

## Clinically Amyopathic, Poikilodermatous Dermatomyositis in a Male Child: Sporadic presentation of an otherwise common entity in a dermatology set-up

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### Case Report

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### Article History

Received: 22.12.2017

Accepted: 27.12.2017

Published: 30.12.2017

### DOI:

10.21276/sjams.2017.5.12.67



**Abstract:** Dermatomyositis is classified as an idiopathic inflammatory myopathy, mainly affecting the skin and the skeletal muscle; with a peak incidence in 5<sup>th</sup>-6<sup>th</sup> decade of life and twice as high in females. We present a case of juvenile onset (< 18 years) dermatomyositis in a 10 year old, male child with suggestive cutaneous manifestations but clinically normal muscle function with an infrequent histological variant.

**Keywords:** dermatomyositis; juvenile onset; normal muscle function.

### INTRODUCTION

Dermatomyositis (DM) is an autoimmune disorder, at one spectrum of which myopathy predominates and at the other end, skin disease is prominent with minimal or absent myositis (amyopathic DM) but deranged muscle testing (hypomyopathic DM). Clinically amyopathic DM (CADM) representing approximately 20% of all cases of DM is a functional designation used to refer to either amyopathic or hypomyopathic DM with cutaneous manifestations of DM in the absence of clinical signs of muscle involvement [1].

### CASE REPORT

We report a case of a 10 year old, school going, male child, presenting with the chief complaints of multiple, asymptomatic, skin to pink coloured lesions over the face, upper trunk and the extremities since 8 months.

There was no history of fever, dyspnoea or dysphagia or use of any medications in the recent past. On examination, there were multiple, well defined, flesh coloured, variably shaped and sized, atrophic papules and plaques, a few with an erythematous margin, present over the chin, the scapular region, dorsa of both hands corresponding the metacarpophalangeal and proximal interphalangeal joints (Gottron's papules), the elbows, and the knees bilaterally (Fig 1-3). Heliotrope rash was evident as violaceous macular erythema present symmetrically involving the periorbital region (Fig.4). Hypertrichosis of knees and nail fold telangiectasias and ragged cuticles were also present.

On muscle testing, clinically, the tone of the proximal muscles (upper arms and thighs) was normal

with grade 5 muscle power. However, serum LDH levels were raised (749U/L) than the upper limit of normal range (313-618U/L) with normal CPK levels (23 U/L) and negative ANA (antinuclear antibodies) profile. Skin biopsy (taken from two sites, one from the lesion on the chin and one from the dorsum of left hand) findings were consistent with changes of early, poikilodermatous DM with orthokeratotic stratum corneum covering slightly atrophic epidermis. Epidermis revealed intercellular edema and basal cell vacuolization with thickening of the basement membrane (further confirmed on PAS staining). Papillary dermis showed foci of melanin incontinence and superficial to deep dermal, mild to moderate, perivascular, periadnexal lymphohistiocytic inflammatory infiltrate (Fig 5).



**Fig-1**



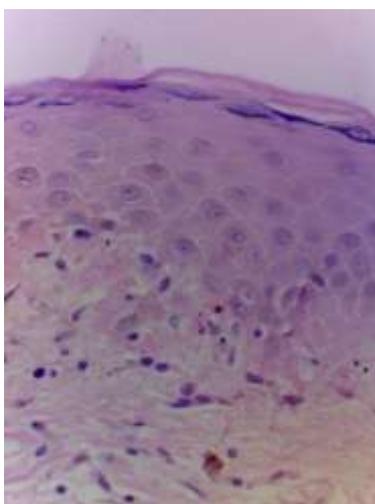
**Fig-2**



**Fig-3**



**Fig-1-4:** showing (left to right) heliotrope rash, atrophic papules and plaques (gottron's papules) over the dorsa of hands, back and the knees



**Fig-5** Photomicrograph at H and E, × 40 showing poikilodermatous variant with basal cell vacuolization, basement membrane thickening and melanin incontinence in the dermis

**DISCUSSIONS**

Bohan and Peter classification is commonly employed to the diagnosis of dermatomyositis which follows as in table-1 [2]. In addition, Sontheimer defines amyopathic DM (ADM) as a subset characterized by biopsy-confirmed hallmark cutaneous manifestations of classic DM occurring for 6 months or longer with no clinical evidence of proximal muscle

weakness and no serum muscle enzyme abnormalities. Exclusion criteria for ADM includes: (i) treatment with systemic immunosuppressive therapy for two consecutive months or longer within the first 6 months after skin disease onset; and (ii) the use of drugs known to be capable of producing DM-like skin changes (e.g. hydroxyurea) [3]

**Table-1: Bohan and Peter classification of dermatomyositis**

Characteristics	Diagnosis
1) Symmetrical proximal muscle weakness	Definitive: 5 plus any three of 1-4
2) Muscle biopsy suggestive of myositis	Probable: 5 plus any two of 1-4
3) Elevation in serum skeletal muscle enzymes	Possible: 5 plus any one of 1-4
4) Characteristic electromyography pattern of myositis	
5) Typical skin rash	

DM occurs in children although it is much rarer in this age group with an incidence of around two cases per million children under 16 years per year, with a median age of onset of 6.8 years, with female preponderance in children being greater at 5:1. [4] Most patients with DM survive but delay in diagnosis may have an impact on children's growth with respect to

both height and weight. This effect on growth can possibly be attributed to cachexia associated with tumor necrosis factor alpha, a cytokine known to play a role in the inflammatory response of JDM and more likely, to decreased gastrointestinal absorption of food, nutrients and oral prednisone secondary to systemic vasculopathy of JDM [5]. Therefore, the key to a favourable outcome

in juvenile DM is early diagnosis and prompt, aggressive immunosuppressive treatment [6].

Despite striking clinical signs in the skin, the dermatopathology of DM is often subtle. A lichenoid tissue reaction with vacuolar changes in the basal layer and occasional Civatte bodies is typical. In acute DM, the changes resemble those of subacute lupus erythematosus. In rarer poikilodermatous DM there is epidermal atrophy, dilatation of superficial vessels and melanin incontinence[7].

For typical cases of JDM, high dose corticosteroids (oral or intravenous) alone or with methotrexate has become the standard treatment regimen[8]. We started our patient on systemic corticosteroids (prednisolone 0.5 mg/kg/day) and topical tacrolimus ointment 0.03% for local application over the lesions. Steroid should be gradually tapered with gradual clinical improvement [8].

### **CONCLUSION**

Rare and severe form manifesting in children and in males, with infrequent poikilodermatous histological variant makes it important to note such a possible case since late diagnosis or delayed, inadequate treatment can result in continued increasing inflammation with potential systemic damage.

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