Case Report

Juvenile Onset Clinically Amyopathic Dermatomyositis Presenting as Calcinosis Cutis

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Abstract

Dermatomyositis (DM) is an autoimmune inflammatory disorder characterized by involvement of muscles and skin. Classical dermatomyositis (CDM) patients display the hallmark cutaneous manifestations of dermatomyositis (DM), proximal muscle weakness, and laboratory evidence of myositis. Rarely, DM presents with cutaneous features of the disease without muscle involvement for a period of more than six months. Such cases are classified into a category of clinically amyopathic dermatomyositis (CADM) which includes amyopathic DM and hypomyopathic DM. We present a case of a 14-year-old child who presented with calcinosis cutis and cutaneous findings suggestive of the disease without muscle involvement. The child was diagnosed as a case of juvenile onset CADM.

Key words*: Dermatomyositis; Children; Calcinosis cutis*

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Introduction

Dermatomyositis (DM) is an autoimmune inflammatory disorder characterized by muscle manifestations (i.e., proximal muscle weakness, elevated serum levels of enzymes derived from skeletal muscle, myopathic changes by electromyography, and muscle biopsy evidence of inflammation) as well as a variety of cutaneous manifestations such as heliotrope rashes and Gottron's papules. Clinically amyopathic dermatomyositis (CADM) as a distinct entity was first proposed by Sontheimer[1] and currently it is used to refer to either amyopathic DM or hypomyopathic DM. Sontheimer defined amyopathic DM as a subset of DM 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.[1] We present a case of a 14-year-old male who presented with chief complaints of calcified lesions over both hands and was subsequently diagnosed as a case of amyopathic DM.

Case report

A 14-year-old male presented to us with lesions over both hands for last seven months which were progressively increasing in size (Fig 1). There was no history suggestive of muscle involvement, Raynaud's phenomenon, photosensitivity, joint pain, oral ulcer or trauma. No other systemic complaints or significant prior medical history were present.

On general physical examination no abnormality was found. Both palms and flexor aspect of left wrist showed multiple skin colored to erythematous hard, noduloplaques with chalky discharge from two lesions (Fig 1). Over bilateral interphalangeal joints and metacarpophalangeal joints, elbows and knees, there were multiple skin colored to violaceous indurated papuloplaques (Fig 2, 3). Over lower trunk

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and posterolateral aspect of bilateral buttocks, there were diffuse violaceous plaques with interspersed hypopigmented macules and patches and follicular hyperkeratosis (Fig 4). Over bilateral upper eyelids, a lilac patch was appreciated (Fig 5). According to relatives they had noticed these lesions two years back, but as they were asymptomatic they never sought any medical advice. No nail or mucosal changes were seen. Motor examination was within normal limit.

Complete blood counts, renal function test and serum calcium were within normal range. Anti-nuclear antibody (ANA) was positive in low titre. Radiography of hands showed calcium deposits in subcutaneous tissue (Fig 6). Muscle enzymes creatinine kinase, lactate dehydrogenase and aldolase were within normal range. Electromyography (EMG) was normal. USG of whole abdomen and chest X-ray were normal. Histopathological examination of skin showed moderately dense superficial and mid-perivascular lymphocytic infiltrate with abundant mucin in reticular dermis (Fig 7). The epidermis showed mild irregular hyperplasia and focal interface change with occasional necrotic keratinocytes. The granular layer was slightly thickened and stratum corneum shows foci of parakeratosis. Findings were consistent with dematomyositis. In view of clinical and laboratory findings a final diagnosis of CADM of amyopathic type was made.

