SELF ASSESSMENT PROGRAMME

A 55 year old male landlord from Andhra Pracesh presented with recurrent erythematous papulo-nodular lesions on the extremities and trunk for 6 months. The lesions would appear on both the upper extremities and spread to the trunk. These were associated with mild fever. There was no history of joint pains or edema feet. The lesions would subside with antibiotics and analgesics in a couple of weeks time. There was no history of sore throat, ingestion of any medicines particularly sulphonamides or aspirin ; no cough with expectoration ; no anaesthesia, epistaxis or nerve pains. At one stage during his investigations his blood serology for syphilis was found to be reactive (VDRL 1:8) for which he was given Benzathene penicillin–9.6 mega units (1.2 × 8), two months following which the serology was found to be non-reactive. For about 3 months before presenting, the patient was on variable doses of corticosteroids, (Betamethasone 0.5 – 1.5 mg per day). When he reported, the patient had papulo-nodular eruption on extremities for 3 weeks associated with malaise, vague body pains and fever.

On examination he showed the presence of livid red, tender nodular lesions on the trunk and the upper extremities ; a few lesions were present on the face. The temperature was 102°F. The nerves were not thickened nor tender ; there was no loss of peripheral sensations. There was no lymphadenopathy. Slight oedema of ankles was present. The blood pressure was normal. Systemic examination was normal.

Which of the following diagnoses is most likely ?

1. Erythema nodosum
2. Erythema nodosum leprosum
3. Nodular vasculitis
4. Polyarteritis nodosa (systemic Vasculitis)
5. Systemic lupus erythematosus
6. Secondary syphilis

Which of the following investigations would be most helpful ?

1. Anti nuclear factor
2. Serum proteins
3. Skin biopsy
4. Nerve biopsy
5. Antistreptolysin O-titre
6. Blood for STS.

The skin biopsy showed inflammatory changes mostly of non-specific nature confined to the upper dermis, with some mononuclear infiltrate present around the appendages. Blood for serology and ANF were negative. Urine and hemogram were normal.

The patient continued on analgesics with which the lesions improved for a while and then relapsed in a big way.

What further investigation would be of help ?

1. X-ray Chest
2. Urinalysis
3. Repeat skin biopsy
4. Sulphones

What is the prognosis of this patient ?

1. Dangerous to life
2. Irreversible kidney damage
3. Mild recurrent nuisance
4. Self limiting course

ANSWER

It would be extremly difficult to be too confident of clinical diagnosis in this patient, though some of the conditions can be regarded as less likely than others. Secondary syphilis would be an unlikely possibility, because of livid red, tender nodules associated with mild constitutional symptoms in spite of adequate antisyphilitic treatment. There was no other cutaneous or systemic evidence of SLE though the rather capricious character of collagen disorders in general and SLE in particular is well known. The normal blood pressure, lack of pulmonary symptoms or any radiological findings in the chest and lack of eosinophilia make polyarteritis nodosa systemic vasculitis an improbable diagnosis. The trunk and upper extremities are unusual sites for erythema nodosum or nodular vasculitis. The look of the lesion, the tenderness, the distribution, all suggested the diagnosis of erythema nodosum leprosum. The absence of any other signs of lepromatous leprosy however was strong point against the diagnosis, even though the patient came from an endemic area. The skin biopsy and the radiological and haematological investigations were of no help.

A repeat skin biopsy, however, showed presence of AFB in abundance and histological features of ENL.

Because of the severity of the disease, the patient was treated with Thalidomide (300 mg. per. day) to which he responded very well.

Prognosis of this patient, despite the severity of ENL is not bad, since the diagnosis has been made and effective therapeutic measures are available. The patient, of course, has to be treated with specific chemotherapeutic drugs.

Comment :

This patient presents a good example of the so called 'invisible leprosy[1]'. A thorough careful examination had failed to reveal any clinical, bacteriological or histological evidence of leprosy on the first occasion. A high degree of suspicion together with persistence in trying to exclude the possibility of leprosy, clinched the diagnosis histologically and bacteriologically, still without any clinical evidence.

The patient had obviously gone from doctor to doctor (Dermatologists included) without a correct diagnosis having been made and re-emphasises the importance of trying to look for leprosy in rather unusual presentations. Canizares[2] has reported a case of ENL presenting with papulonodular eruption with no other clinical signs of leprosy. Jopling[4] suggested that there may be clandestine ingestion of Dapsone before the patient presents with ENL, and Godal[4] and associates expressed similar opinion. Our patient, a highly intelligent individual emphatically and repeatedly denied such history and we have reasons to beileve that ENL can occur without sulphone therapy. The senior author (LKB) has had occasion of seeing a patient presenting with lymphadenopathy, fever and splenomegaly without any skin lesions. Lymph node revealed macrophages riddled with AFB and a diagnosis of ENL since ENL in retrospect was made. In a country such as India, it is important to keep a very high index of suspicion for suspecting leprosy in any one of its protean manifestations.

References :

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