ORIGINAL RESEARCH PAPER

Paediatric Medicine

CLASSICAL CASE OF JUVENILE DERMATOMYOSITIS : A CASE REPORT

KEY WORDS: Juvenile Dermatomyositis, Muscle weakness, Rashes, Myopathy, HLA

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ABSTRACT

Juvenile Dermatomyositis (JDM) is the most common idiopathic inflammatory myopathy in children. It is a systemic autoimmune vasculopathy characterized by muscle weakness and pathognomic skin rashes. It mainly affects skin and muscle but other organ systems can also be involved. Though most common myopathy, the incidence is 2-3 per 1 million children. Pathogenesis involves complement-mediated small vessel damage, type I interferon activation, and genetic susceptibility, with HLA alleles and myositis-specific autoantibodies contributing to risk and disease course.

INTRODUCTION-

Juvenile Dermatomyositis is the most common idiopathic autoimmune inflammatory myopathy in children. The pathogenesis is not clearly understood but it appears that children with genetic susceptibility to JDM (HLA-DQA1*0501, HLA-DQA80301, HLA-DRB1*0301) may have exposure to environmental triggers such as infection. It is a small vessel vasculopathy which typically affects skin and muscle but can also involve other organ systems such as GIT, joints, lungs and heart. Patients typically present with muscle weakness and characteristic skin rashes.

CASE REPORT-

We present a case of 9 years old female child bought with the complaints of rashes over multiple parts of body which first appeared on bilateral elbows and then progressed to involve the knees, face, neck and trunk which was initially erythematous macular type which changed to hyperpigmented lesions over a span of two weeks. Rash spared the palms and soles. The child also developed weakness one week after onset of rash which was proximal in nature which was followed by joint pain and swelling mainly involving large joints. The child also developed dysphagia, both to solids and liquids.

On examination, characteristic findings were noted, namely- On examination classical heliotropic rash, shawl sign, Gottron papules were observed. Proximal muscle weakness was demonstrated. Edema involving bilateral upper and lower limbs was noted.

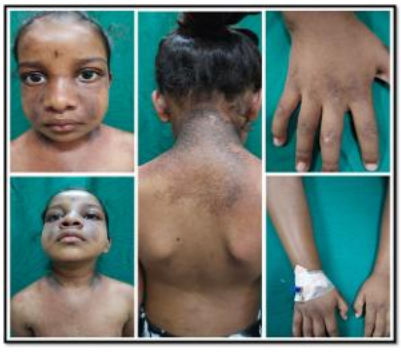

Figure 1- Demonstrating Heliotropic rash, Shawl sign, Gottron's papules, Edema of limbs

Investigations done showed elevated CPK, ESR. ANA profile was sent which was Negative. USG of bilateral knees showed joint effusion. Chest Xray, 2D ECHO and USG Abdomen and Pelvis showed normal study. MRI of bilateral thighs showed diffuse myositis. Myositis specific antibodies panel was strongly Positive for Anti-transcriptional intermediary factor 1 gamma.

The child was treated with IV Methylprednisolone (pulse therapy), switched to oral steroids along with Hydroxychloroquine, Methotrexate, Folic acid, Cyclophosphamide, Vit D & Calcium supplements and sunscreen application. Clinical improvement in the signs and symptoms was noted. Based on the clinical features and supporting lab evidence, the child was diagnosed as a case Juvenile Dermatomyositis.

DISCUSSION-

JDM, though rare is the most common inflammatory myopathy in children. The diagnosis is made on the basis of EULAR/ACR classification criteria for Idiopathic Inflammatory Myopathies. Differential diagnosis include other causes of myositis or myopathy such as infection related myositis, polymyositis, myasthenia gravis, GB syndrome, endocrinopathies (Addison syndrome, Cushing syndrome, hypothyroidism), SLE, Psoriasis, Dystrophic eczema. Steroids remain the mainstay of treatment. Other drugs used- Methotrexate, Hydroxychloroquine, Rituximab, Cyclosporine and Cyclophosphamide. Complications of the disease include calcinosis, GI bleeding, Cardiac involvement, Interstitial lung disease and medication related complications

CONCLUSION-

This case highlights a rare but the most common idiopathic inflammatory myopathy in children and the need for early recognition, multidisciplinary approach, the need for specific investigations for prognostication and aggressive treatment for better long term better prognosis in children.

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