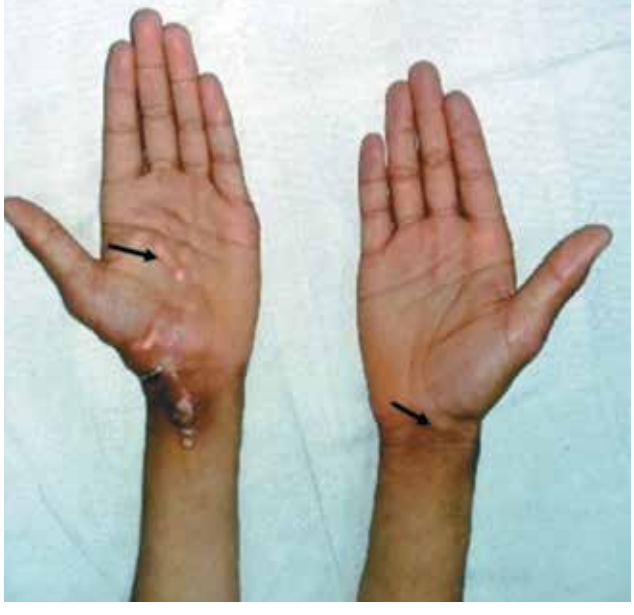

Fig 1. Calcinosis cutis over both palms and flexor aspect of left wrist

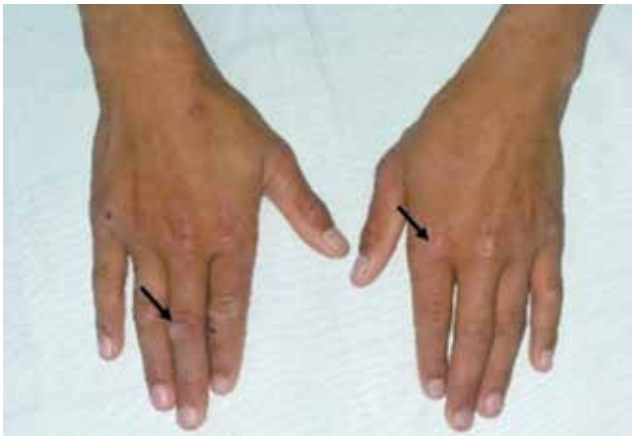

Fig 2. Multiple violaceous papules (Gottron's papule) over IP and MCP joint

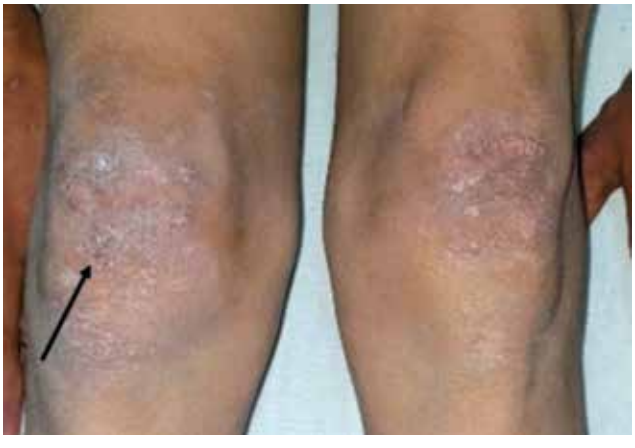

Fig 3. Violaceous plaque over both knees

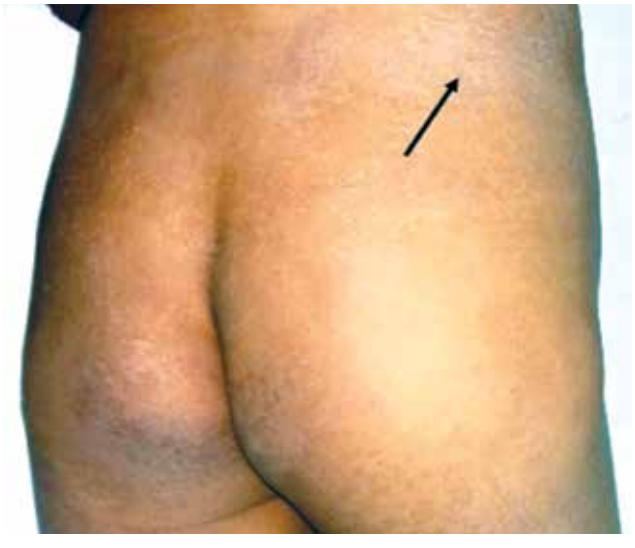

Fig 4. Symmetrical distribution of violaceous plaque with insterspersed hypopigmented macules and patches on trunk

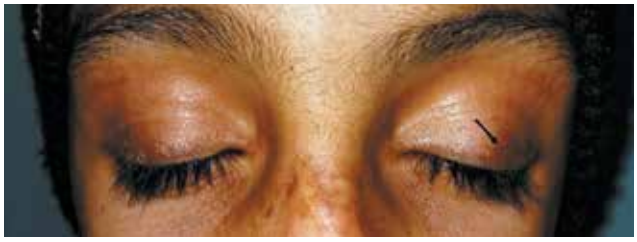

Fig 5. Symmetrical lilac patch over both upper eyelids (Heliotrope rash)

