

Case Report

Paraneoplastic dermatomyositis: a case report from North-East India

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ABSTRACT

Dermatomyositis is a rare autoimmune myopathy with distinctive cutaneous changes. It has been associated with a wide range of malignancies like carcinoma ovary, lung, breast, gastrointestinal tract and non-Hodgkin's lymphoma. The malignancy can be concurrent or can occur before or after the onset of myositis. We report a case of dermatomyositis in a 63-year-old Indian male with nasopharyngeal carcinoma. The characteristic cutaneous features and muscle weakness developed concomitantly with the onset of nasopharyngeal carcinoma.

Keywords: Paraneoplastic, Dermatomyositis, Nasopharyngeal carcinoma

INTRODUCTION

Dermatomyositis is an uncommon inflammatory myopathy associated with characteristic cutaneous findings occurring in an immuno-genetically predisposed individual. It has been linked with various diseases including different internal malignancies. This association was first reported by Shy GM in the year 1962.¹ Dermatomyositis may precede or follow the onset of malignancy or pursue an independent course. We report a case of dermatomyositis associated with nasopharyngeal carcinoma.

CASE REPORT

A 63-year-old male presented with generalized pruritic erythematous rash mostly over the face, chest, back, extensor arms and dorsal hands associated with proximal muscle pain and weakness for 8 weeks. He also had right sided nasal blockage with occasional blood stained nasal discharge. There was no history of photosensitivity, generalized oedema, Raynaud's phenomenon, arthralgia, arthritis or other systemic complaints. He suffered from pulmonary tuberculosis 10 years back. Physical examination revealed firm, non-tender right cervical

lymphadenopathy (3.5cm x 2.5 cm). On cutaneous examination, diffuse erythema on the upper trunk and extensor aspect of upper limbs was noted along with dusky erythema and mild oedema on the eyelids as well as malar area (Figure 1). Poikiloderma was present on the lower abdomen and back (Figure 2). A nail fold change in the form of cuticular thickening was also present (Figure 3). Proximal muscles were mildly tender and muscle power was grade IV. Otorhinolaryngological examination revealed a right sided nasopharyngeal mass.



Figure 1: Dusky erythema and mild oedema on the eyelids and malar area.

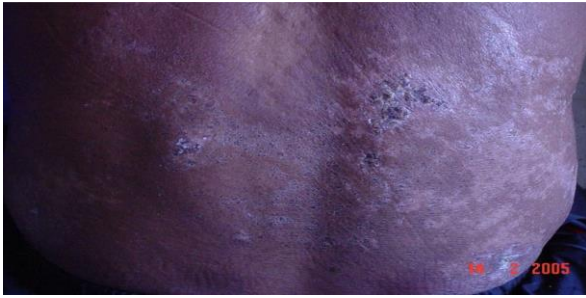


Figure 2: Poikilodermatous changes over back.



Figure 3: Thickened and ragged cuticles of finger nails.

Routine biochemical tests were within normal limit except raised Erythrocyte sedimentation rate (ESR). Antinuclear antibodies were negative. Serum creatine kinase level was high (471 U/L). Chest X-ray showed calcified opacities in the upper zones. Ultrasonography of abdomen showed mild hepatomegaly and mild prostatomegaly. Electromyography showed short duration, low amplitude, polyphasic waves suggestive of myopathy. Electrocardiogram was normal. Muscle biopsy was not done. Computed tomography (CT) scan of nasopharynx revealed soft tissue density mass at the right nasopharynx obliterating fossa of Rosenmuller, extending to parapharyngeal and retropharyngeal space (Figure 4). Biopsy of the nasopharyngeal mass showed features of well differentiated squamous cell carcinoma. Fine needle aspiration cytology (FNAC) of the cervical lymphnode also showed features of well differentiated squamous cell carcinoma. Skin biopsy from the upper back showed an atrophic epidermis, focal vacuolar degeneration of basal cells, sparse perivascular lymphocytic infiltrate and fragmented collagen (Figure 5).

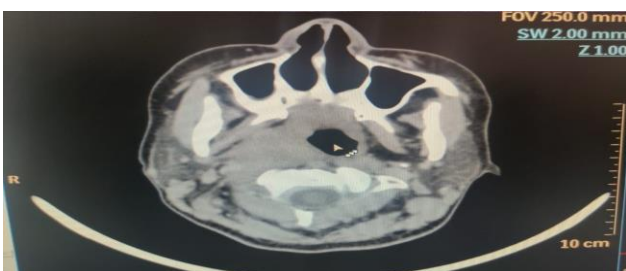


Figure 4: CT scan showing mass in right nasopharynx obliterating fossa of Rosenmuller, extending to parapharyngeal and retropharyngeal space.

