SELF ASSESSMENT PROGRAMME

A 50 year old male councillor presented with asymptomatic nodular lesions on the buttocks, back and shoulders for 1½ years. The patient was otherwise well. There was no history of loss of sensations or paraesthesias. About a year ago he had an episode of sudden diminution of vision which returned to near normalcy after some local injections into the eye. Physical examination revealed a healthy well built individual with succulent firm cutaneous nodular lesions present on the aforementioned sites. The nerves were not thickened or tender. The axillary and inguinal lymphnodes were firm, moderately enlarged and non-tender. The right eye showed circumcorneal congestion. Systemic examination was normal.

(a) Which of the following diagnoses is most likely ?

1. Sarcoidosis
2. Lepromatous leprosy ? (Histoid)
3. Mycosis fungoides
4. Xanthoma tuberosum
5. Benign lymphoplasias of the skin

(b) Which of the following investigations is most likely to help ?

1. Skin biopsy
2. Skin slit and smear for AFB
3. Serum lipids
4. Serum calcium
5. X-ray chest and hands
6. Mantoux test

Skin biopsy revealed a granulomatous infiltrate composed chiefly of epitheloid cells.

(c) Which of the treatments could be most helpful ?

1. Dapsone
2. Chloroquine
3. Systemic corticosteroid
4. Low fat low carbohydrate diet
5. No treatment

(d) What is the prognosis ?

1. to life
2. to a spread of the disease process

ANSWERS

(a) A combination of eye symptomatology and nodular skin lesions suggested the possibility of sarcoidosis. On the other hand the presence of lymphnodes in association with nodular lesions would favour the diagnosis of mycosis fungoides. The lack of involvement of face or other parts of the body, the absence of any paraesthesia or anaesthesia, the lack of thickening of nerves would make leprosy a relatively unlikely possibility. The lesions were not yellowish to favour the diagnosis of xanthoma; the sites would be unusual for a benign lymphoplasia of the skin.

(b) Skin biopsy would be the most useful investigation to differentiate between various conditions listed above. Histology of xanthomas is unmistakable and consists of large fat-laiden macrophages and often giant cells — the Touton giant cells. The histological architecture of benign lymphoplasia of skin and mycosis fungoides is also distinctive. Sarcoid granuloma is classically described as a 'naked' epitheloid cell granuloma which is indeed what was seen in the biopsy of the skin lesion. This was quite distinct from histology either of tuberculoid or lepromatous leprosy. The other investigations would only be of secondary help and except for elevation of serum calcium were normal.

Further investigations would be directed towards the confirmations of the diagnosis and an assessment of the severity of the disease process. The negative acid fast stain for instance, completely ruled out the possibility of lepromatous (histoid) leprosy. X-rays of the chest and hands which in this case were normal, would be expected to help in assessing the severity of the disease. Positive Mantoux test could not be of diagnostic value anyway, though a negative test may favour the diagnosis of sarcoidosis. Serum lipids — cholesterol or triglycerides would normally be expected to be elevated in most forms of xanthomas.

(c) Having made a diagnosis of sarcoidosis, the choice is between no treatment, chloroquine or corticosteroids. If the skin lesions were the only abnormality, one was perfectly justified in withholding all kinds of treatment, since the lesions are benign and most often regress spontaneously. Chloroquine therapy is of particular value in lupus pernio which was not present in this patient. In view of this ocular involvement and hypercalcemia, it was considered advisable to institute systemic corticosteroid therapy.

(d) Sarcoidosis has in general very good prognosis for life though the disease tends to recur and remit. The warning signals are the involvement of the lungs or the kidneys which in the present case were not affected. Ocular involvement if not treated adequately and at proper time can of course, lead to severe disability.

Comment

Systemic or cutaneous sarcoidosis in this part of the world is a relatively uncommon condition, though a few case reports have appeared from time to time in literature[1,2]. In one of these, the nationality of individuals suffering from sarcoidosis is not clear. Sometimes it becomes difficult to differentiate sarcoidosis from leprosy or the opposite could happen depending upon geographical location[3,4]. Indeed, this patient was earlier diagnosed as suffering from leprosy by a competent dermatologist.

Sarcoidosis can be a multisystem disease that may involve the skin, the lymphnodes, the eyes, the lungs, salivary glands, bones and mucous membranes. The present patient showed involvement of skin and eyes without involvement of any of the other vital organs such as the lungs, or the nervous system. While it is undeniable that many patients with sarcoidosis may spontaneously regress, it seemed advisable to administer systemic corticosteroids in the present patient in view of the ocular involvement. Indiscriminate corticoid therapy in every patient of sarcoidosis should be discouraged since in the absence of involvement of the vital organs or the eye, sarcoidosis remains a benign condition and the prognosis to life, in general is good.

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