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Case Study

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CLINICALLY AMYOPATHIC DERMATOMYOSITIS

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ABSTRACT

Clinically amyopathic dermatomyositis (CADM) is a unique subgroup of dermatomyositis, with typical skin manifestations of dermatomyositis but little or no evidence of myositis. The case discussed presented in the OPD with heliotrope rash, gottron papules, periungual hemorrhages on dermoscopy with positive ANA with no evidence of muscle involvement and was managed by hydroxychloroquine and topical corticosteroids.

KEYWORDS: CADM, Dermatomyositis.

INTRODUCTION

CADM is a distinct clinical entity with unique clinical manifestations and autoantibody profiles unlike the classic dermatomyositis. Dermatomyositis is included in idiopathic inflammatory myopathies and shows muscle manifestations such as 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 cutaneous features such as heliotrope rash and Gottron's papules.[1] However, some patients displayed only the hallmark cutaneous manifestations of dermatomyositis as an isolated clinical finding and had not developed muscle weakness for a prolonged period.

CASE

The patient was a 33-year-old female presented with gradually progressing swelling around eyes in the past 6 years, which used to present throughout the day, not associated with itching, associated with reddish discoloration of both upper eyelids. She had pink colored asymptomatic lesions over knuckles of right hand in the past 4 years and developed similar lesions over left hand in last 2 years. She had photosensitivity and history suggestive of Raynaud's phenomenon was present. There was no difficulty in standing from squatting position, climbing stairs, combing hair or difficulty in swallowing or difficulty in speech. On examination, there was violaceous erythema and edema over upper eyelids suggestive of heliotrope rash [Figure 1]. Erythematous papules were present over dorsum of MCP and PIP joint of index, middle and ring finger of right hand and PIP joint of index and middle finger of left hand suggestive of gottron papules [Figure 2]. On dermoscopy, periungual erythema and telangiectasias with ragged cuticles with hemorrhages were seen [Figure 3]. Systemic and musculoskeletal examination was normal. In investigations, ANA was positive, fine speckled. Muscle enzymes and EMG was normal. She was treated with hydroxychloroquine, nifedipine, topical corticosteroids and sunscreen and was responding to the treatment.

Figure legends

Figure 1: Violaceous erythema and edema over upper eyelids s/o heliotrope rash.

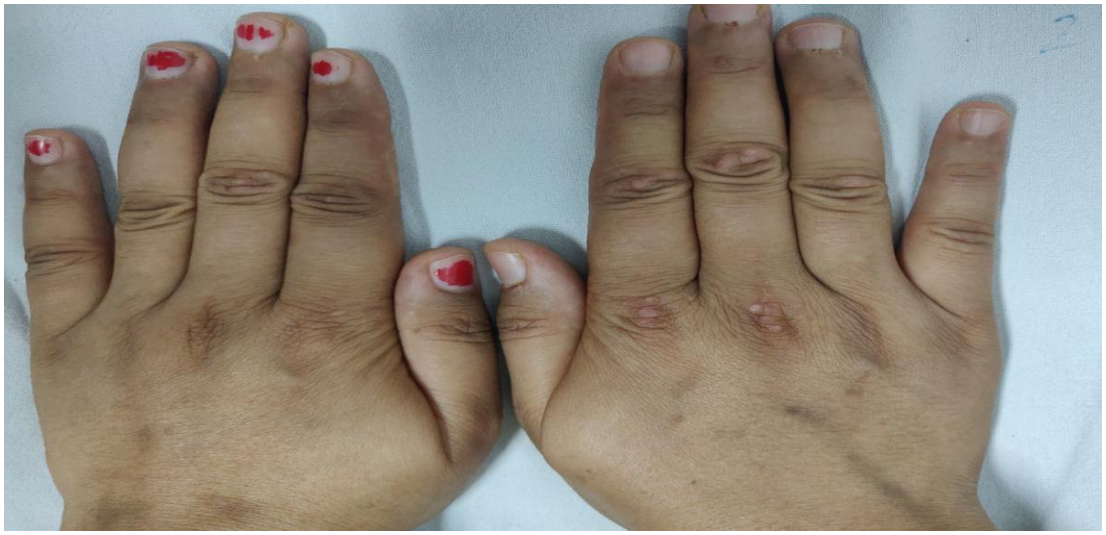

Figure 2: Erythematous papules over dorsum of MCP and PIP joint of index, middle and ring finger of right hand and PIP joint of index and middle finger of left hand.

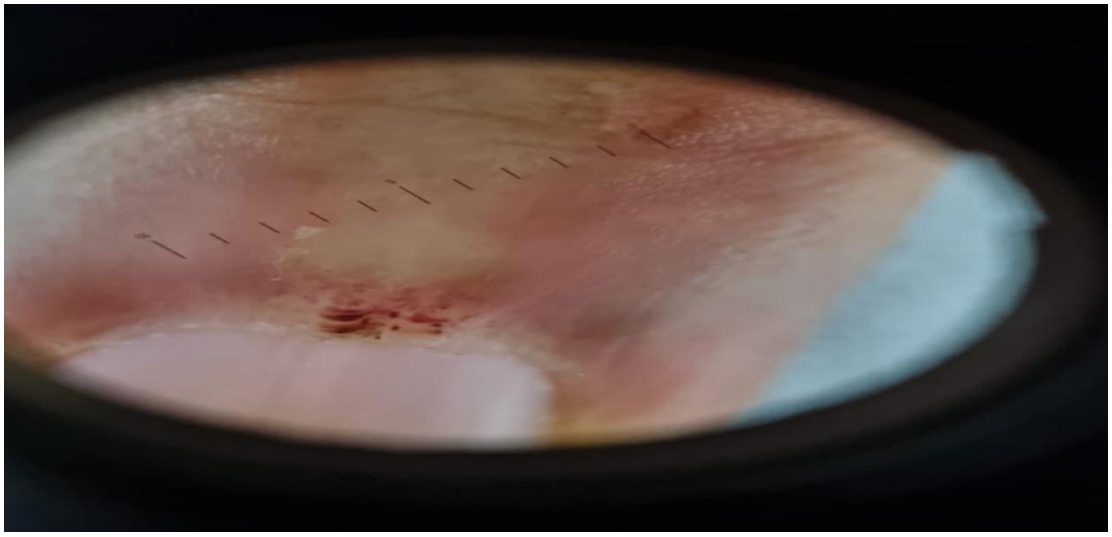

Figure 3: Dermoscopy shows periungual erythema and telangiectasias with ragged cuticles with hemorrhages.

DISCUSSION

The term "amyopathic dermatomyositis" was first coined by Pearson to distinguish patients with hallmark cutaneous manifestations of dermatomyositis who do not show clinical evidence of muscle involvement. This was later refined to include true "amyopathic dermatomyositis" patients who have no clinical or laboratory evidence of muscle involvement, as well as "hypomyopathic dermatomyositis" patients who had no clinical evidence of muscle disease, but do show evidence based on EMG, or muscle biopsy.[2] Data suggest that cutaneous and histopathologic findings in amyopathic dermatomyositis patients are no different from the "classic" dermatomyositis patients, and the same systemic disease associations are present in both groups including an association with malignancy and lung disease.[3]

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