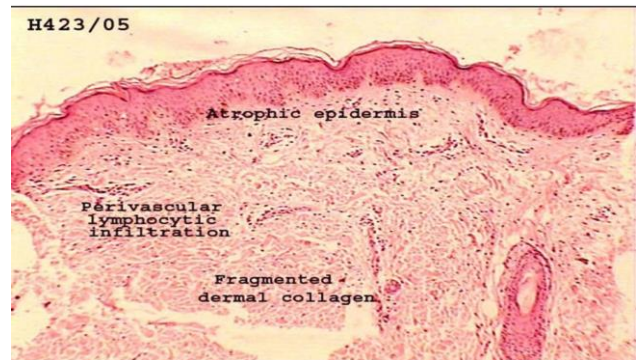


Figure 5: Atrophic epidermis, focal vacuolar degeneration of basal cells, sparse perivascular lymphocytic infiltrate and fragmented collagen in dermis (H & E, 40X).

On the basis of the characteristic cutaneous eruption coupled with increased creatine kinase, myopathic features of electromyography and evidence of squamous cell carcinoma in the nasopharyngeal mass, the diagnosis of paraneoplastic dermatomyositis was established. Prednisolone 40mg daily was started which was tapered gradually over a period of 6 months. Simultaneously external radiation with a total dose of 60Gy was given over a period of 6 weeks along with Carboplatin 150 mg weekly for 7 cycles. Adjuvant chemotherapy with injectable Carboplatin 450 mg and 5-Fluorouracil 1gm was further given for another 7 cycles. Three months later, cervical lymphadenopathy had disappeared, nasal blockage had decreased and muscle power had returned to normal. Serum creatine kinase had also returned to normal values (178 U/L). Repeat CT scan 6 months later however still showed mass in the right sided nasopharynx. The cutaneous changes showed significant improvement except the post inflammatory hypopigmentation on dorsum of hands.

DISCUSSION

Dermatomyositis is an idiopathic inflammatory myopathy with cutaneous involvement. The incidence of paraneoplastic dermatomyositis ranges from 4.4- 60% according to different series.² Elderly females are more commonly affected.³ In this case, the patient was a 63-year-old male.

Carcinomas are more commonly associated with dermatomyositis than sarcomas. Carcinoma of ovary, lung, gastrointestinal tract, nasopharynx and haematological malignancies are known to be associated with dermatomyositis.⁴ The type of associated tumours also vary with the ethnicity of the patients or certain geographic regions. Nasopharyngeal carcinoma is more prevalent in South-East Asia and Southern China.⁵ Our case belonged to North-East region of India who was diagnosed with nasopharyngeal carcinoma following onset of rash. Gupta, et al⁶ had also reported a case of paraneoplastic dermatomyositis associated with

nasopharyngeal carcinoma from this region. The pathogenesis of paraneoplastic dermatomyositis is not clear. It is presumed to be an immune response by the body against foreign antigens liberated by tumour tissue sharing common antigen with skin and muscle or a result from the damaging effect of cellular autoantibodies from mutant lymphocyte clones.³

Clinically, paraneoplastic dermatomyositis have more rapid onset, absence of Raynaud's phenomenon, vesicular and bullous lesions, bright erythema on the face, neck and shoulders. Moreover cutaneous necrosis, generalized hyperkeratotic follicular papules and laboratory findings of increased ESR, C-reactive protein, CA-125 as well as therapeutic resistance are also reported to be present.⁷In our case, the patient demonstrated dusky erythema and edema of face and poikiloderma with proximal muscle weakness. Although physical examination and imaging techniques may fail to detect malignancies early, CT scan of chest, abdomen with pelvis and mammography in women must be performed.^{7,8}

Dermatomyositis with its various forms of presentations can act as a clinical indicator for an associated malignancy. This case is being reported to highlight the uncommon association of dermatomyositis with nasopharyngeal carcinoma and distinctive ethnic connection. Improvement in clinical and biochemical parameters after institution of treatment was, however, not correlated with tumour regression.

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Ethical approval: Not required

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