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Juvenile Dermatomyositis and Posterior Reversible Encephalopathy Syndrome: A Rare Association.

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JUVENILE DERMATOMYOSITIS AND POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME: A RARE ASSOCIATION.

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INTRODUCTION

Juvenile dermatomyositis or JDM is a rare but debilitating pediatric disease that affects the proximal muscles and skin, as implied by its name. Although rare, it is the most common childhood idiopathic inflammatory myopathy [(1).] The estimated incidence is 3.2 cases per million children [(2).] The diagnosis is based on clinical signs and symptoms supported by investigations including, but not limited to, muscle biopsy, muscle enzymes and EMG studies. Bohan and Peter's criterion is most widely used for diagnosis. It includes the presence of Gottron's rash, raised muscle enzymes, proximal muscle weakness and suggestive findings on muscle biopsy [(3).] Treatment is done with a combination of steroid and immune-suppressing agents, mainly methotrexate [(4).]

This report illustrates a case of JDM that presented with Posterior Reversible Encephalopathy Syndrome (PRES) and vision loss which are unusual presentations among children. Only a handful of studies have been published in existing literature where children with JDM presented with neurological symptoms [(5).]

CASE PRESENTATION

An 8-year-old boy from Karachi was referred to a hospital with a 4-month history of diffuse erythematous rash, generalized body ache, and fatigue. He was bedridden for the past two months. On examination, the child was fully oriented in time, place, and person. Child was drooling, dysphonic, and had impaired swallowing. Power in all limbs were 2/5, proximal muscle weakness was present, and there was partial neck holding. There was generalized swelling and an erythematous rash all over the body, but no ulcers or calcinosis were present. Abdominal, cardiovascular, and respiratory examinations were normal.

Lab reports showed increased CPK of 2490 U/L, antinuclear antibody test was positive. Extractable nuclear antigen antibodies (ENA) panel was negative for systemic lupus erythematosus and juvenile idiopathic arthritis. Rheumatoid factor and anti-dsDNA antibody tests were negative. EMG studies were suggestive of irritable myopathy. MRI of quadriceps showed post-contrast enhancement of muscles, and biopsy from the left quadricep muscle was taken which showed perifascicular atrophy and necrosis, thus confirming the diagnosis of Juvenile dermatomyositis.
Management was initiated with nasogastric feeding and three doses of pulse Methylprednisolone (30mg/Kg), followed by subcutaneous Methotrexate (MTX) (15mg/m2) and hydroxychloroquine (HCQ) for 6 months. Once stable, the child was discharged on physiotherapy. After one week of discharge, he presented again in the emergency department with severe respiratory distress. Examination and chest x-ray suggested right-sided lung collapse (Figure 1) due to excessive secretions. He required suctioning and mechanical ventilation. The next day, he developed hypertension with blood pressure exceeding 95th centile for his age (120/78 mmHg), vision loss and seizures. Cerebral fluid analysis and cultures were negative. MRI brain (figure 2) showed asymmetrical bilateral signals in the parieto-occipital regions that were hyperintense on T2 and flair images. These findings were consistent with the diagnosis of posterior reversible encephalopathy syndrome (PRES). The child was weaned off from ventilator after six days. However, due to his continued breathing and swallowing problems, tracheostomy and gastrostomy was done.
Pharmacological treatment was escalated with two doses of I/V Immuno-globulin (2g/kg) followed by pulse Methylprednisolone for three doses. After discussion within a multidisciplinary team, MTX was replaced by intermittent Cyclophosphamide injections given every three weeks for six doses.

The child was followed fortnightly, and he showed gradual improvement in vision and muscle strength. However, he required excessive suctioning due to copious secretions. To mitigate this problem, injection

Botox was administered in both parotid glands. At the end of six months, remission in the disease was noted. Rash had disappeared, muscle power had become 4 in all limbs, neck holding was complete, and vision had returned. Muscle enzymes also significantly decreased. The child was again shifted to subcutaneous MTX and HCQ. In his last follow up, the child could talk, was taking a significant amount of oral feed but could not walk. Tracheostomy and gastrostomy tubes were removed, MTX and HCQ were continued along with physiotherapy.

DISCUSSION

A large retrospective study consisting of 116 JDM patients was published by authors from France, wherein the most common complications identified were related to gastrointestinal tract (11/116), respiratory failure (1 patient), cardiac complications (1 patient), thrombotic microangiopathy (2 patients). Only one child was found to have PRES [6]. We reported an unusual neurological complication of JDM where the vasculopathic nature of this disease can be appreciated.

