

SARCOIDOSIS: A REPORT OF TWO CASES

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SYNOPSIS

Sarcoidosis is a rarity in South East Asia. Only two cases were diagnosed in the University Hospital between the period January 1967 to April 1977. Both the cases were Indian females. Review of previously documented cases of sarcoidosis from Malaysia, Singapore and Brunei suggests that there is a greater susceptibility among the Indians as compared to other ethnic groups.

INTRODUCTION

The incidence of sarcoidosis shows geographical and racial variation. It is relatively common in Western countries and is rarely seen in certain ethnic groups e.g. Maoris in New Zealand, North American Indians and Chinese. Previous publications (Tang, Khoo and Tan, 1964; Snelling and Chooi, 1966; Da Costa, 1973 and 1975; Ong, Chew and Chiang, 1975; Poh and Ong, 1976) have emphasised the rarity of sarcoidosis in our geographical areas.

To date, two cases of sarcoidosis were diagnosed in the University Hospital between the period January 1967 to April 1977. Both the cases were Indians females. Review of previous publications show only a total of seven cases have been documented in Malaysia, Brunei and Singapore. With the exception of one Malay case from Brunei and one Chinese case from Singapore all the remaining cases were Indians. As the Indians constitute less than ten per cent of the population in the geographical area of Malaysia, Brunei and Singapore, it appears that there is a greater susceptibility among the Indians as compared to other ethnic groups.

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CASE REPORTS

Case 1

A forty-three year old Indian female was admitted to the University Hospital in December 1973 with a history of

bilateral ankle swelling for the past one month. She did not have cough or dyspnoea. Clinically she had moderate anaemia and minimal pitting oedema of both legs. The cervical and axillary regions lymph nodes were enlarged. The liver was enlarged 5 cm below the costal margin and the spleen was enlarged 3 cm below the costal margin. No other abnormality was detected. Investigations revealed a haemoglobin of 6.0 G%, E.S.R. 45 mm/hour, total W.B.C. 5,200/c.mm (normal differential count). Chest radiograph revealed bilateral hilar and right paratracheal lymph nodes enlargement. The lung parenchyma was normal. The electrocardiogram was within normal limits. L.E. cells negative, blood urea 30 mg%, serum protein 7.4 G% (albumin 2.6 G%, globulin 4.8 G%), serum calcium 9 mg% and serum phosphate 3.3 mg%. The peripheral blood film suggested a iron deficiency anaemia. The serum iron 82 ug/100 ml, total iron binding capacity 330 ug/100 ml, serum folate 4.7 ng/ml and serum vitamin B₁₂ 532 pg/ml. The urine examination was normal.

Biopsy of the cervical lymph node and the liver was taken. Histological section of the cervical lymph node (Fig. 1) revealed almost complete replacement of the lymph node by confluent non-caseating granulomata. Acid-fast bacilli and fungal organisms were not detected. Histological section of the liver revealed multiple discrete non-caseating granulomata in the portal tracts. The Mantoux test was negative twice. Acid-fast bacilli was not detected in the sputum. The intradermal Kveim test was not performed as the antigen was not available.

While the patient was in the ward a transient erythema nodosum was observed on both legs. The anaemia of the patient improved with Fe SO₄ and Vitamin B complex. The cortico-steroid therapy was withheld. The patient was followed-up for two subsequent years. She was asymptomatic. The chest radiographs showed normal lung parenchyma and there was some degree of regression in the size of hilar lymph nodes.

Case 2

A fifty year old Indian female was admitted to the University Hospital in November 1975 with a history of exertional dyspnoea for the past ten months. She had cough with expectoration of whitish sputum for the past one week. Clinically there was intercostal recession during inspiration. There were crepitations on both bases. The liver was palpable 3 cm below the costal margin. Clinically no other abnormality was detected.

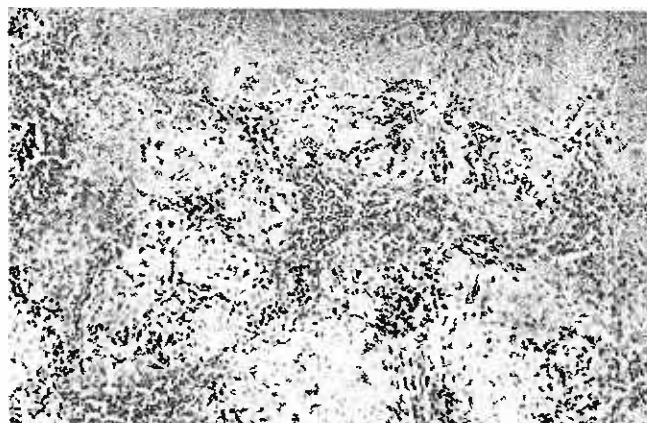


Fig. 1. Cervical Lymph Node. Confluent non-caseating granulomata. (Haematoxylin and eosin stain, X 40).

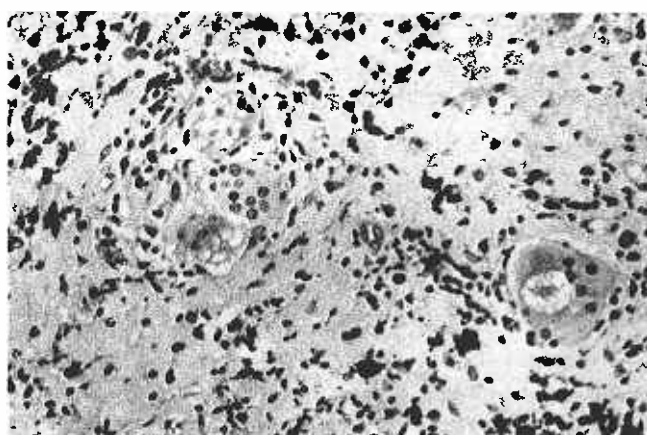


Fig. 2. Cervical Lymph Node. Higher magnification of non-caseating granulomata. (Haematoxylin and eosin stain, X 150).

