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A Rare Case Report of Juveile Dermatomyositis

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ABSTRACT

Juvenile dermatomyositis is a rare autoimmune myopathy of childhood. It is primarily a capillary vasculopathy which is distinguished by profound proximal muscle weakness and characteristic skin lesions. Here we report a rare case of JDM in a 6 year old female child who was previously misdiagnosed as a case of atopic dermatitis and was being treated for the last three years, presented to us with proximal muscle weakness of all four limbs and cutaneous rash. The aim of this case report is to make clinicians aware of the manifestations and complications associated with this disease so that early diagnosis and intervention can halt the progress and reduce the morbidity as well as mortality associated with the disease.

Keywords: Gottron Papule, Heliotrope rash, Prednisolone, Calcinosis cutis, proximal myopathy

INTRODUCTION

Juvenile dermatomyositis is an immune mediated disorder primarily affecting the muscles and skin which is characterized by proximal muscle weakness in addition to typical skin lesions, calcinosis and underlying vasculopathy.[1] Though the etiology remains unknown , it is seen that an autoimmune mediated activation of antibodies and complement occurs due to interaction with various environmental factors and infectious trigger.[1]

The major cause of morbidity in this conditon is calcinosis which is a soft tissue calcification, having a prevalence range between 12-30 %. The younger age of onset and severe disease activity has been associated with calcinosis.[2]

The quick recognition of cutaneous signs before the appearance of muscle inflammation may help the clinician in early diagnosis of JDM. The early initiation of appropriate therapy provides a favourable outcome for normal childhood and adolescent development.[3]

CASE REPORT

A 6 year old female child, born of non-consanguinous marriage, presented with rash on face and joint pains with restricted movements involving both the knees, elbow and shoulder joints for the past three years. The pain aggravated with daily activity, however there was no early morning stiffness. The patient also had pus discharge from the skin overlying the right knee for the last 15 days. The redness over the cheeks and bridge of the nose increased on exposure to sunlight and was persistently present for the last three years. The patient had difficulty in walking, sitting from lying down position and lifting hands above the shoulder, however she had no difficulty in combing hair or writing.

During the last 3 years patient had developed reddish itchy lesions on both the arms, legs, trunk with subsequent formation of erosions, covered by crustations which would heal leading to scar formation. She was misdiagnosed as a case of atopic dermatitis and was being treated with emollients and ointments.

She belonged to lower middle socioeconomic status according to modified Kuppuswamy scale, with uneventful antenatal, natal and neonatal period and normal development, immunized till 3 years as per the National Immunization schedule of India. The family history was unremarkable.

On examination, the vitals were stable, had a thin built with a weight of 12 kg (< 3rd centile), height of 102 cm (< 3rd centile) and BMI of 12.49 Kg/m2 (between -2 to -3SD). She was pale, had alopecia, malar rash which crossed the nasolabial folds [Figure 1] a blue-violet discoloration of eyelids which was associated with periorbital edema (Heliotrope rash) . There were pale, shiny, thickened plaques over the proximal interphalangeal and metaphalangeal joints (Gottrons Papule) [Figure 2] and the skin over the trunk showed hypopigmented

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area. There was swelling present over right knee with pus discharge and multiple healed scars present all over the body. The power in the proximal group of muscles (2/5) was less compared to distal muscles (3/5). In addition to this, child had a waddling gait.

Figure 1: Showing malar rash

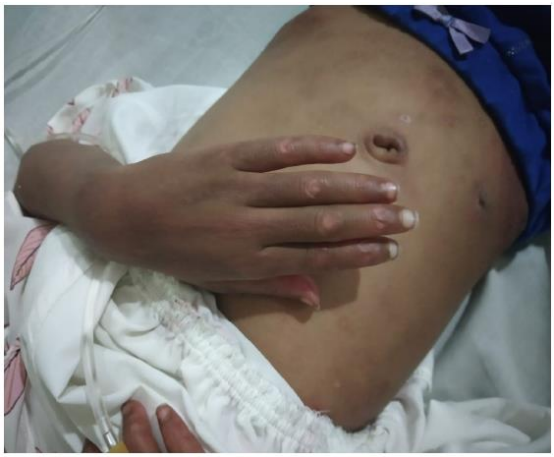

Figure 2: Showing Gottron papule

Investigations included a complete hemogram, renal function tests, urine routine which were normal. The Liver function tests showed an elevated Aspartate transaminase 270 U/L and Alanine transaminase 145 U/L. CRP was positive (59), ESR-28, pus culture showed MRSA with a moderately positive ANA (54.66 U) while Anti DsDNA and Smith antibody was negative. The serum aldolase and LDH were highly elevated however CK was normal. The Xray of knee, pelvis and shoulder joint showed streaks of linear calcification in the muscle plain. [Figure 3] We got the MRI of lower limbs done which was suggestive of inflammation in the muscles. [Figure 4,5]

With the typical clinical features and characteristic laboratory and radiological findings, a diagnosis of Juvenile Dermatomyositis was made.

The patient was started on antibiotics (for pus discharge according to sensitivity), two immunomodulators (prednisolone (2 mg/kg/day), methotrexate (0.5 mg/kg/day), folic acid and supportive therapy.

