Juvenile Dermatomyositis in a 12 years old girl: A case report.

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Abstract

Juvenile Dermatomyositis (JDM) is a rare but potentially life threatening autoimmune disease of childhood. We report the case of a 12 years old female child with proximal muscle weakness of four limbs, heliotrope rash on the eyelids and Gottron papules was diagnosed to have JDM. Since it was early diagnosed and treated, the patient was recovered quickly.

Introduction

Juvenile Dermatomyositis (JDM) is a rare inflammatory myositis in children, distinguished by proximal muscle weakness and characteristics rash. Inflammatory cell infiltrates result in vascular inflammation is underlying pathology in this disorder. It has an incidence of 1.9 - 4 per million children and prevalence of 2.5 per 100,000.[1] Peak age of onset is between 4 and 10 yr.There is a second peak of dermatomyositis onset in late adulthood(45-64)yr.[2]

Etiology of JDM is multifactorial, based on genetic predisposition and an unknown environmental trigger. Human leukocyte antigen (HLA) alleles such as B8, DRB1*0301, DQA1*0501,and DQA-1*0301 are associated with increased susceptibility to JDM in selected populations. Common environmental triggers like enterovirus and group B streptococcus infection are likely to play an important role in the etiology.[2]

JDM has been associated with significant mortality and morbidity in developing countries.[3,4] Early diagnosis and treatment with adequate dose with corticosteroid and other immunosuppressive drugs have improved mortality and morbidity in children.

This paper reports the rare case of JDM with clinical features, laboratory findings and treatment with its response.

Case description

A 12 year old female child, admitted to paediatric department of Rajshahi Medical College hospital, born to a non-consanguineous parents belonging to lower socioeconomic with uneventful antenatal, natal and neonatal period and normal development, immunized as per the EPI schedule of Bangladesh, presented with proximal muscle weakness of all four limbs with generalized muscle pain since one and a half years and photosensitive rash over the face, trunk and upper limbs since one year. She also had low grade irregular fever for same duration.

Figure1: A 12 years girl with proximal weakness and rash .

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There was no history of altered speech or difficulty in swallowing. She also deny history joint pain, oral ulceration and color change of fingers during cold exposure. She got treatment from homeopathic and kobiraz before admission.

On examination the girl was severely ill febrile and mildly pale. She had violacious rash over the both eyelid with periorbital edema (Heliotrope rash), Gottron's papules over metacarpophalangeal and proximal interphalangeal joints of both hands. There was no cutaneous ulceration and sign of arthritis. Muscle wasting was present over proximal muscle group and Power of proximal muscles (2/5) was less compared to distal muscles (4/5). In addition to this, child had a waddling gait and positive Gower sign.

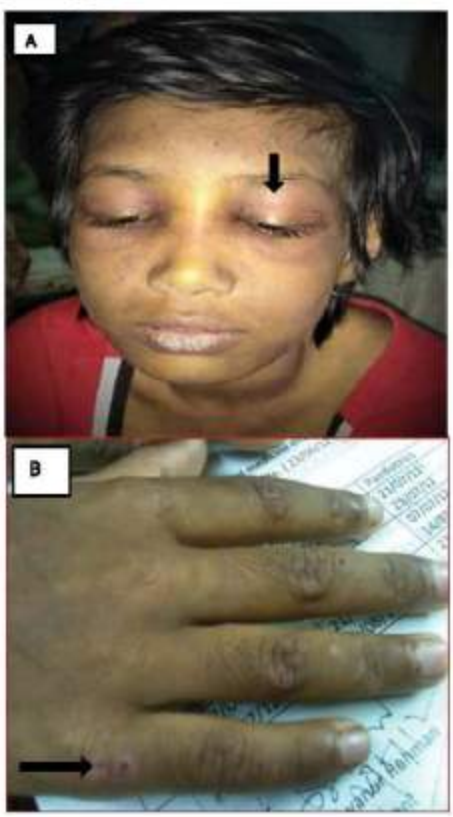

Figure2: A-Heliotrope Rash. B- Gottron's papules

Investigations included a complete haemogram, renal function tests, urine routine which were normal except mild normocytic normochromic anemia (Hb-10.5 gm/dl). ESR-60 mm in 1st hour, ANA positive but anti Ds DNA,Anti Jo-1, Anti Mi-2 were negative. Additional investigations included:
Creatine phosphokinase >13845U/L, alanine aminotransferase 176U/L, MRI of Muscle revels myositis and fascitis of proximal muscle . EMG showed features of myositis and NCS of crossed limbs were normal. These findings clinched the diagnosis of Dermatomyositis.