PRES, also known as Reversible Posterior Leukoencephalopathy Syndrome (RPLS), is a clinico-radiologic condition that presents with headache, encephalopathy, seizures, and vision abnormalities. On imaging it is seen as reversible vasogenic subcortical edema, most commonly in part supplied by posterior cerebral artery. Generally, pathophysiology of PRES is linked to hypertensive encephalopathy [7]. While in JDM, complement system is responsible for causing PRES. Complement mediators attack the microvasculature and get deposited in endothelium of the vessels, leading to hypertension [8]. We postulate that vasculitis and complement mediated injury was responsible for PRES in our child and hence he responded to immunosuppressive treatment. JDM associated neurological complications have been treated by immunosuppressants in other cases as well (5,6). However, our patient despite of having severe course of illness responded to treatment and recovered with disease sequalae.

CONCLUSION

Dermatomyositis in children can present in various ways, and with severely debilitating associations. A timely diagnosis, appropriate treatment, and a high index of suspicion for associated conditions like PRES needs to be practiced for proper management.

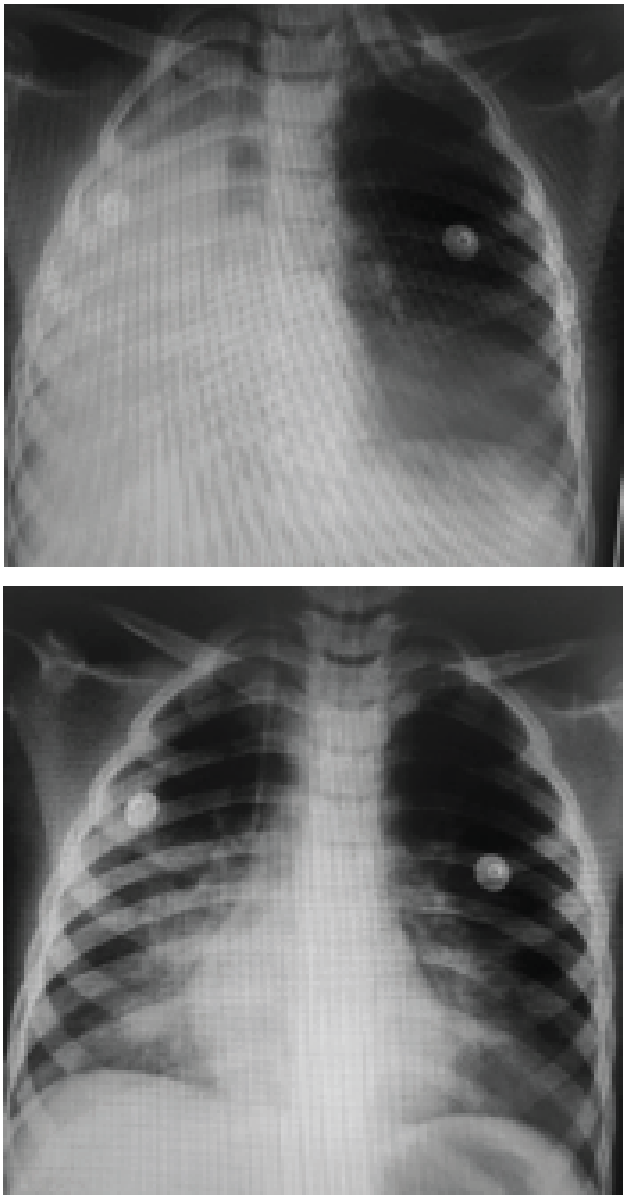

FIG.1: Chest X-rays. Top: Right lung collapse due to excessive secretions. Bottom: Re-expansion of lungs after suctioning and ventilation.

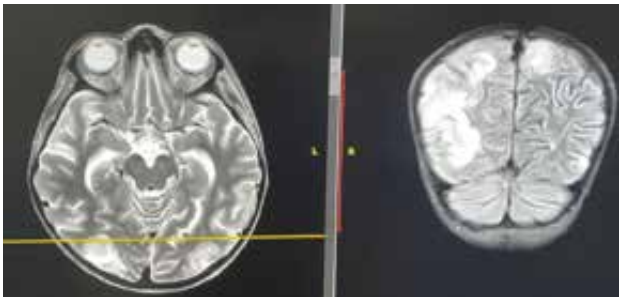

FIG.2: MRI Brain. Left: T2 weighted axial image showing asymmetrical hyperintense signals in parieto-occipital region. Right: Flair coronal image

showing asymmetrical hyperintense signals in bilateral parieto-occipital regions.

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