Clinical diversity of dermatomyositis – illustrated by three cases

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Abstract

Although dermatomyositis (DM) is an uncommon connective tissue disease, we came across three patients with many of the clinical features of DM, within a short period. One of the patients had an underlying malignancy in addition. We report these three cases to highlight the clinical diversity of dermatomyositis, in order to prevent delay in diagnosis and treatment of these patients.

Introduction

Dermatomyositis (DM) is an idiopathic inflammatory polymyositis with characteristic cutaneous manifestations. The exact cause is currently unknown but various viral infections and autoimmune mechanisms have been proposed. The incidence of carcinoma associated with DM varies from 15% to 34%[1]. DM precedes the neoplasm in 40%, occurs simaltaneously in 26% or may follow in 34%[1]. DM has bimodal age distribution with both juvenile and adult forms. Male: Female ratio of the disease is 1:2[1]. Older age of onset and female gender are associated with a high risk of internal malignancy[2]. All adult patients especially if they are more than 40 years require a complete evaluation and investigations for underlying malignancy. In this paper we describe three patients with different clinical presentations encountered by us within a short period.

Case report

All three patients were adults. Out of three patients two were elderly females and one was a young male. Two of the patients developed cutaneous manifestations prior to myopathy and one developed both features simultaneously.

First patient was a 62 year old female with poor appetite and weight loss and a BMI of 17.2. She had many of the typical clinical manifestations of DM, including heliotrope rash, periorbital swelling, facial erythema and pigmentation, shawl sign, photodermatitis, diffuse alopecia with seborrhoeic capitis, poikiloderma, Gottron's sign, ragged cuticles, urticarial lesions and marked pruritus. In addition she had hyperkeratotic pigmented lesions suggestive of mechanic's hands. One month later she developed proximal muscle weakness and generalized lymphadenopathy. Rest of the clinical examination was normal.

