Sarcoidosis in a Sri Lankan male

G M P Sirimanna¹ and J B C Wijesinghe²

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Introduction

Sarcoidosis is a chronic multisystemic disorder of unknown origin, which affects lungs most frequently. Skin eyes and lymph nodes also are commonly involved. Compared to the west, sarcoidosis is very rare in Southeast Asia.

We report a young Sri Lankan male who presented with skin lesions of sarcoidosis. He had generalised peripheral lymphadenopathy and radiological evidence of bilateral hilar lymphadenopathy. To our knowledge sarcoidosis presenting with skin lesions has not been reported in Sri Lanka before.

Case report

In September 1999, a 39 year old Sri Lankan male presented to the skin clinic of the North Colombo Teaching Hospital with a skin eruption of three months duration. The skin lesions were non-itchy and non-tender. He did not have any other symptoms.

Examination revealed multiple shiny yellowish papules, distributed almost bilaterally and symmetrically over the lumbar region. Their diameters ranged from 1 - 10 mm and shapes were circular or oval. Some coalasced to from plaques.

On general examination he was found to have generalised lymphadenopathy. Enlarged nodes were firm, rubbery, discrete and non-tender. Ulceration was not noted.

Cardiovascular, respiratory, gastrointestinal

and nervous systems were clinically normal. Examination of locomotor system and eyes did not reveal any abnormality.

Investigations revealed the following. ESR was 8 mm/1st hour. Total white blood count was 7800 with N 52%, L 42%, E 6%. Haemoglobin level was 11.8 G/dl. Blood picture was normal. Lipid profile was normal.

Chest x-ray showed bilateral symmetrical hilar lymphadenopathy. Tuberculin test was negative. Sputum for acid fast bacilli was negative on three occasions. Lung function tests including lung volumes, spirometry and diffusion capacity were within normal limits.

Ultrasound examination of the abdomen did not reveal any organomegaly or lymphadenopathy. ECG and echocardiography were normal.

Immunological assessment revealed normal CD4 & CD8 counts. Total serum proteins were 8.2 G/dl; Albumin 4.1 G/dl; Globulin 4.1 G/dl. Serum protein electrophoresis demonstrated an increase of beta and gamma globulins. Serum Immunoglobulin levels were normal.

VDRL and HIV screening test were negative. Serum Calcium and 24 hour excretion of urinary calcium were normal. Angiotensin converting enzyme level was normal.

Histological findings in skin and lymph nodes were identical. Epithelioid cell granulomata containing Langhan's type and foreign body type multinucleated giant cells were numerous. Necro-

¹Consultant Dermatologist, Teaching Hospital, Colombo North.

²Medical Officer, Department of Dermatology, Teaching Hospital, Colombo North.

sis or caseation was not seen. Mild to moderate lymphocytic infiltrations were seen around the granulomata.

Culture for mycobacteria and fungi were repeatedly negative in material obtained from skin biopsy and lymph node biopsy.

Discussion

There are no specific diagnostic tests for sarcoidosis¹. Diagnosis is established most securely when clinical and radiological findings are supported by histologic evidence of widespread noncascating epithelioid granulomata in more than one organ or a positive Kveim test.

In our patient the combination of clinical chest x-ray findings, typical histological findings in skin and lymph node specimens and the presence of skin anergy as evidenced by a negative Tuberculin test were supportive of the diagnosis of sarcoidosis. Kveim test is not available in Sri Lanka and is not in general use at present².

Other diseases such as lymphoma, fungal disease and mycobacterial diseases, which can give rise to similar findings were excluded by investigations.

Skin manifestations are found in 25% of

patients with sarcoidosis. Erythema nodosum is the common non-specific skin lesion seen in sarcoidosis, especially in Europe. Other skin lesions are plaques, maculopapular eruptions, subcutaneous nodules and lupus pernio. Papular sarcoid lesions usually occur on face and extensor aspects to the limbs and are noted to be especially common among Afro-Carribeans³. Our patient's lesions were waxy yellow papules distributed on the back. Use of potent topical steroids is an accepted method of treatment in skin lesions of sarcoidosis. With three months of topical steroids the lesions became flattened and less firm.

Enlarged lymph nodes in our patient were firm, rubbery, discrete, mobile and non-tender as typically described in sarcoidosis. Considerable reduction in the size of the lymph nodes occurring spontaneously was evidenced over a follow up period of ten months. This is a commonly observed feature in sarcoidosis.

Bilateral hilar lymphadenopathy is the best recognized x-ray finding in sarcoidosis. It is classified as the stage I of thoracic sarcoidosis. It is usually considered as a benign reversible manifestation. However 1 in 10 patients with bilateral hilar lympathadenopathy will progress to chronic sarcoidosis. During the follow up period of ten months no further pulmonary changes were noted and the lung function tests including diffusion capacity were normal.



Figure 1. Yellowish papules on the back.

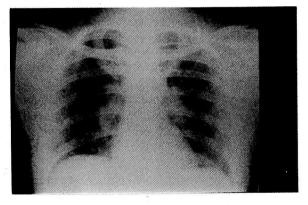


Figure 2. Chest X ray showing bilateral hilar lymphadenopathy.

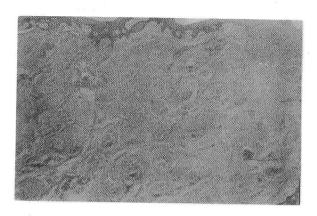


Figure 3. Granulomata in the skin. (H & $E \times 40$)

Sarcoidosis is rare in Southeast Asia. In 1954 a case of sarcoidosis affecting the orbit has been reported in Sri Lanka⁴. To our knowledge, occurrence of sarcoidosis affecting the skin has not been documented previously in Sri Lanka. It is important to recognize the skin lesions of sarcoidosis. Differential diagnosis of sarcoidosis has to be considered in skin lesions showing granulomatous histology. Recognition of the skin lesions and regular follow up may save the patients from potentially life threatening systemic complications of the disease.

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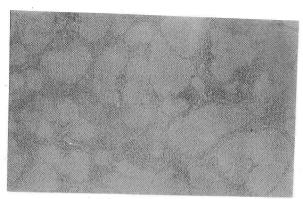


Figure 4. Granulomata in the lymphnode. (H & $E \times 40$).

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