ORIGINAL RESEARCH PAPER

Volume-8 | Issue-7 | July - 2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A CASE OF JUVENILE DERMATOMYOSITIS

Paediatrics

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ABSTRACT

Juvenile Dermatomyositis (JDM) is an autoimmune connective tissue disease. It is prevalent in children less than 16 years of age. JDM is an inflammatory disease of the muscle (myositis), skin and blood vessels. The primary symptom of JDM includes muscle weakness and skin rash. It involves predominantly proximal muscles. We hereby report a case of 12 years old girl who presented with muscle weakness and later diagnosed as Juvenile Dermatomyositis.

KEYWORDS

JDM, Autoimmune disease, Muscle weakness

INTRODUCTION

Juvenile dermatomyositis is an inflammatory disease of the muscle (myositis), skin, and blood vessels that affect about three in one million children each year.[1] The cause is unknown. The primary symptoms of JDM include muscle weakness and skin rash.[2] Patients with juvenile dermatomyositis (JMD) develop weakness in the proximal muscles around the neck, shoulders, and hips.[3] This causes difficulty in climbing stairs, getting into cars, getting up from a chair or off the floor, or brushing hair.[4] Most patients have little, if any, pain in their muscles, which distinguishes them from patients with other forms of muscle disease.[5]

CASE PRESENTATION

A 12-year-old girl presented to Paediatric OPD with complaints of not able to get up from the chair, not able to climb stairs and not able to comb hair for past 6 months which was insidious onset and gradually progressive. She also had rashes over the hands and periorbital region for past 3 months. On physical examination, the child was noticed to have Gottron papules which were pink, shiny, thickened plaque over the proximal and distal Interphalangeal Joints Fig.1 . Heliotrope rash was present in the periorbital region Fig.2. Proximal muscle weakness was observed and muscle power was grade 3 as per MRC grading. Investigations revealed thrombocytosis with neutrophilia. A rheumatoid factor was negative. C-reactive protein (CRP) was high – 46 mg/L; Lactate dehydrogenase was high 550 U/L; Creatine phosphokinase (CPK) and ferritin were within normal limits. MRI of thigh muscles showed myositis. In view of clinical rash and evidence of myositis by MRI and elevation of muscle enzymes a diagnosis of JDM was made. The patient was given methotrexate 5mg/week initially for 2 months and was maintained on 7.5mg/week. Methylprednisolone was started on 20mg/day tapered to 2.5mg for 3 months and later maintained on 2.5mg every alternate day for 1 year. On follow up after 3 months, skin rash disappeared and muscle enzymes improved. The patient is in remission for 6 months.

Fig.2 Heliotrope rash

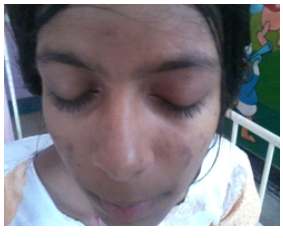
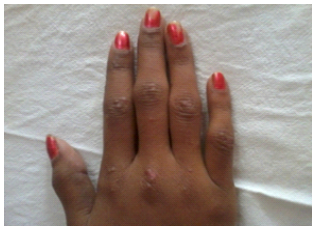


Fig.1 Gottron papules



DISCUSSION

Juvenile dermatomyositis (JDM) is a rare but serious systemic autoimmune condition of childhood primarily affecting proximal muscles and skin. Polymyositis and inclusion body myositis are rare in children. [6,7] The cutaneous manifestations of JDM can be serious and difficult to treat and may progress to ulcerative disease and or subcutaneous calcification, impacting seriously on quality of life in the long term. Clinically children with JDM present with either rash, insidious onset of weakness involving the proximal muscles or both. The characteristic heliotrope rash of eyelids, shawl sign, Gottron papules, ulcerations, calcinosis, Lipodystrophy, Raynaud phenomenon are the cutaneous manifestations of JDM Extracutaneous manifestation is the presence of GI symptoms, ILD, joint contracture &cardiac disease.

Genetic factors have been associated with specific manifestations, disease susceptibility, and severity in both adult onset myositis syndromes and JDM.[8] The tumor necrosis factor α variant, TNFα-308A, carries a higher risk of prolonged disease course, calcinosis, and ulcerative skin disease, Diagnosis requires the presence of characteristic rash as well as at least 3 signs of muscle inflammation (elevated levels of muscle enzymes, EMG changes, muscle biopsy showing signs of necrosis and inflammation.) & weakness.[9] Latest modality involves MRI which reflects the inflammatory response in involved muscle and helps in directing the location of muscle biopsy or EMG. The degree of muscle change on MRI can be quantified on T2 STIR images and has been shown to correlate with disease activity.

While classical proximal muscle weakness is typical, it is also important the child undergoes thorough investigation for other organ involvement, such as of the gastrointestinal tract, including a speech and swallowing assessment. or respiratory system (typically a high-resolution computed tomography scan). In addition to MRI, some centers still perform a biopsy, most usually from quadriceps or biceps muscle; a proposed muscle score tool assessing pathological severity in four domains (inflammation, vasculopathy change, muscle fiber, and connective tissue) has been shown to have good interobserver reliability and to correlate with clinical severity.[10] Treatment involves the use of corticosteroids (oral prednisone 2mg/kg/day) which is the mainstay of treatment. Other treatment modalities involve the use of Methotrexate, IVIG, MMF, cyclosporine, and cyclophosphamide. The mortality rate in JDM has decreased since the advent of corticosteroids from 33% to currently approximately 1%.

CONCLUSION

The prognosis of JDM and the related childhood inclusion myositis has improved dramatically in recent years. Despite this, these conditions remain serious and still potentially life-threatening/ Since they are rare, an evidence base for treatment is difficult to build. High index of suspicion is required in any child presenting with muscle weakness especially in adolescence age group.

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