PLEURAL SARCOIDOSIS

By J.L. Da Costa* and S.C. Chiang

SYNOPSIS

Histologically-proven sarcoid involvement of the pleura is documented in a 23-year old Indian female for the first time. The presenting features were pyrexia, arthralgia affecting the hands, elbows and ankles with subsequent development of bilateral hilar lymphadenopathy.

The patient is now asymptomatic but the hilar lymphadenopathy persists. It is noted that sarcoidosis is uncommon in S.E. Asia and the rarity of pleural sarcoidosis is emphasized.

Pleural involvement is rare in sarcoidosis (Scadding, 1967). Since the original report by Nickerson (1937), histologically-proven sarcoid involvement of the pleura has been reported in only four other cases (Berte and Pfotenhauer, 1962; Kovnat and Donohue, 1965 and Macquet *et al*, 1965). Sarcoidosis is uncommon in S.E. Asia (Da Costa, 1973) and it is therefore of interest to document a further case of sarcoidosis with pleural involvement in an Indian female.

CASE REPORT

A twenty-three-year old Indian female was admitted to hospital in March 1973 with a history of fever for the past 4 months associated with arthralgia involving the metacarpophalangeal joints of both hands, the elbows and ankles. Clinically she had a low grade fever and was asthenic but otherwise no other abnormality was detected. Investigations revealed a haemoglobin of 11 G%, E.S.R. 135 mm./hour, total W.B.C. 8,500/c.mm. (normal differential count), blood rheumatoid factor negative. L.E. cells negative, blood urea 32 mg.%, serum proteins 7·6 G% (albumin 3·2 G%, globulin 4·4 G%), serum calcium 10 mg.%, serum phosphate 3·4 mg.%. The electrocardiogram was within normal limits. Chest and abdominal radiographs were normal.

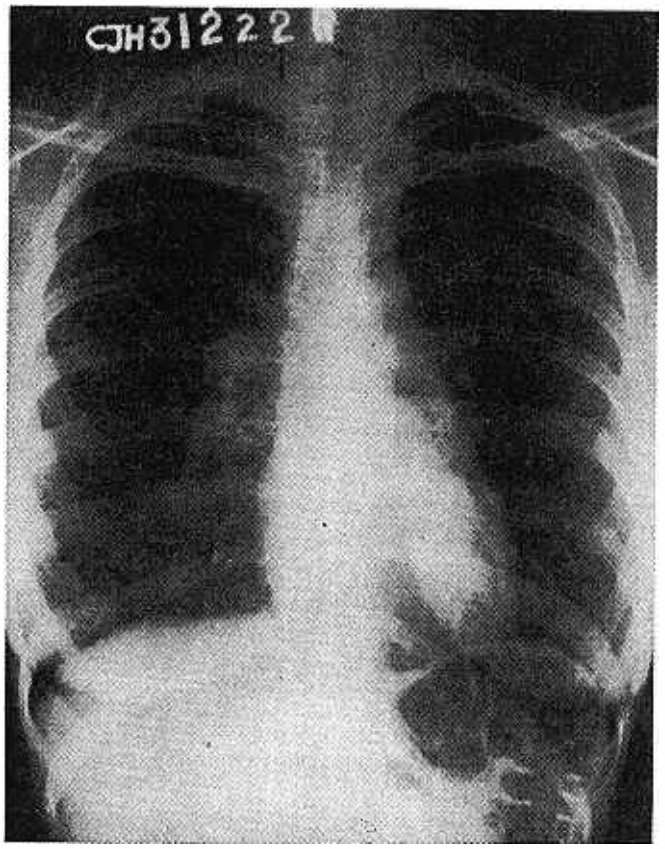

Fig. 1. Chest radiograph (P—A view) of patient showing bilateral hilar lymphadenopathy. No parenchymal lesions are visible.

She was thought to be probably suffering from a mild rheumatoid arthritis and was given phenylbutazone 200 mg. t.d.s. with good symptomatic improvement. She was discharged asymptomatic after one week with the E.S.R. down to 32 mm./hour. Subsequently, in April 1974, she complained of a dry persistent cough for 3 weeks. Clinically, no abnormality was detected but a chest radiograph revealed

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bilateral hilar lymphadenopathy without any parenchymal lesions (Fig. 1). A Mantoux test (PPD 10TU/ml.) was negative. Lung function tests showed normal dynamic lung volumes with a mild decrease in the transfer factor.

