ABSTRACT

A 6-year-old girl presented with irregular fever for last two months and difficulty in standing from sitting position for last 1½ months. She had pathognomonic heliotrope rashes on both eyelids, Gottron’s papules in proximal interphalangeal and metacarpophalangeal joints of both hands and papules on elbow, knee and ankle joints. She had elevated serum muscle enzyme levels and electromyogram was in favor of juvenile dermatomyositis (JDM). She is now under steroid treatment and showing signs of improvement.

Keywords: Juvenile dermatomyositis, heliotrope rashes, Gottron’s papule, proximal myopathy, MRI

CASE REPORT

A 6-year-old girl presented with irregular fever for last two months and difficulty in standing from sitting position for last 1½ months. She had pruritic, erythematous rashes on face, trunk and extremities. She was treated for malaria and enteric fever elsewhere and was referred to our hospital as the symptoms did not improve.

Her anthropometric measures were normal for age, she had shiny erythematous papules on proximal interphalangeal joints, metacarpophalangeal joints (Fig. 2), elbow, knee and ankle joints of both sides suggestive of Gottron’s papule. There were erythematous rashes on face and bridge of nose along with violaceous rashes on both eyelids consistent with heliotrope rashes (Fig. 1). There was weakness of proximal muscles of legs with positive Gower sign. There was no neurodeficit and deep tendon jerks were normal.

Investigations revealed normal hemogram, erythrocyte sedimentation rate (ESR) was 70 in first hour, rheumatoid factor, antinuclear antibodies (ANA), lupus erythematosus cell phenomena were negative, C-reactive protein (CRP) level was elevated (>0.6 mg/dl). Creatine phosphokinase (CPK) was 72 U/l (21-215), alanine transaminase (ALT) = 63 (normal 3-65), aspartate transaminase (AST) = 48 (15-37), lactate dehydrogenase (LDH) = 379 (100-190). Electromyogram (EMG) revealed denervation along with myopathic changes characteristic of juvenile dermatomyositis (JDM). Magnetic resonance imaging (MRI) of muscles revealed hyperintense signals in bilateral gluteal muscles. Muscle biopsy from biceps was normal.

She was diagnosed as a case of JDM and treated with oral prednisolone 2 mg/kg/day. Follow-up after three weeks showed improvement in muscle power and decrease in rashes with resumption of normal day-to-day activity. However, at second follow-up, she had an ulceration around left elbow joint with whitish discharge, which was diagnosed as calcinosis cutis.

DISCUSSION

JDM is a rare autoimmune vasculopathy of childhood that preferentially affects dermal and muscular vessels. By definition, the onset of JDM is prior to the age of 18, whereas the average onset is in the 7th to 8th year of life, with a slight preference for the female gender.1 The disorder is rare, with a prevalence of 1-3.2 cases per million in children.2

While the etiology of JDM remains unclear, the working hypothesis is that this involves environmental triggers, immune dysfunction and specific tissue responses (in particular those of muscle, skin and small vessel endothelium) in genetically susceptible
individuals. Environmental factors such as bacterial and viral infections (Group A β-hemolytic streptococci, Enterovirus, Coxsackie virus) and exposure to UV light have to be considered as important trigger factors. Anecdotal reports of JDM onset after exposure to drugs, biological therapies, vaccines also exist. At present, the most widely used set of criteria remain those defined by Bohan and Peter in 1975, which require the presence of one of the characteristic rashes, combined with three of the following features, for definite JDM: Symmetric proximal muscle weakness, raised serum muscle enzymes (which may include creatine kinase (CK), transaminases, LDH and aldolase) and abnormal findings on muscle biopsy and EMG. The presence of the rash with two of these features make a diagnosis of probable JDM. These criteria, now over 30 years old, no longer reflect modern diagnostic investigation of suspected cases of JDM. Although MRI findings are not part of the Bohan and Peter criteria, MRI is now widely used to detect typical inflammatory changes in proximal muscles, and quantitation of such changes has been shown to correlate with disease activity. Myopathy, mostly affecting the proximal muscles, is present in about 95% of dermatomyositis cases. 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. Approximately 50% of children present with rash as their initial symptom and 25% present with weakness as their first symptom. 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 (present in more than two-thirds of patients), facial malar rash, Gottron papules (livid scaly plaques on the extensor surface of joints), and nailfold changes that may present as periungual infarcts. Calcinosis is common in JDM. Our patient developed calcinosis around elbow joint, two months after starting treatment. Blood tests to measure specific markers of muscle inflammation should be ordered including creatine kinase, LDH, aldolase, ALT and AST. Because muscle inflammation is patchy, EMG and muscle biopsy is not always diagnostic. MRI scans have become more

Figure 1. Heliotrope rashes.

Figure 2. Gottron’s papules on hand.

Figure 3. Calcinosis cutis in elbow joint.
widely used and can be helpful in choosing the correct muscle biopsy site.\textsuperscript{8}

The course of the disease is difficult to predict, but it is known to have a long course with remissions and exacerbations or, in some cases, a chronic course with a severe debilitating morbidity. Systemic glucocorticosteroids are the mainstay of therapy; they are administered orally (upto 2 mg/kg/day of prednisolone) or as intravenous pulses (usually 30 mg/kg/day of methylprednisolone). Therapy is continued until there is improvement of clinical and laboratory parameters.\textsuperscript{6}

The incidence of calcinosis is decreasing but, for some patients, it remains a debilitating and disfiguring problem. Many treatments have been tried, including diltiazem, aluminum hydroxide, probenecid, bisphosphonates and local corticosteroid injections, amongst others. No treatment has been proven to be effective. Calcinosis in many patients tends to regress over time (often years).\textsuperscript{9}

**Therapeutic Options for JDM\textsuperscript{3}**

Prednisolone, intravenous methylprednisolone, methotrexate are used as first-line therapy and adjunctive therapies to these are hydroxychloroquine, physical therapy, photoprotective measures, topical therapies for skin rashes, calcium and vitamin D for bone protection.

Second-line therapies include intravenous gammaglobulin, cyclosporine, azathioprine, while third-line therapies include cyclophosphamide, mycophenolate mofetil, tacrolimus, rituximab and anti-tumor necrosis factor alpha agents.

Combinations of the above agents as first-line therapies are among those most often used in the initial treatment of JDM, whereas second- and third-line therapies are most often used in the treatment of refractory patients, patients considered to have severe features, or patients with unacceptable medication toxicities.

**SUMMARY**

JDM is a very rare autoimmune disorder characterized by skin changes like Gottron's papule and heliotrope rashes along with proximal myopathy. MRI is helpful in demonstrating muscle edema and choosing the site of muscle biopsy. The disease has a long course with remissions and exacerbations. Steroid therapy is the cornerstone in the management of JDM.

**REFERENCES**