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Case Report

Juvenile dermatomyositis in an eleven-year-old boy: a case report and review of literature

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ABSTRACT

Juvenile dermatomyositis (JDM) is a rare chronic autoimmune inflammatory condition characterized by systemic capillary vasculopathy and idiopathic inflammatory myopathy (IIM) that primarily affects the skin and musculoskeletal system, resulting in a characteristic rash and symmetric muscle weakness of the upper and lower proximal muscles, increased levels of serum muscle enzymes and myopathic electromyography. Dysphagia, respiratory problems, and aspiration pneumonia can occur due to the involvement of respiratory and oropharyngeal muscles in severe cases. Both immune dysfunction and environmental factors are thought to contribute to etiopathogenesis. It could be linked to genetics, an immune response to a virus, an adverse drug reaction, exposure to sunlight or internal malignancy. JDM is characterized by two periods of peak occurrence: one during infancy which is called juvenile DM (JDM), typically between the ages of 5 and 15, and another during maturity, typically between the ages of 40 and 60. Histologically, it is characterized by the presence of lymphocytic vascular inflammation and endothelial swelling. Management of JDM is complex and warrants a multidisciplinary approach including physiotherapists, dermatologists, specialist nurses and pediatric rheumatologists, with other specialists like cardiologist/pulmonologist. The long-term complications of JDM include prolonged and severe muscle weakness with muscle atrophy, cutaneous calcifications, scarring or atrophy, and lipodystrophy. Calcinosis cutis, a form of dystrophic calcification, is a late sequel of the disease and occurs in up to 40% of patients with this disorder, a major cause of morbidity. JDM has poor prognosis with one third of patients recovering without complication, one third develop disability and one third die. Herein, we reported a case of an 11-year-old boy who presented to us with facial puffiness & weakness in both limbs, classic clinical and histopathological features of Juvenile dermatomyositis (JDM). As early diagnosis and timely aggressive treatment are associated with better outcomes.

Keywords: Juvenile dermatomyositis, Proximal myopathy, Heliotrope rash, Gottron sign, Proximal muscle weakness, Elevated muscle enzymes

INTRODUCTION

Juvenile Dermatomyositis (JDM) is a rare multisystemic autoimmune disease, characterized by chronic inflammation of striated muscles and skin, though other organs may also be affected. It is reported at an annual incidence of 2 to 4 cases per million children per year.[1] JDM has peak occurrence during infancy with ages of 5 and 15. There is a 2:1 ratio of JDM in girls compared to boys.[2] The disease affects all races and ethnicities. Some studies showed that the condition is more common in

Asian children.[3] Clinically, the condition is characterized primarily by a heliotrope rash, Gottron papules, an erythematous photosensitive rash on the face, neck and shoulders, anomalies in the capillary loops and symmetrical proximal muscle weakness.[2] Gottron papules, heliotrope rash and periungual erythema are pathognomonic. Calcinosis, which is the deposition of calcium salts in the skin, subcutaneous tissues or the muscles, is often seen as a late sequel of this disorder.[1] JDM is characterized by symmetrical proximal muscular weakness, particularly noticeable in the neck flexors, hip flexors, and shoulder girdle.[4] 10% of patients experience muscular symptoms before to the development of skin abnormalities, and 30% to 50% of patients have cutaneous disease 3-6 months before developing myositis.[5] The etiopathogenesis of JDM involves a complex interplay of environmental triggers, immune dysfunction, and specific tissue responses. Both cellular and humoral immune mediated destruction of the vasculature, skin, and the muscles are believed to be involved in its pathogenesis.[1] Juvenile dermatomyositis is characterized by systemic capillary vasculopathy that primarily affects the skin and striated muscles.[6] The diagnosis of JDM is made using the "Bohan and Peter criteria" developed in 1975. The diagnostic criteria include characteristic skin rash, symmetric muscle weaknesses of the upper and lower proximal muscles, increased levels of serum muscle enzymes, myopathic electromyography, and characteristic pathologic changes revealed by a muscle biopsy.[7,8] Muscle weakness is the primary feature of the JDM, usually progressive, symmetric, non-selective and greatly affects proximal muscles.[9] Elevated blood muscular enzymes levels are indicative of muscle involvement. Serum CK is a highly sensitive muscle enzyme that is released when muscle damage occurs, particularly during the early stage of the disease. LDH and ALP return to normal levels 2.53 and 3.68 months, respectively. CK and aldolase, on the other hand, take approximately 4.5 months to normalize. Thus, whereas CK is commonly used in international categorization criteria, its reliability diminishes after approximately 4 months from the onset of symptoms to the clinical evaluation.[10]

