# Dermatomyositis associated with ovarian malignancy

F M Srisaravanapavananthan[1], G M P Sirimanna[2]

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## Introduction

Dermatomyositis is a rare inflammatory myopathy with characteristic skin manifestations and muscle weakness. In the female it is well known to be associated with ovarian malignancy. This association was first reported by Stertz in 1916[1]. Diagnosis of malignancy can be antecedent (40%), concurrent (26%) or subsequent to the occurence of dermatomyositis (34%)[2].

## Case report

A 39-year old female presented with erythematous malar and periorbital rash of one month duration. She had palatal ulcers and photosensitivity too. Six weeks later she developed progressive dysphagia and proximal muscle weakness.

Examination revealed, painful superficial necrotic ulcers with ragged margins located on face, ears, nape of the neck, upper chest and back. There was periorbital oedema with heliotrope discoloration and periungual erythema. Non tender firm lymph node enlargement was noted in left supraclavicular fossa. Thyroid, breasts and abdomen were normal.

Grade 4 muscle weakness with mild tenderness was noted in proximal girdle muscles

## Investigations

The ESR was 40mm 1st hour, ANA titre was >1/80 and the CPK was raised on two occasions 322IU and 413IU(normal 24-195IU). CA 125 was 222 u/ml (normal <35u/ml).

EMG did not reveal any abnormality.

Skin biopsy showed *Leucocytoclastic vasculitis* with secondary abscess formation.

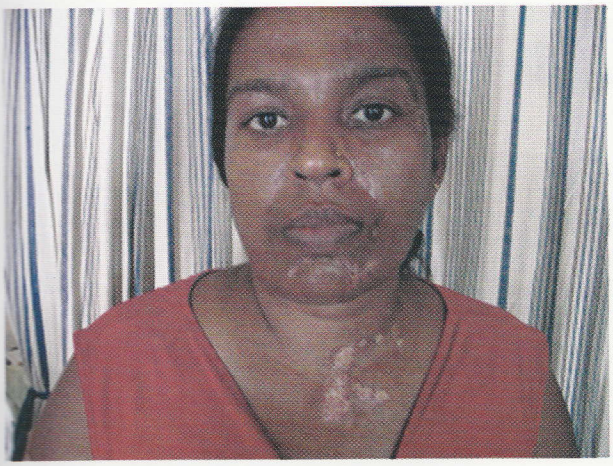
Fine needle aspiration and cytology of left supraclavicular LN showed metastatic deposit of adenocarcinoma.

Contrast enhanced CT scan showed a pelvic mass on either side of uterus appearing to be ovarian in origin. Evidence of supraclavicular and para aortic lymphadenopathy was also noted. Mammogram of both breasts was normal.

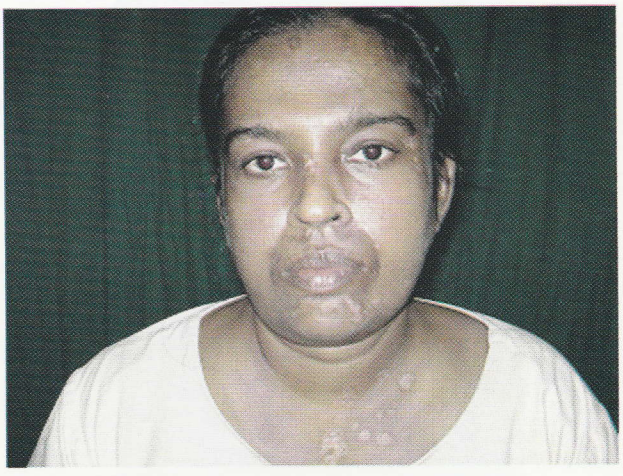
Histological examination showed poorly differentiated papillary adenocarcinoma of both ovaries. Extensive lymphatic invasion of myometrium was noted.

## Management

Once the clinical diagnosis of dermatomyositis was made, the patient was initiated on 30mg of



*Necrotic skin lesions.*



*Periorbital oedema with heliotrope rash.*

[1]*Registrar in Dermatology,* [2]*Consultant Dermatologist, National Hospital of Sri Lanka, Colombo.*

prednisolone daily. However, a poor response to treatment lead to further evaluation, and subsequently to the detection of malignant deposits in left supraclavicular lymphnodes. The patient was referred to an oncologist. Laporotomy was done at the gynecology unit of Cancer Institute, Maharagama. It showed bilateral ovarian tumours. Total abdominal hysterectomy and bilateral salpingooophorectomy was done. After debulking surgery she was treated with six cycles of chemotherapy (Paclitaxel and Cisplatin). Skin lesions resolved completely at the end of treatment.

## Discussion

Although several cutaneous changes that are highly suggestive of dermatomyositis are described in literature often the skin findings are non specific. Rarely necrotic lesions can be seen[3]. Our patient presented with well known features such as peri-orbital oedema and heliotrope discoloration as well as necrotic lesions.

Skin biopsy from the necrotic lesion showed leucocytoclastic vasculitis. Due to progression of dysphagia and proximal myopathy despite treatment, we evaluated her repeatedly regarding the possibility of an occult malignancy.

Ultimately the patient was found to have adenocarcinoma of ovaries with metastasis to left supraclavicular lymph node. Ovarian carcinoma is the commonest malignancy associated with dermatomyositis in females.

## Conclusion

Skin rash with progressive muscle weakness in an adult should be thoroughly investigated in search of occult malignancy. Repeated examination and investigations are necessary to detect occult malignancy in treatment resistant dermatomyositis or relapse of previously stable dermatomyositis[2].

### References

1. Kovacs SO, Kovacs CS. Dermatomyositis. *J Am Acad Dermatol* 1998; **39**: 899-913.
2. Rook's textbook of Dermatology. Burns T, Breathnach S, Cox N, Griffiths C. 7th edition, Blackwell Science, Chapter 56, 127-38.
3. Dermatology, Braun-Falco O, Plewig G, Wolff HH, Burgdorf WHC. Second edition, Spinger, Chapter 18, 813-18.
4. Guidelines of care for dermatomyositis. *J Am Acad Dermatol* 1996; **34**: 824-29.