## SELF ASSESSMENT PROGRAMME

A 50 year old male office worker presented with an 8 month history of erythematous, hypopigmented skin lesions on the face, the trunk and the extremities. He also complained of difficulty in swallowing and pain and weakness of the girdle muscles for 7 months. The complaints started with high fever and erythematous macular rash on the face, the scalp and the extremities. A few days later he noticed difficulty in swallowing solid foods. His symptoms were relieved after a month of systemic corticosteroid therapy but the symptoms recurred soon after corticoids were discontinued. There was no history of Raynaud's phenomenon, arthralgia, respiratory symptoms, oliguria or edema.

The patient was well built but looked ill. He had multiple erythematous hypopigmented macular lesions on the scalp, the forehead, the chest and the arms with some bluish hyperpigmentation around the border ; the lesions were mildly scaly. Liver was palpable, 3 cm below right costal margin and non tender. He had weakness of shoulder and thigh muscles. His blood pressure was normal. Rest of the examination was non contributory.

A. What is the likely diagnosis ?

1. Systemic lupus erythematosus
2. Dermatomyositis
3. Mixed connective tissue disease
4. Systemic Sclerosis
5. SLE + Steroid myopathy

B. Which of the following investigations are most useful ?

1. Barium swallow
2. X - ray Chest
3. Muscle biopsy
4. Skin biopsy
5. Antinuclear factor (ANF)
6. Muscle enzymes
7. Electromyography
8. LE cell test
9. Urinary creatine excretion.

The muscle biopsy showed variable degeneration of the muscle fibres with oedema and inflammatory cells. The skin biopsy was non-specific.

C. What are the chances of associated malignancy in this patient.

1. Rare
2. Occasional
3. Invariable
4. Quite often

D. Which of the following treatment is preferred ?

1. Corticosteroids
2. Anabolic hormones
3. Immunosuppressants
4. Chloroquine
5. Potassium para-aminobenzoate (POTABA)

E. What is the likely prognosis ?

1. Fatal
2. Gradual worsening
3. Remissions and recurrences.

## ANSWERS

A. The most likely diagnosis seems to be dermatomyositis because of his proximal muscle weakness and skin lesions. SLE, particularly associated with steroid myopathy cannot be confidently excluded in this case even though other manifestations are absent. Mixed connective tissue disease can only be diagnosed after various investigations the results of which may be difficult to explain by dermatomyositis alone. Muscle weakness and difficulty in swallowing suggest systemic sclerosis but the absence of Raynaud's phenomenon and the type of skin lesions seen disfavour this diagnosis. Steroid myopathy simulates polymyositis but is not associated with skin lesions for which an alternative explanation will be necessary.

B. Muscle biopsy, muscle enzymes and ANF would help in differentiating the above conditions in this case. A negative LE cell test and the absence of antinuclear anti-bodies excluded SLE. Elevated muscle enzymes and muscle biopsy showed features of dermatomyositis. The electromyography showed features of polymyositis, further confirming the diagnosis.

C The association of malignancy with dermatomyositis is seen quite often. The frequency is reported to vary from 6.7 to 50% in cases of dermatomyositis. The incidence is said to be more in patients over 40, particularly males. There is no association with dermatomyositis seen in children.

D. The preferred treatment is systemic corticosteroids monitoring the activity by clinical picture, the muscle enzymes and urinary creatine excretion. Methotrexate and Azathioprine have been tried in cases not responding to corticosteroids.

Methandienone, though given along with systemic corticoids is said to help the disease. Potaba and chloroquine have not been proven to be effective.

E. The prognosis is unpredictable and the patient has to be maintained on systemic steroid therapy for a longtime. If associated with an internal malignancy, the disease may regress after the removal of the malignancy. The disease may fluctuate but generally takes a gradual down-hill course.

**Comment**

Steroid myopathy shows the same pattern of muscle weakness as that seen in polymyositis but can be differentiated by serial estimations of muscle enzymes which are elevated in polymyositis and unchanged in steroid myopathy[1]. Urinary creatine excretion varies directly with the activity of the disease in dermatomyositis and with the dosage of steroids in steroid myopathy. The association of internal malignancy with dermatomyositis has been well established, and chances of upto 50% have been claimed by some[2]. The progress of the disease in that case depends upon the neoplasm and may improve when the latter is treated. It is always worthwhile to search for an occult neoplasm particularly in a patient over 40 years, since this may have a great bearing on the progress of the disease.

**References**


1. Ali Aksari et al: Steroid myopathy in connective tissue disease, Amer J Med, 61 : 485, 1976.

2. Arundell FD, Wilkinson RD, Haserick JR : Dermatomyositis and malignant neoplasms in adults, Arch Dermatol 82 : 772, 1960.