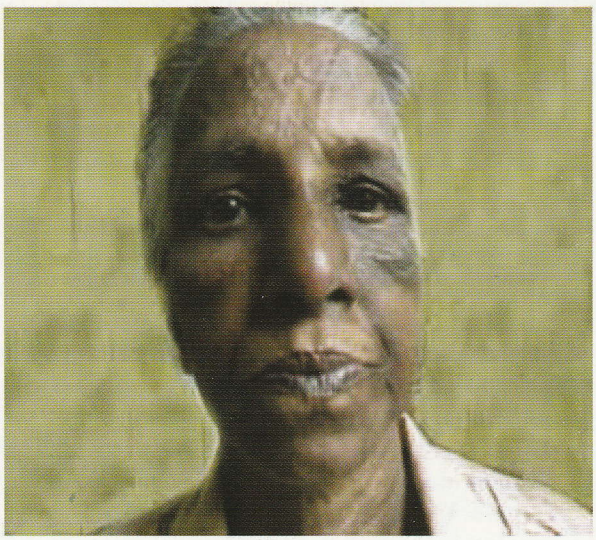

Figure 1. *Case 1 – Heliotrope rash and periorbital swelling.*

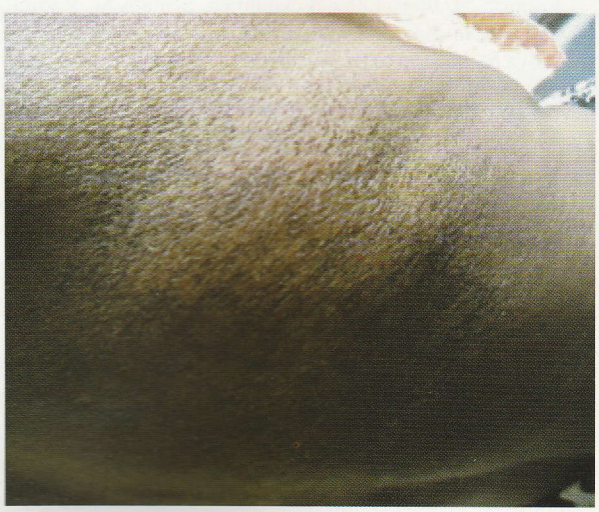

Figure 2. *Case 1 – Poikiloderma.*

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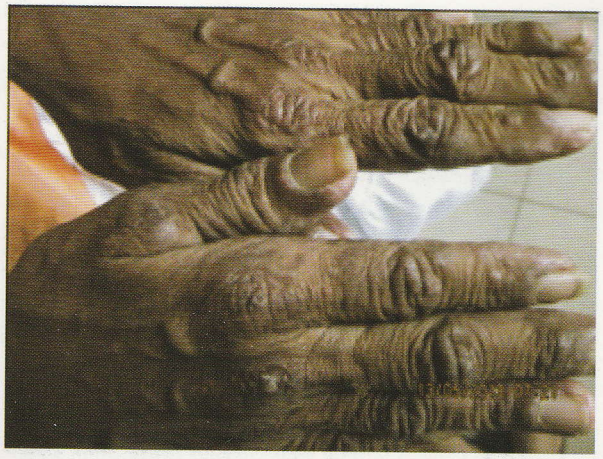
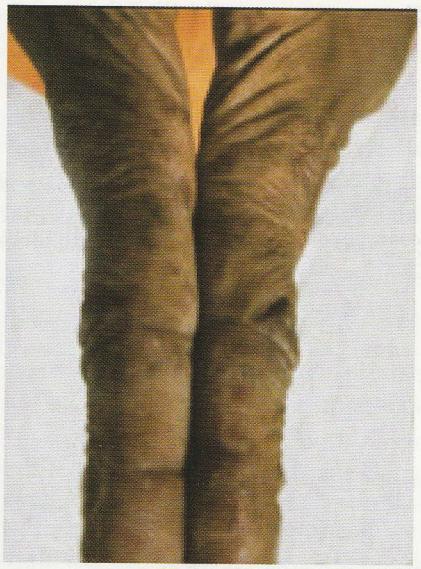


Figure 3. *Case 1 – Gottron's sign.*

A



B

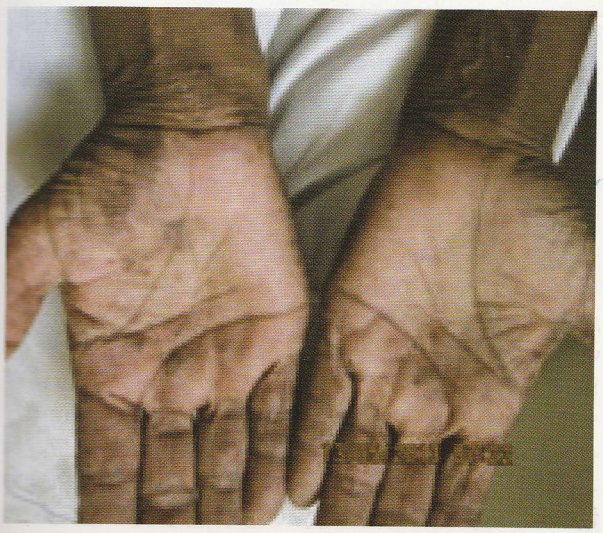


Figure 4 A, B. *Case 1 – Mechanic's hand.*

Our second patient was a 53 year old female who had developed a facial erythematous eruption along with weakness of the body. A right sided breast lump noted at the same time, which was proven to be an invasive ductal carcinoma of breast. Subsequently, right sided mastectomy was done followed by chemotherapy. Meanwhile her weakness progressively deteriorated until she became bed bound. At presenation she had diffuse non scarring alopecia, poikiloderma, ragged cuticles and cutaneous ulceration.