In acute stage, the erythematous eruption reveals mild hyperkeratosis and epidermal atrophy with effacement of the rate ridge. Basal cell liquefactive degeneration is a common occurrence, and occasionally cytoid body could be detected. Evidence upper dermal edema is present. Additionally, the presence of melanophages could be observed. There is an occurrence of activated T-lymphocytes and macrophages infiltrating the perivascular area. Mucin deposition in the dermis, are indicative of dermatomyositis.[11] Testing for myositis-specific antibodies such as antibodies against histidyl-tRNA synthetase (anti-Jo-1), nuclear helicase (anti-Mi-2), melanoma differentiation associated gene 5 (anti-MDA5), nuclear matrix protein-2 (anti-NXP2), anti-p155/140 and myositis-associated antibodies such as antibodies against signal recognition particle (anti-SRP) and U1-ribonucleoprotein (anti-U1-RNP) should be considered. These antibodies have diagnostic and prognostic value. Myositis-specific antibodies are found in most children with juvenile dermatomyositis but not in healthy children or other autoimmune diseases or muscular dystrophy.[12] Management of JDM is complex and warrants a multidisciplinary approach. The mainstay of therapy is high-dose corticosteroid initially in combination with disease modifying drugs like MTX or cyclosporin A (CsA). The combination of steroids and MTX had the best outcome for efficacy and safety.[13] Since the introduction of corticosteroids, mortality rate dropped and significant improvement in clinical and functional outcome has been achieved.[14].

CASE REPORT

An 11- year- old boy presented to the pediatric outpatient department with a history of insidious onset and gradually progressive pain and weakness, predominantly affecting the proximal muscles of both upper and lower limbs, for the last six months and inability to walk for last one month. Initially, he had difficulty raising his arms and legs, later it was getting worse, harder to move, walk and sit. For the last one month he became bed bound. History of seizures, loss of consciousness and trauma were denied by his mother. His mother also complained of dusky red rash over eyelids with swelling, itching, and photosensitivity and puffiness of face since last three months, pain and swelling over bilateral knee joints for last three years. His past medical and family history was unremarkable. On general examination, the child appeared to be emaciated with a weight 20 kg and height 125 cm. Dermatological examination revealed violaceous erythematous macules on upper eyelid (Heliotrope sign) and edema on upper eyelid (Figure 1a), erythematous papules over the proximal and distal interphalangeal joints (Gottron papules) with a dystrophic and ragged cuticle (Samitz sign) (Fgure 1b), ill-defined erythematous scaly plaques (Gottron sign) over knee joints were found (Figure 1c).

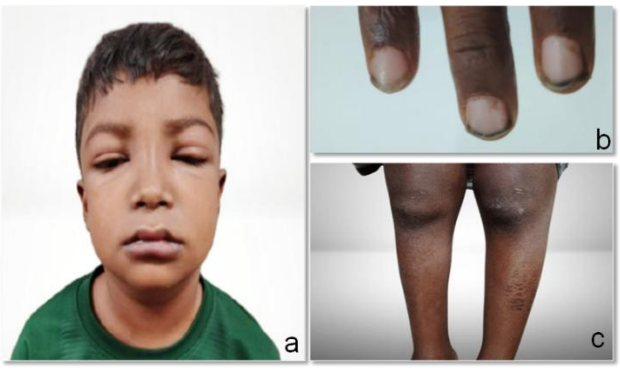

Figure 1: a) Heliotrope rash (violaceous erythema on upper eyelid) and edema on upper eyelid, b) Positive Samitz sign dystrophic and ragged cuticles, c) Gottron sign ill-defined violaceous erythema and hyperpigmentation on bilateral knee.

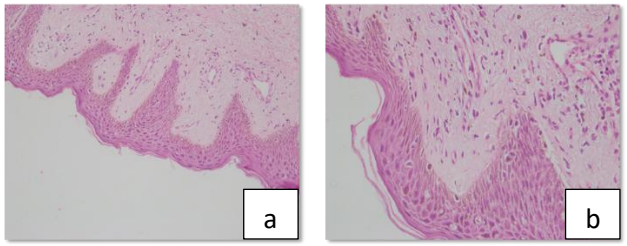

Figure 2 (a and b): Skin histopathology (hematoxylin and eosin, 100X) showing vacuolar degeneration of basal layer and perivascular lymphocytic infiltration.

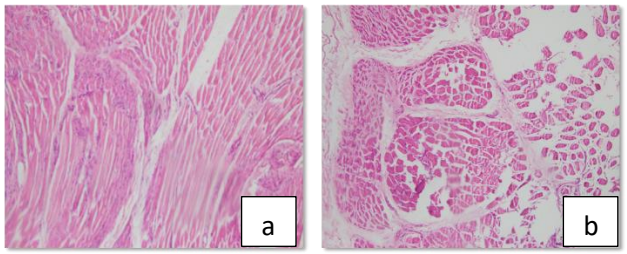

Figure 3 (a and b): Muscle histopathology (hematoxylin and eosin, 100X) showing perivascular and perimysial lymphocytic infiltration.

