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JUVENILE DERMATOMYOSITIS : A CASE REPORT

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Summary

Juvenile Dermatomyositis, most common pediatric inflammatory myopathies, distinguished by characteristic rash and proximal, symmetric muscle weakness. A 7yrs old girl admitted to Dept. of Pediatric, Chittagong Medical College Hospital with difficulty in stand up from sitting and lying position for 3months, multiple erythematous papule in both upper limbs, face and extensor surface of the body for one and half months. On examination there is violaceous periorbital erythema(Heliotrope), raised papules overlying the metacarpal, interphalangeal, elbow and knee joints (Gottron's papules). There are also erythmatous papules on both upper limbs. On laboratory examination, serum creatine kinase raised, musle biopsy showed feature of Dermatomyositis, Electromyogram reveal proximal myopathy. Based on clinical appearance, laboratory data, the diagnosis of juvenile dermatomyositis was made and treated accordingly. After initial treatment patient condition improved. She also adviced to avoid sun exposure.

Key words

Juvenile dermatomyositis (JDM); heliotrope; gottron's papule

Introduction

JDM , most common pediatric inflammatory myopathies, distinguished by characteristic rash and proximal, symmetric muscle weakness[1]. The inflammatory pathway may be driven by the introduction of genetic predisposition with antigen stimulation and other environmental factors leading to disease. Genetic markers on chromosome 6, DQA1*0501 and DRB1*0301 associated with JDM susceptibility in the united states, while the DRB1*0501 locus more closely linked to cases with JDM in Asia . Environmental factors like bacterial and viral infections, excessive sun exposure may play a contributory role.

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The incidence of JDM is 3.2 cases/ million/ yr. The average age is 6.9yrs and ratio to girls to boys is 2.3:1[1]. Diagnostic criteria for JDM are currently still based on those established by Bohan and Peter in 1975[2], which include a characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes. The presence of 3 of these criteria characterizes definite JDM, whereas the prevalence of 2 criteria makes the diagnosis probable. In affected skin, the epidermis thins and the dermis demonstrates edema and vascular inflammation.

Case Report

A 7 yrs aged girl of non consanguineous parent from Tabalchary, Rangamati admitted to Chittagong Medical College Hospital for treatment of difficulty in stand up from sitting and lying position for 3months, multiple erythematous papule in both upper limbs, face and extensor surface of the body for one and half months. There is no positive family history, all the family members are in good health. On examination patient is well alert, co-operative, mild oedema on lower limbs. There is violaceous periorbital erythema(Heliotrope), raised papules overlying the metacarpal, interphalangeal, elbow and knee joints(Gottron's papules). There are also erythmatous papules on both upper limbs.

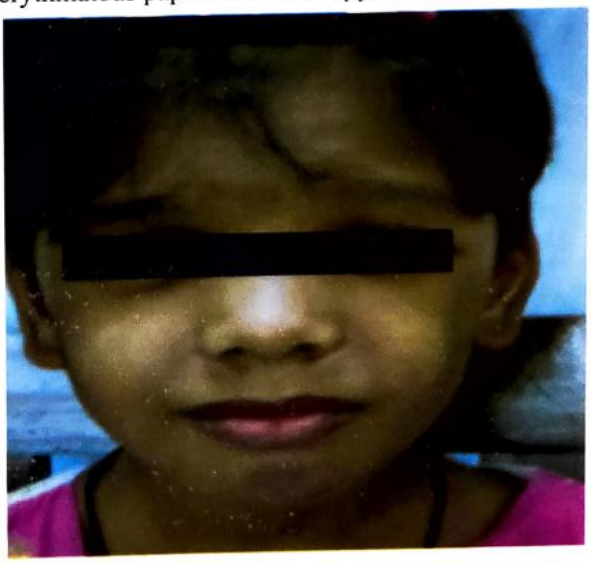

Fig 1: Violaceous periorbital erythema (Heliotrope rash)

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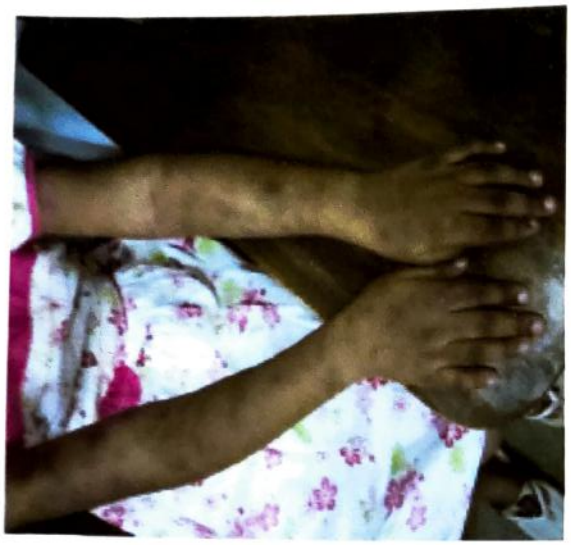

Fig 2: Symmetrical erythematous & bluish patches and plaques of both upper limbs

On laboratory examination Serum creatine kinase 910U/l (N:21-215 U/L), Serum aldolase 12.6U/L (N <7.6U/L). Serum calcium, Serum T3, T4, TSH, CXR and other laboratory investigations revel normal. On muscle biopsy section shows perifascicular atropy, necrosis of fibers and perivascular infiltration with lymphocytes. Focal calcification is also noted. EMG revels proximal myopathy. ANF and MRI was not done due to financial incapability. Based on clinical appearance, laboratory data, the diagnosis of juvenile dermatomyositis (JDM) was made. Pulse corticosteroid therapy with intravenous methylprednisolone was initiated and was administered on 3 consecutive days, then oral prednisone(2mg/kg) added, in addition, omeprazole (40 mg per day) was given. Serum level of anti-jo 1 antibodies one of the markers to detect interstitial lung disease was done, which was negative. The therapy was well tolerated without apparent side effects. Creatine kinase gradually improved. The patient has now been followed-up. At her last presentation, muscle power and skin manifestations improved and muscle enzymes were within normal range.