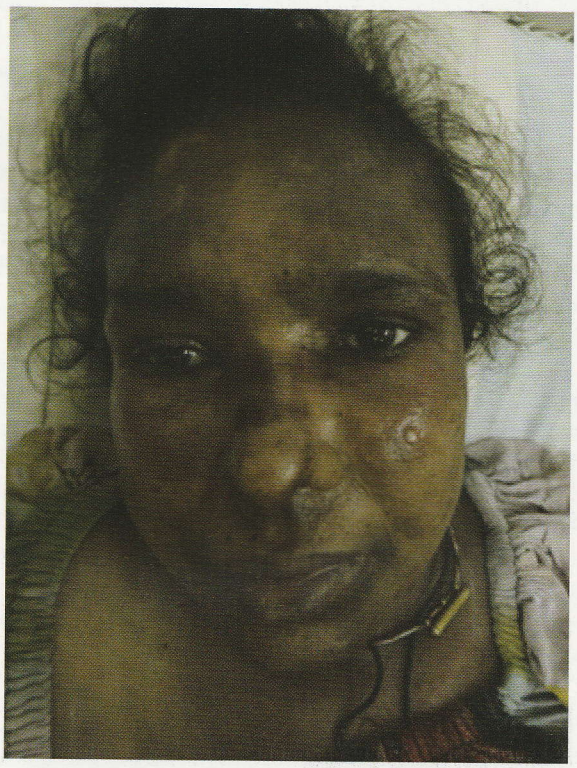


Figure 5. *Case 2 – Facial erythema and diffuse alopecia.*

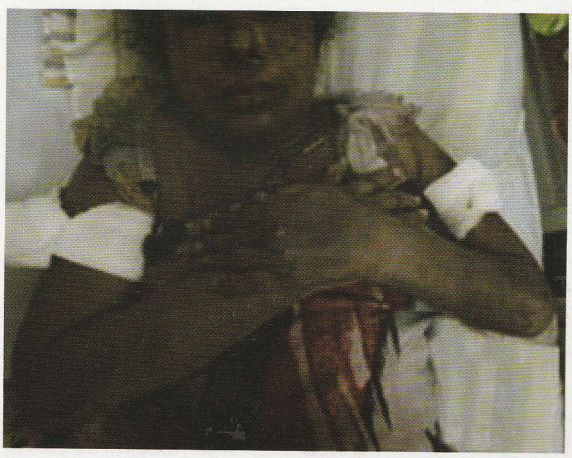


Figure 6. *Case 2.*

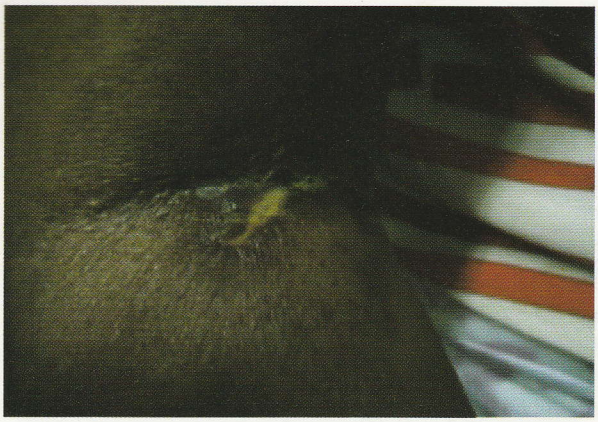


Figure 7. *Case 2 – Cutaneous ulcer.*

Third patient was a 26 year old male who presented with erythema over both cheeks and nasal bridge which increased with sun exposure. Subsequently he developed swelling, redness over periorbital region and few inflammatory papules which resembled rosacea.

Later he developed hyperpigmented macules, discoid lupus erythematosus like lesions involving face, both ears and neck, photodermatitis over upper chest and shawl sign. Nail fold ragged cuticles and capillary dilations, telangiectasia were also present. Palatal ulcers, vasculitic lesions or features of SLE were not observed. Proximal muscle weakness developed two weeks later and then flexors. Axillary and inguinal lymph nodes were palpable but there was no organomegaly.

