid in our patient. Dermatomyositis differs from polymyositis not only by the presence of skin manifestation but also by clinical presentation and biopsy findings(1-4), also the presence of extra muscular features such as fever, dysphagia and dysphonia suggest more of dermatomyositis. The presentation of juvenile dermatomyositis differs from that of adult. Some important differences include the presence of fever, prominent muscle pains, more rapid onset and good response to treatment. It also differs in the increased incidence of overlap with other connective tissue diseases as well as presence of calcinosis. Our patient presented with fever, muscle pain, but did not have features of other connective tissue disease. Anti-Jo-1 was also negative. Calcinosis and metabolic abnormalities such as insulin resistance were not present in our patient possibly because of the short duration of the illness, while such are usually late sequelae of the disease. The patient was managed with corticosteroids with resultant improvement of the condition. Although no established protocols exist at present for the management of juvenile dermatomyositis, evidence suggests that early aggressive management improves outcome whereas a longer duration of untreated disease is associated with a longer time to reach remission and higher rates of complications. Improving both long-term and short-term outcomes requires early recognition, prompt referral to specialist care and appropriate treatment of patients with JDM and other forms of idiopathic inflammatory myositis(5-6). This case report is presented to increase the awareness of this condition in Saudi children who present with manifestations of proximal muscle weakness.

Conclusion:

Juvenile dermatomyositis (JDM) is a rare multisystemic autoimmune disease of uncertain origin that results in chronic inflammation of striated, smooth muscles and skin in children less than 16 years of age. Bohan and Peter developed the diagnostic criteria to JDM(8), sometimes it doesn't come with all skin manifestations and it becomes hard to diagnose if no one considered JDM as one of differential diagnosis. So early diagnose and treatment of JDM will improve the outcome for the patients(6).

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