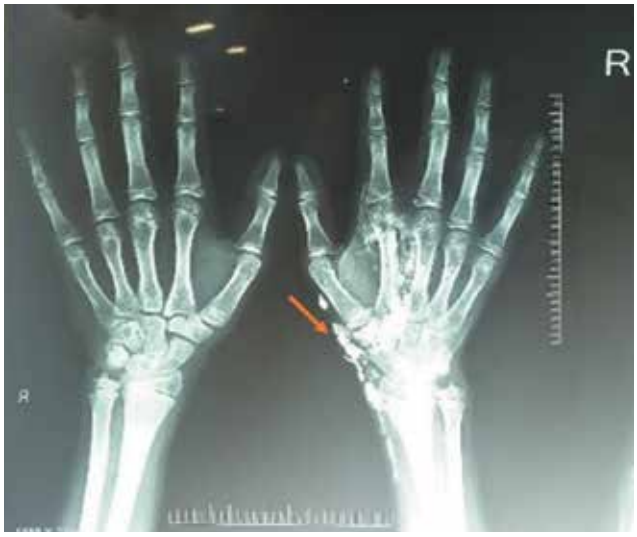

Fig 6. X-ray of both hands revealing calcinosis cutis

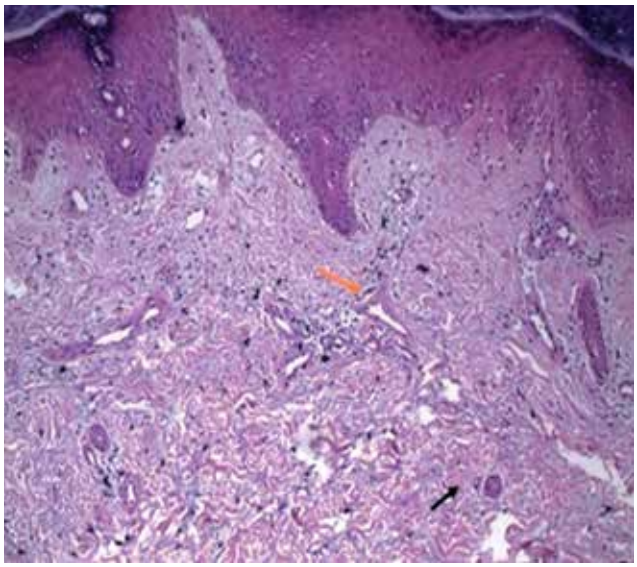

Fig 7. Biopsy shows moderately dense superficial and mid–perivascular lymphocytic infiltrate (red arrow) with abundant mucin in reticular dermis (black arrow)

Discussion

Dermatomyositis (DM) is a progressive autoimmune condition characterized by inflammatory skin changes and muscle weakness. Clinically amyopathic dermatomyositis (CADM) is a rare variant seen in 20% cases.[2] The diagnostic criteria of CADM as given by Euwer and Sontheimer[3] in 1991 are cutaneous changes pathognomonic for DM with Gottron's papule as the essential criteria. Their features with skin histopathology were suggestive of DM, no clinical evidence of proximal motor weakness or derangement of muscle enzymes within two years of onset of skin disease.[3] Euwer later stated that provisional diagnosis can be made if this interval is reduced to six months.[4] History of immunosuppressive intake at least for two consecutive months prior to patient presentation should be excluded.[1] Stonecipher et al[5] also proposed a classification system and divided patients with amyopathic dermatomyositis into three main groups depending on whether there was no muscle involvement, subsequent clinical or subclincal myositis.

In adults both DM and CADM may show internal malignancy or interstitial lung disease (ILD) which is infrequently reported in juvenile variants. A retrospective study by Gerami et al[6] over a period of 43 years (1963–2006) identified only 68 patients with juvenile onset CADM (JCADM) who had a mean age of onset 10.8 years with no significant difference in female to male ratio. They observed that 26.4% of JCADM progressed to juvenile onset classic DM (JCDM).[6] El-Azhary et al[7] in his 18 years retrospective study found that JCADM comprised of 18.9% of total CADM. A ten years study at Taiwan showed only 9.5% of total juvenile DM.[8] Vasculopathy and calcinosis cutis are more commonly seen in JCDM and very rare in JCADM. Gerami et al[6] observed absence of vasculopathy and presence of calcinosis cutis in 4% cases of JCADM. In our case calcinosis cutis was the presenting feature and diagnosis of JCADM was made only after examination.

Isolated cases of systemic complications in patients of JCADM were reported. Abe et al[9] reported rapidly progressive ILD in a 16-year-old child suffering from

JCADM. Kobayashi et al[10] reported five cases of rapidly progressive ILD in patients of both the juvenile variants of DM. These patients responded poorly to potent immunosuppressives. A vigilant approach is required in all variants of DM.

Conclusion

Patients of CADM may not present early because of lack of muscle involvement despite cutaneous findings and hence diagnosis is often delayed. There are 26% chances of juvenile onset clinically amyopathic dermatomyositis (JCADM) progressing to juvenile onset dermatomyositis (JDM). There is high risk of developing rapidly progressive interstitial lung disease, vasculopathy and calcinosis cutis in JDM while in JCADM they are rarely seen. JDM may require aggressive therapy while JCADM can be treated by topical steroids and antimalarials and a regular follow-up is required.

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