AUTOIMMUNE CONNECTIVE TISSUE DISEASES

VARYING PRESENTATIONS OF JUVENILE DERMATOMYOSITIS: SERIES OF 30 CASES

Sandipan Dhar [(1)]

Institute Of Child Health, Hospital, Kolkata, India [(1)]

Introduction: Juvenile dermatomyositis is a rare complex autoimmune disorder of childhood primarily affecting the proximal muscles and skin. Classical skin findings include heliotrope rash, gottron papules, shawl sign and nail fold capillary changes. Apart from the above classical findings childhood dermatomyositis can present with unique skin changes like calcinosis cutis, skin ulcers, panniculitis, lipodystrophy and rarely can be associated with other autoimmune disorders. As compared to adults, juvenile dermatomyositis is not associated with malignancy.

Objective: The aim of the study was to review the varied cutaneous manifestations of juvenile dermatomyositis presenting at a tertiary care children hospital.

Materials and Methods: All children less than 18 years presenting at department of pediatric dermatology at our institute from January 2005 through July 2018 with clinical diagnosis of juvenile dermatomyositis was included in the study. All the demographic details, clinical features and laboratory investigations were collected on a predesigned proforma.

Results: During the study period 30 children presented with clinical features suggestive of juvenile dermatomyositis. The study population included 8 males and 22 female children. Age group of presentation was between 3 years and 15 years. Heliotrope rash, gottrons papule and periungual telangectasia was seen in 22 children, calcinosis cutis was seen in 15 children, linear excoriation was seen in one child, ichthyosis was seen in 5 child, skin ulcers and atrophic scars was seen in 7 children, palmar hyperkeratosis was seen in one child, panniculitis and lipodystrophy was seen 5 children. One child had flagellate erythema or zebra stripes like macules on trunk and extremities. Three children in our study had pansclerotic morphea with dermatomyositis. Muscle weakness was seen in all children.

Conclusions: Juvenile dermatomyositis presents with classical features along with some rare presentation like extensive calcinosis cutis, lipodystrophy, panniculitis and flagellate erythema.