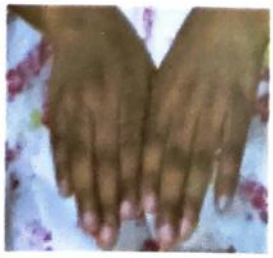

Fig 3: Violaceous papules overlying the metacarpal and interphalangeal joints (Gottron's papules) of both hands

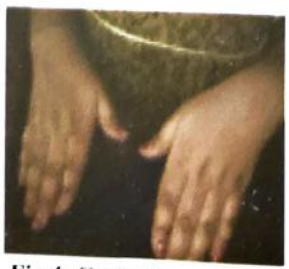

Fig 4: Gottron's papules disappeared after 3 months treatment of corticosteroids (Prednisolon 40 mg /day)

Discussion

Our patient was diagnosed as JDM, as her symptoms and signs fit the diagnostic criteria like characteristic skin eruption, symmetrical proximal muscle weakness, elevated muscle enzymes, pathological muscle histology, and myopathic electromyographic changes. The average age is 6.9yrs and ratio to girls to boys is 2.3:1. Our patient is 7yrs, female.

As fatigue, fever, weight loss, irritability, myalgia, arthralgia are initial nonspecific signs of JDM, early diagnosis is often difficult. Characteristic skin lesions include periorbital heliotrope rash that present in more than two-thirds of patients, facial malar rash, Gottron papules, and nailfold changes which may present as periungual infarcts may help to establish an early diagnosis. Nonspecific eruptions on the extremities and mouth, skin ulcers, lipodystrophy[3], psoriasiform scalp dermatitis[4], and limb edema have been described. Dystrophic calcification can occurs in up to 30 percent of cases[5], may lead to long-term disabilities and ulceration.

Another characteristic feature of JDM is myopathy, mostly affecting the proximal muscles, is present in about 95 percent of dermatomyositis cases; the existence of amyopathic dermatomyositis is controversial. Myalgia may precede skin rashes, thereby posing a diagnostic challenge[5,6]. MRI is used in pediatric patients as an alternative diagnostic procedure due to invasive nature of muscle biopsy[7]. Vasculopathy may affect the vessels of the GI tract, potentially resulting in malabsorption, ulceration, acute hemorrhage, and infarction. In respiratory tract, muscle weakness may lead to aspiration. Moreover, an increased incidence of interstitial lung disease has been reported. Therefore, GI symptoms and dyspnea should always be taken seriously and assessed clinically and radiologically[8]. JDM is not associated with malignancies as adult dermatomyositis, so corresponding assessments does not require. Autoantibodies such as ANA, anti-Mi-2 or anti-Jo-1 detected in a minority of JDM cases. If a patient with clinical complaints resembling dermatomyositis suffers from persistent arthritis, mixed connective tissue disease is an important item in the differential diagnosis. However, our patient did not complaint arthralgia. Histologic hallmarks of muscle biopsy are perifascicular atropy and occlusion of capillaries and arterioles, which is mediated by a primarily mononuclear cell infiltrate. Damage to the muscle is indicated by infarcted tissue, muscle fibres with central nuclei and increased fibrosis composed of type 1 collagen, which is associated with loss of range of motion.

Children with only mild cutaneous findings, normal immune and serological markers of disease activity, and a negative history of color blindness are administered hydroxychloroquine (maximum: 5mg/kg/day) with daily oral prednisone (1mg/kg)[1]. With evidence of minimal inflammation and muscle damage, oral prednisone (1-2mg/kg) may suffice. Children with extensive vasculopathy, high dose intermittent intravenous methylprednisone (30mg/kg/day for 3 days; maximum dose 1g/day) rapidly normalizes the muscle enzymes. After the initial pulse, lower dose of methylprednisolone may be required daily in extremely ill child. Weekly methotrexate (15-20mg/m2/wk) is started at the same time as methylprednisolone, along with oral folic acid (1mg/day). Cyclosporine (3mg/kg/day) helpful for persistent cutaneous involvement. A few case reports have shown disease control by the B lymphocyte-depleting antibody rituximab[9]. All children with JDM should avoid exposure to sun and also use sunscreen[1]. Vitamin D and calcium supplements sould be given to repair the osteopenia and to decrease the fequency of bone fracture.

Conclusion

JDM is most common pediatric inflammatory myopathies which can lead to life-threatening complications such as systemic calcification, sepsis, hemorrhage, and infarction. So to avoid disability and to minimize serious complications it should be diagnosed and treated as early as possible.

Disclosure

All the authors declared no competing interestes

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