SELF - ASSESSMENT PROGRAMME

A fifty-six year old house wife presented with gross puffiness of face of three months' duration and generalised erythematous scaly lesions for a fortnight. The lesions came on abruptly and had gradually progressed. There were no constitutional symptoms. For a fortnight, the patient also complained of hoarseness of voice and difficulty in swallowing liquids as well as solids. Patient was well built and obese and had diffuse, fairly widespread erythematous, warm, slightly tender and indurated areas on the back and front of trunk and to a lesser extent on extremities. Her eyelids were swollen, red and scaly. Similar lesions were present on front of the neck and 'V' of chest. Patient was afebrile, normotensive and in mild discomfort. Systemic examination did not reveal any significant abnormality.

1. Which of the following diagnoses is most likely ?
   - A. Dermatomyositis
   - B. Airborne contact dermatitis
   - C. Photosensitive dermatitis
   - D. Cellulitis
   - E. Trichinosis

2. Which of the following investigation(s) will be most useful?
   - A. Skin biopsy
   - B. Muscle biopsy
   - C. Patch test
   - D. Photo-patch test
   - E. Estimation of muscle enzymes (CPK, LDH)

Skin biopsy revealed degeneration of the basal cell layer, atrophy of the epidermis and hyalinized dermis. Muscle enzymes, more particularly lactic dehydrogenase (LDH) and creatinine phosphokinase (CPK) were grossly elevated. Muscle biopsy was normal. A tentative diagnosis of dermatomyositis was made.

3. What further investigation ought to be carried out ?
   - A. X-ray chest
   - B. Barium meal
   - C. Gynaecological check up
   - D. Electromyogram

   All these examinations were normal.

4. What should be the line of management?
   A. Systemic steroids
   B. Topical steroids
   C. Systemic steroids with Methotrexate or Azathioprine.

5. What is the prognosis of this patient?
   A. Potentially fatal
   B. Depends on associated malignancies
   C. Spontaneous recovery

ANSWERS

1. The most probable diagnosis in this patient would be dermatomyositis because of puffiness and redness of the eyelids and the exposed and unexposed areas of the skin. Photocontact dermatitis should not have severely involved the trunk, an argument that would also hold for airborne contact dermatitis. Absence of itching and presence of fair degree of induration with relatively minimal epidermal changes would also disfavour the diagnosis of any dermatitic process. The patient was afebrile and the lesions were not acutely tender; hence a generalized cellulitis could also not be regarded as a very likely possibility. Clinically it is very difficult to distinguish trichinosis from dermatomyositis.

2. The most useful investigations would be muscle biopsy and a study of muscle enzymes since they would give us an indication about the degeneration of muscles in the presence or absence of any clinical features of myositis. A normal muscle biopsy as in this case does not however exclude the diagnosis of dermatomyositis. Skin biopsy can also render useful supportive evidence.

3. In further investigations of this patient, two points have to be borne in mind: first, to confirm the diagnosis of dermatomyositis by other laboratory investigations such as electromyogram and secondly to exclude the possibility of any associated malignancy. The patient should therefore be thoroughly screened from "head to toe" for a proper exclusion or otherwise of a malignant growth. Breast examination and gynaecological examination are particularly valuable.

4. Topical steroids are of no value in a systemic disease such as dermatomyositis. Systemic steroids, preferably not triamcinolone, should be the first line of therapy. If the patient does not show significant improvement within a reasonable period of time, say two to four weeks, the treatment may be supplemented with methotrexate or azathioprine. Maintenance dose of one or more of these drugs ought to be continued for a fair period of time, say a couple of years, even if there is no relapse.

5. The prognosis of dermatomyositis is unpredictable. Lack of detection of any malignancy on first examination dose not exclude the possibility that a malignancy may not appear subsequently. The prognosis would thus depend firstly on the severity and acuteness of the disease itself and secondly on the nature of the associated malignancy.

Comments :

Dermatomyositis is one of the relatively uncommon connective tissue disorders. It is a polymyositis with variable cutaneous manifestations. Most often the face, particularly the eyelids, neck and 'V' of the chest or other light exposed areas are preferentially affected. The skin involvement can, as in present case, however, be very widespread and diffuse. Muscle involvement does not run pari pasu with the skin involvement. Most commonly involved msucles are those belonging to the pelvic and shoulder girdles, but any muscle such as laryngeal, oesophageal or cardiac may be affected. This patient had evidence of laryngeal and oesophageal muscular involvement. Association between dermatomyositis and malignancy, has been variously reported in the adults[1]. Frequency has varied between 15–34%. It is, therefore, essential that every patient should be thoroughly investigated for an associated neoplasm which should be adequately and properly treated. Complete reversal of dermatomyositis has been known after removal of the tumor.

In the absence of an associated malignancy, most patients would need systemic corticosteroids with or without antimitotic agents. All fluorinated steroids, but more particularly triamcinolone, result in myopathy which may complicate the picture. A serial estimation of muscle enzymes would help differentiate active damage due to dermatomyositis or steroids. Malavia et al for the first time reported the utility of using methotrexate in patients who do not respond to systemic steroids[2].

A word about differentiation of dermatomyositis from trichinosis. The clinical, biochemical and histological picture may so closely mimic trichinosis that it may become well nigh impossible to distinguish between the two conditions except by demonstration of encysted larvae in the muscle. Incidentally, trichinosis is a rare disease in this country.

References :

1. Callen JP, Hyla JF, Bob GG and Kay DR : The relationship of dermatomyositis and polymyositis to internal malignancy. Arch Dermatol 1980 ; 116 : 295-298.

2. Malavia AN, Many A and Schwartz RS : Treatment of dermatomyositis with methotrexate, Lancet 1968; 2:485-488.

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