Figure 3: MRI of Right thigh muscle revels myositis and fasciitis of quadriceps muscle.

Differential diagnosis considered was collagen vascular disease like Systemic Lupus Erythematosus (SLE), but SLE was ruled out based on American College of Rheumatology 1997 revised criteria. Anti DS DNA was also negative.

Diagnosis of juvenile Dermatomyositis was confirmed since the diagnostic criteria included classic rash. Heliotrope rash of the eyelids, Gottron papules, plus 3 of the following:
1) Symmetric and proximal muscle weakness
2)Muscle enzyme elevation:(1 or more) Creatine kinase, Alanine transaminase,
3) Characteristics EMG findings

After proper counseling child was given prednisolone at 2 mg/kg/day. Weekly oral Methotrexate 1 mg/kg was also used as a steroid sparing agent. Folic acid was given with Methotrexate, started at a dose of 1 mg daily to reduce toxicity and side effects of folate inhibition.

Follow up after 4 weeks of treatment muscle weakness was reduced and skin rash were mostly disappeared . Muscle enzyme returned to normal level after 8 weeks of treatment. Then we started to tapering the dose of prednisolone. At present overall condition of the child is better with normal muscle power.

Discussion

JDM is an autoimmune connective tissue disease occurring in children less than 16 years old.[5] The etiology of JDM is multifactorial, based on genetic predisposition and an unknown environmental trigger. Human leukocyte antigen (HLA) alleles such as B8, DRB1*0301, DQA1*0501, and DQA1*0301 are associated with increased susceptibility to JDM in selected populations. Maternal microchimerism may play a part in the etiology of JDM by causing graft-versus-host disease or autoimmune phenomena.[2]

Children with JDM present with either rash, insidious onset of weakness, or both. Fevers, dysphagia or dysphonia, arthritis, muscle tenderness, and fatigue are also commonly reported at diagnosis. Rash develops as the first symptom in 50% of cases and appears concomitant with weakness only 25% of the time. Heliotrope rash of eyelids and Gottron's papules are two most important pathognomic findings of JDM present 66-83 % and 57-91% of patient respectively.[2]

Diagnosis of dermatomyositis requires the presence of characteristic rash as well as at least three signs of muscle inflammation and weakness.

Diagnostic criteria of JDM[6] developed in 1975 by Bohan A and Peter are as:[6]

Classic rash: Heliotrope rash of the eyelids
Gottron papules
Plus 3 of the following:
Weakness Symmetric Proximal
Muscle enzyme elevation (≥1):
Creatine kinase
Aspartate aminotransferase
Lactate dehydrogenase
Aldolase
Electromyographic changes:
Short, small polyphasic motor unit potentials,Fibrillations
Positive sharp waves
Insertional irritability
Bizarre, high-frequency, repetitive discharges
Muscle biopsy: Necrosis, Inflammation

There are three patterns of disease in JDM.[7,8]

Monocyclic course, in which there is one disease episode that responds to standard treatment without relapse (approximately one-third of patients),

Polycyclic course with multiple remissions and relapses (approximately 3 percent).

Chronic persistent course, sometimes with persistent complications (approximately two-thirds)

An increased severity of muscle histo pathologic features was associated with an increased risk of a chronic persistent course (as judged by the need for a longer course of treatment), whereas presence of anti-Mi-2 autoantibodies was, perhaps, associated with a decreased risk.[9] Chronic continuous or polycyclic disease is predictive of a poorer outcome.[10,11] These patients are at increased risk for persistent pain, calcinosis, and disability.

Most complications from this disease are related to prolonged and severe weakness, including muscle atrophy, to cutaneous

calcifications and scarring or atrophy, and to lipodystrophy.[2] Children with acute and severe weakness are at risk for aspiration pneumonia and respiratory failure.[2]Crampy abdominal pain and occult GI bleeding may indicate bowel wall vasculitis and lead to ischemia, GI bleeding and perforation.[2] Cardiac involvement is rare but includes arrhythmias.[9] Malignacy is common in adult onset dermatomyositis but usually not associated with JDM.[2]

Advances in the treatment have improved mortality and morbidity rates in children with JDM. Early treatment may limit JDM to a monocyclic pattern.[12]At 7 yr of follow-up, 75% of patients have little to no residual disability, but 25% continue to have chronic weakness and 40% have chronic rash.[2]

The reported mortality rate has declined from greater than 30 percent in the 1960s[13], before routine glucocorticoid therapy was administered, to less than 2 or 3 percent in the 2000s with the advent of early combination immunosuppressive therapy.[8,10,14,15] So increased awareness should be made for early diagnosis and treatment for better outcome and to prevent long term complications

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