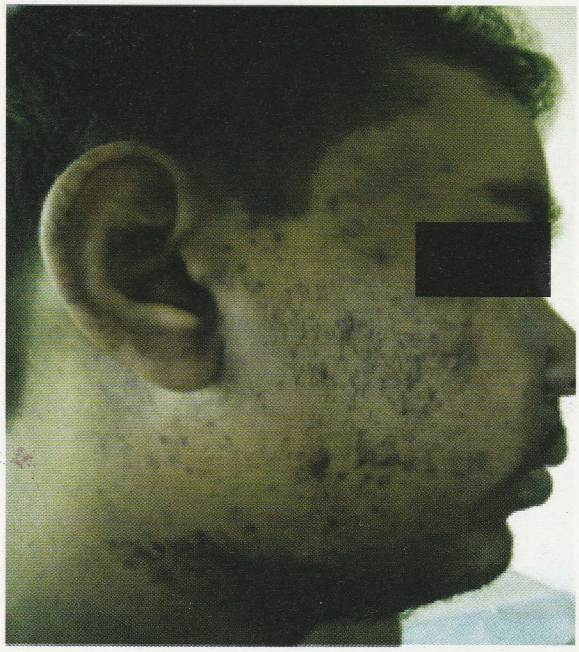


Figure 8. *Case 3 – Pigmentation and DLE like lesions.*

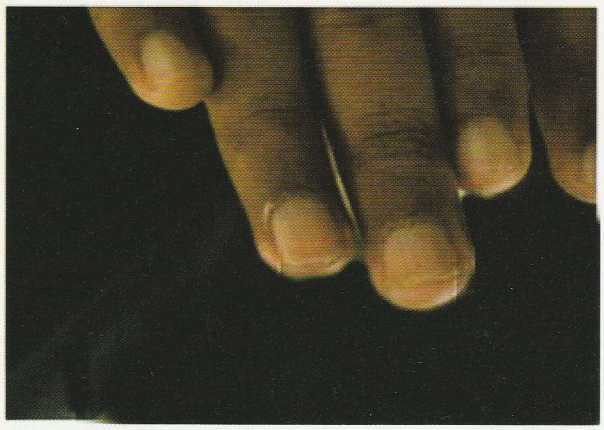


Figure 9. *Case 3 – Nail fold capillary dilatation and ragged cuticles.*

Investigations were done mainly to confirm the diagnosis of DM and additionally focusing on evaluation of underlying malignancy. Significant investigation results are summarized in Table 1.

In case 1 we investigated the patient thoroughly for an underlying malignancy by performing thyroid USS, thyroid profile, USS guided FNAC, USS abdomen, CT abdomen, mammogram, UGIE (Upper Gastro Intestinal Endoscopy), stool for occult blood, CEA (Carcinoembryonic antigen), CA 125 (Carcinoma antigen 125), but all were normal.

In patient no. 3, lymphoma was excluded by lymph node biopsy.

Discussion

DM is not a common entity, however we were intrigued to come across three cases within a space of six months with entirely different and interesting clinical presentations. We wish to highlight the following features. In case no. 1, elderly female with typical clinical features of DM did not have any demonstrable malignancy. However, she needs periodic evaluation to rule out occult malignancy.

Second patient was also an elderly woman with typical clinical features of DM associated with carcinoma of breast. She had poor prognostic features like severe disabling proximal muscle weakness, old age, pro-gressive disease, dysphagia, associated malignancy and cutaneous ulcers[1,3]. Cutaneous ulceration is a risk factor for internal malignancy in DM[4]. Although this patient improved with oral steroids (1 mg/kg/d) initially, she subsequently developed hypokalemia. Unfortunately inspite of typical clinical features and an obvious neoplasm being diagnosed, her dermatomyositis had been overlooked.

Table 1.

Investigation	*Case 1*	*Case 2*	*Case 3*
CPK	320 U/L	447 U/L	6136 U/L
SGOT	89 U/L	72 U/L	81 U/L
SGPT	38 U/L	57 U/L	107 U/L
EMG	Myositis	Myositis	Myositis
Skin Biopsy	Non specific	Non specific	Non specific
Muscle Biopsy	Non specific	Non specific	Non specific
LN Biopsy	Dermatopathic Lymphadenitis	LN are free of tumor deposits	Dermatopathic Lymphadenitis
ANA	Negative	Negative	Positive> 1/80
Anti Jo Ab	Negative	Negative	Negative
USS Abdomen	Normal	Grade ll Fatty Liver	Normal
CXR PA	Normal	Normal	Normal
ESR (mm/1st hr)	40	65	33

Patient no. 3 is a young male and no underlying malignancy has been detected so far and is more likely to be idiopathic DM. His cutaneous features were initially subtle and mimicked rosacea.

All 3 patients were treated with steroids and methotrexate. In 2nd patient who had carcinoma of breast, response to treatment was less satisfactory.

We reported these three cases of dermatomyositis, although not a common disease in dermatology field, to highlight the variations in presentations and fascinating cutaneous features.

References

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