Musculoskeletal examination revealed minimal tenderness of the proximal muscles of upper and lower limbs. Gower's sign (use of hands and arms to "walk" up the body from squatting position due to proximal myopathy affecting lower limbs) could not be elicited. Muscle power in proximal muscles of both upper and lower limbs was 3/5.

Laboratory examinations revealed complete blood count, renal function tests, thyroid function tests, serum electrolytes, complement C3/C4 levels were within normal limits. Test for ANA and anti-ds-DNA were negative. Muscle enzymes were elevated like lactate dehydrogenase (LDH) 646 U/l (N:120-246U/l), creatine kinase (CK) 553U/l (N:30-200 U/l) aldolase 13.2U/l (N:2.5-10U/l).Myositis-specific antibodies such as antibodies against histidyl-tRNA synthetase (anti-Jo-1), nuclear helicase (anti-Mi-2), U1-ribonucleoprotein (anti-U1-RNP), anti -Ku antibodies were negative.

A punch biopsy from skin, which on histopathology revealed atrophy, hyperkeratosis in epidermis, vacuolar alteration of basal layer, basement membrane thinning. Dermis showed moderate perivascular lymphocytic infiltration with melanophages and mucin deposition.

Muscle biopsy and histopathology revealed moderate infiltrations of mixed inflammatory cells seen in the perivascular & perimysial connective tissue and a good number of muscle fibre showed nuclear internalization, which are compatible with inflammatory myositis.

Based on clinical, biochemical, and histopathological features, the boy was diagnosed with JDM. Treatment was initiated after consultation with pediatric rheumatology department, with two immunomodulator drugs prednisolone 20 mg daily and methotrexate 7.5 mg per week, along with physiotherapy, calcium and vitamin D as supportive treatment for muscle weakness and slow improvement was reported by the patient.

DISCUSSION

The case presented above fulfilled all the components of the diagnostic criteria for JDM. McCann et al conducted a study with the largest European cohort of children with dermatomyositis. A total of 63 out of 175 children with a new diagnosis of myositis were recruited at the time of diagnosis. The most common presenting features were characteristic rash, weakness, tiredness, Gottron's patches and myalgia. Muscle biopsy, magnetic resonance imaging and muscle enzymes were abnormal.[15] Jun HOU et al reported the clinical data of three patients of Juvenile Dermatomyositis (P1-P3), aged from 10 years 3 months to13 years 4 months. All presented with rash and received oral methylprednisolone and cyclophosphamide pulse therapy, while human immunoglobulin was given only to P1 and P2. P1 developed rapid progressive pulmonary interstitial disease (RPILD) and died of respiratory failure after 2 months. While P2 and P3 were followed up for 1 to 2 years, who had complete remission, as anti-MDA5 antibody turned to negative and ILD improved significantly. They concluded that the cases of JDM with positive anti-MDA5 antibody mainly presented with rash and mild muscle weakness, and could be complicated with ILD, pneumothorax and mediastinal emphysema without respiratory symptoms at early stage. Anti-MDA5 antibody titer is related to disease activity and can turn to negative after treatment.[16]

Varnier GC et al develop and test a new multidimensional questionnaire incorporating all main parent/child-reported outcomes (pcros) in pediatric rheumatology practice to be used in the assessment of children with JDM in standard clinical care.[17] The Juvenile Dermatomyositis Multidimensional Assessment Report (JDMAR) includes 15 parent/child-centered measures that assess well-being, pain, functional status, health-related quality of life, fatigue, disease activity, disease status and course, disease manifestations, side effects of medications, therapeutic compliance, problems at school, and satisfaction with illness outcome. A total of 107 consecutive JDM patients were included in the study and the proportion of parents who reported normal scores on the various JDMAR scales are summarized in table.[17]

Ishaque et al reported a 14-year-old girl, with a history of fever, joint pain, easy fatigability and a rash since the age of 3 years at Aga Khan University, Pakistan. Physical examinations, laboratory evaluation, electromyography (EMG) and muscle biopsy were suggestive of dermatomyositis. The report highlighted the importance of muscle biopsy as the gold standard for diagnosing dermatomyositis.[18]

Table 1: Multidimensional assessment report of juvenile dermatomyositis.[17]

JDMAR assessments on a 10-cm VAS	Pts assessed	No. positive (%)	Others JDMAR assessments	Pts assessed	No. positive (%)
Normal well-being	98	39 (39.8)	Normal functional ability	97	53 (54.6)
No pain	99	52 (53.6)	Normal HRQL - Total score	94	27 (28.7)
No fatigue	73	24 (32.9)	Remission	92	50 (54.3)
No disease activity	67	31 (46.3)	Satisfied with illness outcome	93	66 (71.0)

CONCLUSION

A case of a young boy with classical clinical and histopathological findings of a rare autoimmune disorder has been presented. It is important to diagnose and initiate immunosuppressive treatment early to reduce long-term complications. This case has been reported for its rarity and to emphasize the importance of early and aggressive treatment to prevent long-term disease sequels.

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