A right antero-lateral thoracotomy was performed. At operation the hilar lymph nodes were found to be enlarged, hard and gray in colour. Multiple, gray, hard nodules, 2-3 mm. in diameter, were seen scattered over the visceral and parietal pleura. The right lung and pleura felt gritty on palpation. A right hilar lymph node and the lateral edge of the middle lobe were taken for histological examination.

Histological examination of the hilar lymph node (Fig. 2) revealed almost complete replacement of the lymph node by whitish firm tissue composed of confluent non-caseating granulomata. Acid-fast bacilli were not detected.

The lung biopsy specimen showed multiple small hard nodules studded on the pleural surface (Fig. 3). A cut section showed similar nodules in the underlying lung parenchyma. Histologically these nodules were found to be made up of confluent non-caseating granulomata with multinucleated Langhans' giant cells, epithelioid cells and histiocytes (Fig. 4). Acid fast bacilli were not detected.

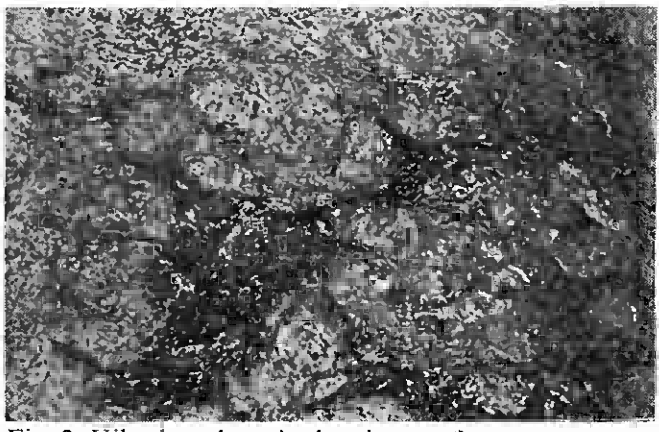

Fig. 2. Hilar lymph node showing confluent non-caseating granulomata (Haematoxylin and eosin stain, ×40).

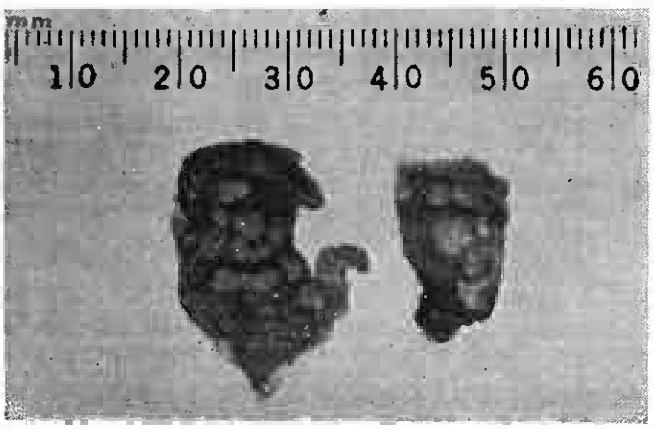

Fig. 3. Lung biopsy specimen showing multiple gray hard nodules studded on the pleural surface.

The intradermal Kveim test (Murdock antigen) was reactive at 5 weeks (Fig. 5). Follow-up at 6 months on symptomatic treatment shows persistence of the hilar lymphadenopathy. No lung parenchymal lesions are visible on the

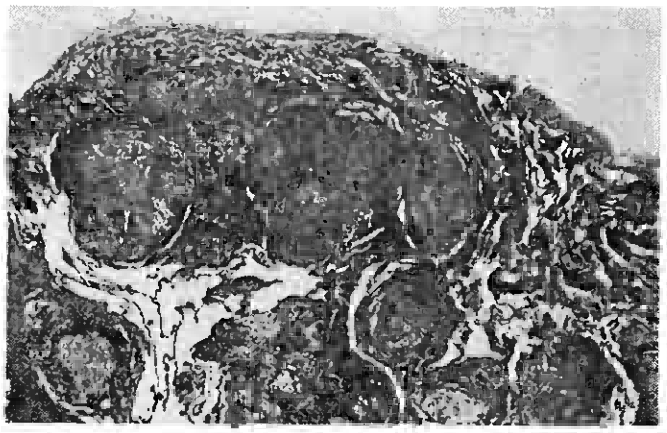

Fig. 4. Lung biopsy specimen showing non-caseating granulomata involving the pleura (Haematoxylin and eosin, × 27).

