

A Rare Case of Juvenile Dermatomyositis (JDM)

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Abstract: JDM is a rare autoimmune vasculopathy affecting children and adolescents under the age of 18. A 5 year female child presented with myalgia, skin rash, abdominal pain and fever. On Observation Gottron papules, Heliotrope rash were present; EMG showed myopathic pattern. The Childs symptoms and clinical findings improved with corticosteroid and Methotrexate therapy. JDM is a rare vasculopathy, should be diagnosed and treated early to minimise disability and life-threatening complications.

Keywords: vasculopathy, myopathy, skin rash.

INTRODUCTION

Juvenile dermatomyositis is a rare auto immune vasculopathy of childhood that preferentially affects dermal and muscular vessels. By definition the onset of JDM is prior to age of 18 years [1]. The peak age of onset is between 4 year to 10 years. Male/female ratio is 2: 1. The incidence of JDM is 3 cases / million children / year without racial predilection. The etiology is not clear yet, but increased susceptibility to HLA alleles such as HLAB10301, HLAB8, HLADQA10501 and HLADQA301 has been reported suggesting genetic susceptibility [5]. Polymorphisms of TNF alpha and IL 1 receptor is identified as additional risk factors. Environmental factors such as bacterial and viral infections (group A beta haemolytic streptococci enterovirus, coxsackie virus) act as important triggers [2].

CASE REPORT

A 5 year female child presented to us with myalgia and non-itchy maculopapular rash all over body for 1.5 month with past history of intermittent low grade fever for 10 days 1.5 months back. The patient also developed gradual swelling around eyes and lips for last 7 days. Examination revealed HR-98/minute, RR-16/minute, BP-124/70 mm of hg, Gottrons papules over small joints of both hands, Heliotropes rash around eyes with facial puffiness, mucosal ulceration and maculopapular rash all over body. Muscle tenderness

was significant. Rest of systemic examination was normal. Investigations revealed Hemoglobin - 11.1gm/dl, Total leukocyte count-6410/mm³ with Neutrophil-80%, Lymphocyte-18%, serum urea-20.4mg/dl, serum creatinine-0.38 mg/dl, serum albumin-2.88 mg/dl, serum sodium-137.44 mmol/dl, potassium-4.08 mmol/dl, calcium-1.2 mmol/dl, serum bilirubin total- 1.06 mg/dl , direct-0.53 mg/dl, ALT-158 IU/L, AST- 610 IU/L, ALP 330 U/L, Urine albumin- nil, pus cell1-2/hpf, RBC -nil. Serum CPK 3960 U/L, ESR -7mm, ANA Profile normal and EMG showing myopathic pattern , Ultrasound of abdomen revealed hepatomegaly. Based on clinical appearance, laboratory and imaging data the diagnosis of juvenile dermatomyositis (JDM) was made. Corticosteroid therapy with intravenous methylprednisolone (30 mg/kg body weight per day) was initiated and was administered for 5 consecutive days followed by oral prednisolone (2 mg/kg per day). The patient was discharged with oral prednisolone and Methotrexate. The therapy was well tolerated without apparent side effects. The skin lesions completely resolved. The patient has now been followed up after 6 months. At his last presentation, skin manifestations had disappeared and muscle enzymes were within normal range. Continuation of this treatment is planned for at least 1 year.



Fig 1: Showing Heliotrope Rash



Fig 2: Showing Gottron Papule

DISCUSSION:

Diagnostic criteria for JDM are currently still based on those established by Bohan and Peter in 1975 [3], which include a characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic EMG changes. The presence of 3 of these criteria characterizes definite JDM, whereas the prevalence of 2 criteria makes the diagnosis probable.

However, early diagnosis is often hampered by the nonspecific nature of the initial signs of JDM, such as fatigue, fever, weight loss, irritability, myalgia, and arthralgia. Identification of characteristic skin lesions may help establish an early diagnosis. Typical cutaneous lesions include a characteristic periorbital heliotrope rash (figure1), facial malar rash, Gottron papules (figure2) and nailfold changes that may present as peri ungual infarcts. Nailfold capillaroscopy shows reduced capillary density, capillary dropout, branching, and dilatation [4]. In addition, nonspecific eruptions on the extremities and mouth, skin ulcers, lipodystrophy, psoriasiform scalp dermatitis, and limb edema have been described. Particular attention should be given to dystrophic calcification that occurs in up to 30 percent of cases [5] and may lead to longterm disabilities and ulceration.

Myopathy, mostly affecting the proximal muscles, is present in about 95 percent of dermatomyositis cases. Myalgia may precede skin rashes, thereby posing a diagnostic challenge. With respect to the invasive nature of muscle biopsy and electromyography, MRI is frequently used as an alternative diagnostic procedure in pediatric patients [6]. In the international consensus survey of the diagnostic criteria for JDM, MRI was appreciated as one of the most important diagnostic methods to be added to the revised criteria.

Autoantibodies such as ANA, antiMi2 or antiJo1 can only be detected in a minority of JDM cases and were negative in our patient.

Systemic glucocorticosteroids are the mainstay of therapy; They are administered orally (up to 2 mg/kg per day of prednisolone) or as intravenous pulses (usually 30 mg/kg per day of methylprednisolone). Therapy is continued until there is improvement of clinical and laboratory parameters. Corticosteroids may be combined with Methotrexate (15 mg/m² once weekly) to allow reduction of cumulative corticosteroid doses and associated side effects. Other therapeutic options include combination of (methyl) prednisolone

with Azathioprine, Cyclosporin or Hydroxychloroquine. High dose intravenous Immunoglobulins have been administered as adjuvant therapy in steroid resistant cases. A few case reports have shown disease control by the B lymphocyte depleting antibody Rituximab. Adjunctive measures such as physiotherapy and the use of photo protective agents may contribute to a better outcome of the disease. In addition, calcium and vitamin D should be administered as co medication in situations of long term corticosteroid use.

CONCLUSION:

JDM is a rare but serious autoimmune vasculopathy of childhood that should be diagnosed and treated as early as possible to avoid disability and to minimize life-threatening complications such as systemic calcification, sepsis, hemorrhage, and infarction.

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