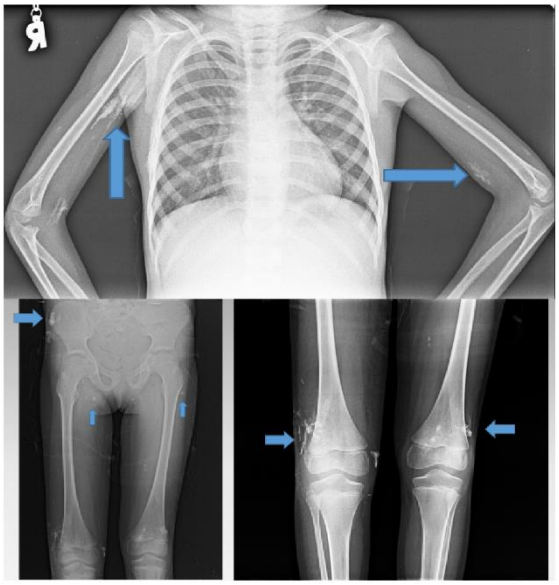

Figure 3: Plain radiograph chest, arms hip and knee joints showing well defined areas of linear sheet like soft tissue calcifications, which were not related to bone or periosteum.

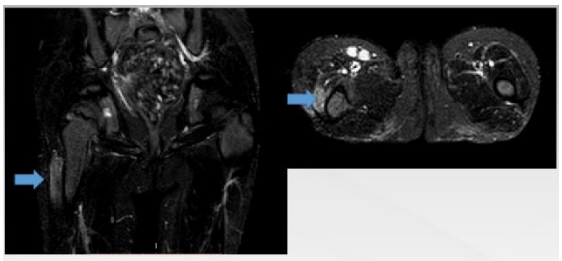

Figure 4: Axial and coronal fat suppressed STIR sequence also showed hyperintensity of same muscle s/o intra muscular inflammation/edema of the proximal part of right vastus lateralis muscle.

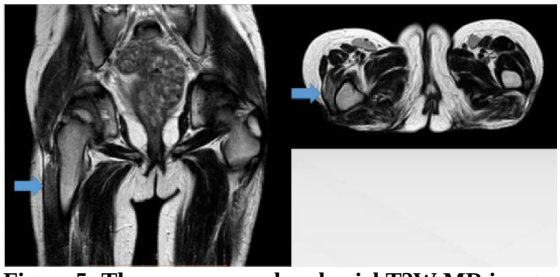

Figure 5: These are coronal and axial T2W MR images of hip at the level of procimal femur which showed hyperintensity in the proximal part of right vastus lateralis muscle.

DISCUSSION & CONCLUSION

Juvenile dermatomyositis is a part of heterogenous group of muscle disease called Idiopathic Inflammatory myopathies.[4] It characteristically involves the muscle and skin. The annual incidence of the disease is 2 to 4 cases per million children.[5-9] The peak age is 5 to 10 years with a 2 to 5 folds greater incidence in girls.[7,8,10]

The etiopathogenesis is unknown, however an autoimmune response with an activation of antibody and complement factors have been seen due to interaction with environmental and infectious trigger.[11] The various clinical manifestations of JDM are proximal muscle weakness, malar rash, heliotrope rash, gottrons papule and periungual nail fold capillary changes and arthritis or arthralgia, .However if remain undiagnosed, the patients can present with complications involving various organ systems, the most common being calcinosis, dysphagia, interstial lung diseases and cardiac arrhythmias.

The diagnosis of JDM was earlier made according to "Bohan and Peter classification and diagnostic criteria" given in the year 1975 which included classic rash plus 3 of the following: Weakness (proximal muscle), elevated muscle enzymes (CK, Aldolase, LDH, Aspartate transaminase), Electromyography changes and muscle biopsy showing necrosis and inflammation. A diagnosis of definite JDM was made if a patient fulfiiled 3 out of the 4 criterias.[12]

Interestingly, it was seen that there were many patients who had only cutaneous manifestations with no muscle involvement known as Amyopathic dermatomyositis who remained undiagnosed because they could not fulfill the criteria. .Besides EMG and muscle biopsy being an invasive procedure, patients were reluctant in getting further investigated.

Hence in 2017 ACR/EULAR came up with a new classification criteria with a score point in patients who underwent muscle biopsy and a separate score for those who did not. The variables included were age of onset, muscle weakness, skin and other manifestations and laboratory investigations.[13]

According to EULAR/ACR classification criteria our patient had a score of 8.1 (Definite IIM).

In the mid1950s and 60s prior to introduction of steroids, the prognosis of JDM was very poor as one third patients died , one third survived with disability while only one third recovered. With the introduction of steroids, the mortality decreased to < 10%. However there were many side effects associated with a long term use of steroids.

In 2012 CARRA protocol came up with the use of combination of prednisolone and methotrexate or cyclosporine as it showed better response and less side effects.[14] Where as in the year 2016 PINTRO trial suggested that a combination of prednisolone and methotrexate had lesser side effects than prednisolone and cyclosporine as the patient can be discontinued from steroids earlier.[15] Hence the standard treatment regimen for the patients with JDM has been the use of high dose corticosteroids with methotrexate .Other agents such as intravenous cyclophosphamide, immunoglobulin, mycophenolate mofetil may be used in severe and refractory cases. Though the biological agents like rituximab and abatecept are increasingly being used in autoimmune diseases, there use is very limited in children with refractory cases of JDM and still under trial.

The complications associated with JDM are Osteoporosis, Calcinosis intestinal perforation, cardiovascular and cerebrovascular comorbidities whereas the risk of malignancy is very rare as compared to adults.

Though the mortality has decreased to less than 2% in the last few decades, it has been found on long term follow up that the cumulative risk of organ damage is greater in patients with high score of disease activity.

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Abbreviations:

JDM- Juvenile dermatomyositis, ACR- American College Of Rheumatology, EULAR- Europeon League against Rheumatism, IIM- Idiopathic Inflammatory Myositis, PRINTO – Pediatric Rheumatology International Trials Organisation, CARRA- Childhood Arthritis And Rheumatology Research Alliance, MRSA- Methicillin Resistant Staphylococcus Aureus.

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