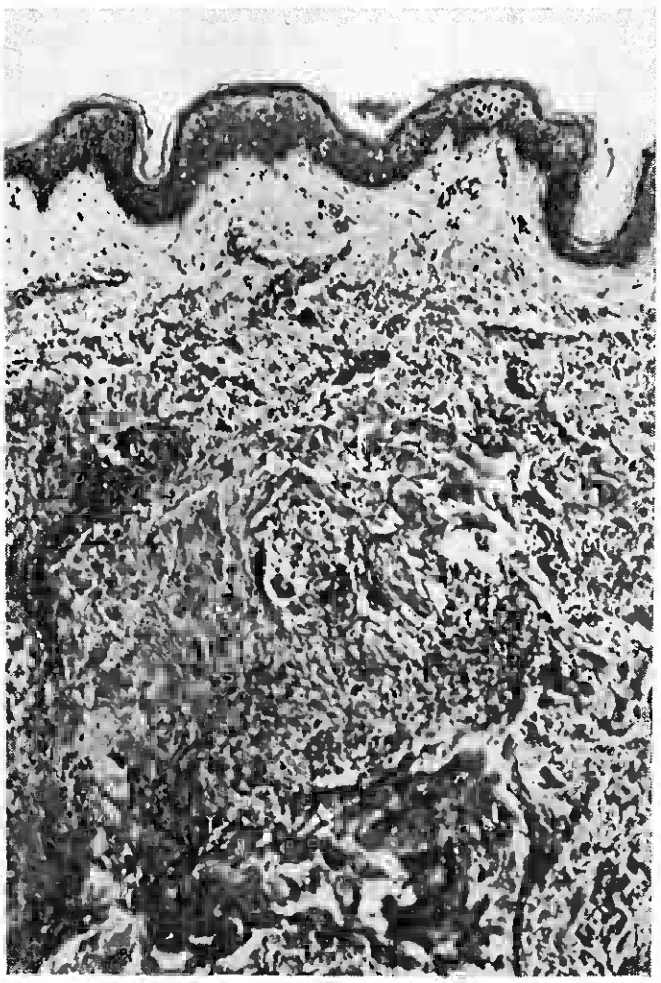

Fig. 5. Skin biopsy specimen at site of positive Kveim test showing few non-caseating granulomatous foci with multinucleated giant cell, epithelioid cells and histiocytes. (Haematoxylin and eosin stain, × 27).

chest radiograph. The patient feels well and is asymptomatic.

DISCUSSION

Nickerson (1937) reported the necropsy findings in 6 cases of sarcoidosis and described a case in a 58-year-old negress with irregular nodules coalescing to form plaques scattered over the parietal pleura and pericardium; histologically these showed sarcoid granulomata. Scadding (1967) in a detailed review of 275 cases of sarcoidosis did not document any case of pleural involvement with sarcoid granulomata although he described a case of sarcoidosis with an associated pleural effusion. Pleural sarcoidosis is thus seen to be extremely uncommon. As sarcoidosis in S.E. Asia is rare per se, the present case in an Indian female is thus unique. The other presenting features in this case, namely, arthralgia, fever and bilateral hilar lymphadenopathy with a positive Kveim test were characteristic of sarcoidosis, the pleural involvement being detected coincidentally at operation.

Other associated pleural changes have been noted in sarcoidosis e.g. pleurisy due to intercurrent infective inflammatory episodes and spontaneous pneumothorax due to rupture of an emphysematous bulla, especially in the fibrotic stage (Scadding, 1967). However, these changes are not directly due to sarcoid involvement.

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