

Juvenile Dermatomyositis presenting with Anasarca

Juvenile dermatomyositis (JDM) is a multisystem disease characterized by non-suppurative inflammation of striated muscles with characteristic cutaneous findings in the form of erythematous (heliotrope) periorbital rash(1). The classical features of disease include proximal muscle weakness, dermatitis associated with constitutional symptoms and edematous and indurated muscles. Localized edema of muscle is a common feature but anasarca as a presenting feature is exceedingly rare(2).

An 8-year-old female child presented with irregular fever, anorexia, myalgia and generalized arthralgia. It was associated with muscle weakness and progressive deformity of both elbow and knee joints making her bedridden. Patient received irregular treatment prior to admission. On admission child was febrile, had mild pallor, generalized nonpitting edema of upper and lower extremities and facial puffiness. There was ascites and mild hepatomegaly. Neurological examination revealed grade III muscle power in all four limbs, muscle atrophy with flexion contractures at elbows and knees. Plantar reflex was flexor. The joint mobility was limited due to flexion contractures and the movements were painful. There was no joint swelling but there was tenderness and induration of muscles in both extremities. There was an erythematous periorbital violaceous rash. Rest of the systemic examination was normal.

Investigations revealed Hb 11g%, TLC 11, 300, DLC P80, L16, M2, E2, ESR-14 mm Urea, Creatinine, Bilirubin, SGOT, SGPT,

Alk Phosphatase and electrolytes were normal. Blood culture sterile. CRP-positive. RA factor - negative. ANA-negative. Total serum proteins- 7.8, (5.5-8.0 g/L), serum albumin-2.8, CPK-220 units/L (55-170 U/L), LDH-2033 units/L (150-500U/L). Urine albumin - nil (on repeated examinations), 24-hrs urinary protein-normal. Urine culture - sterile. USG abdomen revealed mild hepatomegaly, ascites and mild pleural effusion. Kidneys - normal in size and echotexture. (An USG repeated 15 days later revealed no free fluid). EMG revealed MUAP Duration and Amplitude reduced in Deltoid R, Biceps Brachii R and Tibialis Ant R. Muscle biopsy revealed infiltration with inflammatory cells and scattered areas of degeneration and necrosis.

The diagnosis of juvenile dermatomyositis was based on clinical features, muscle biopsy and EMG findings. The patient was treated with prednisolone 2 mg/kg/24 hr. Oliguria resolved within a few days and pedal edema subsided a little but weakness and non pitting edema of both upper and lower extremities persisted. Her general well being and appetite improved in 5 weeks of therapy but later on she developed severe mucosal ulcers, dysphagia and fresh bleeding per rectum. Patient died due to septicemia and disseminated intravascular coagulation.

JDM presents characteristically with proximal muscle weakness associated with periorbital heliotrope rash(1,3). Localized muscle edema is commonly encountered but generalized edema is extremely uncommon (2). In our case generalized non pitting edema was presenting feature along with muscle weakness and arthralgia. The absence of clinical features and laboratory parameters that ruled out any other cause of anasarca suggests that anasarca in this case was due to dermatomyositis.

Generalized edema in association with JDM is infrequently reported(2). Such patients respond poorly to oral steroid therapy. The exact mechanism of generalized edema in JDM is not known. Autoantibodies are believed to play an important role in pathogenesis. The pathological hallmark is an immune complex vasculitis - the damage to endothelium caused by complement C5b-9 attack complex(4,5). It is suggested that as a sequel to diffuse and widespread capillary endothelial damage there is increase in capillary permeability in muscles leading to generalized edema.

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