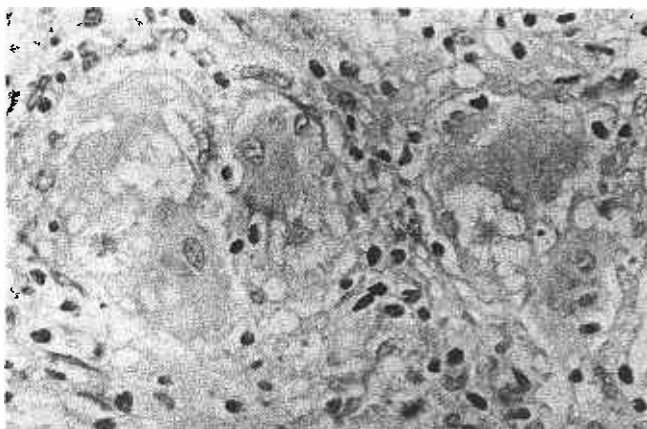


Fig. 3. Lung. Area of fibrosis with multinucleated giant cells and lymphocytes. A giant cell with an asteroid body. (Haematoxylin and eosin stain, X 250).

Chest radiograph revealed infiltrates in both lung fields suggestive of interstitial fibrosis. Investigations revealed a haemoglobin of 14.1 G%, total W.B.C. 10,800/c.mm (normal differential count),

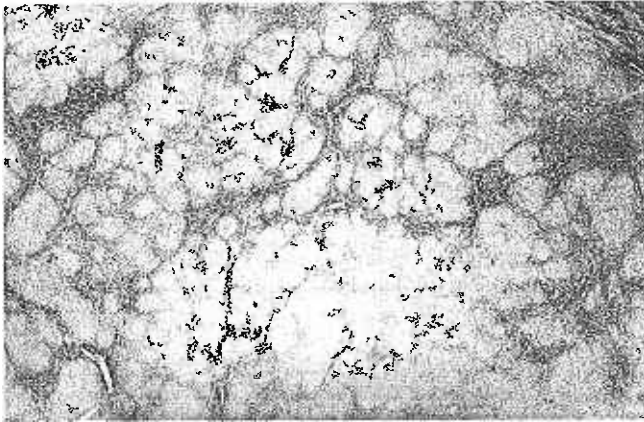


Fig. 4. Liver. Non-caseating granulomata with three multinucleated giant cells showing asteroid bodies. (Haematoxylin and eosin stain, X 250).

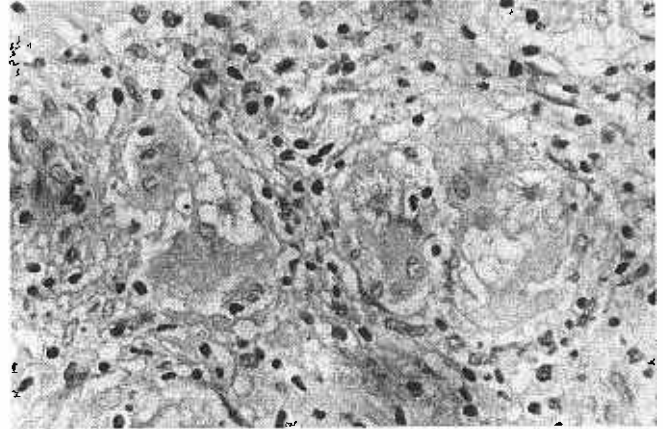


Fig. 5. Liver. Higher magnification showing the asteroid bodies in multinucleated giant cells. (Haematoxylin and eosin, X 500).

L.E. cells negative, blood rheumatoid factor negative and anti-nuclear antibodies negative. Blood urea 20 mg%, serum protein 7.9 G% (albumin 3.4 G%, globulin 4.5 G%), serum calcium 9.2 mg%, serum phosphate 2.8 mg%. The electrocardiogram was within normal limits.

Drill biopsy of the lung parenchyma revealed interstitial fibrosis and inflammatory cells comprised of lymphocytes and plasma cells. Multinucleated giant cells with well defined asteroid bodies were present (Fig. 2). Schaumann's bodies were present.

The liver biopsy revealed non-caseating granulomata in the portal tracts. Multinucleated giant cells with asteroid bodies were present (Fig. 3). Acid fast bacilli and fungal organisms were absent in the sections.

The Mantoux test was negative and the sputum for acid fast bacilli was negative. The intradermal Kveim test was not performed as the antigen was not available. Treatment with prednisolone resulted in considerable clearing of the lung fields. The patient was followed up for one year.

DISCUSSION

Sarcoidosis is a systemic disease and in order to establish its presence, systemic involvement must be demonstrated. In both these cases biopsies were performed on two different anatomical sites. The clinical and radiological features were consistent with the diagnosis. In view of the rarity of sarcoidosis, there was careful exclusion of tuberculosis which may closely simulate sarcoidosis. The Mantoux test was negative in both these cases and all the seven cases previously published from this region. Kveim test was not performed on both the cases as the antigen was

not available. However, the Kveim test is positive in only 60 to 80% of patients with sarcoidosis.

Although the aetiology of sarcoidosis is still unknown, the epidemiological studies suggest that genetic and environmental factors are important. The causative factor, if there be a single one, remains unknown. It is probable that no single aetiological agent exists, and that the histological reaction is a common response to a number of differing causative agents. Alternatively, there may be a specific sensitivity to a particular antigen, as yet unidentified, which develops only in genetically predisposed individuals. The occupational history of all the seven Indian patients was diverse and did not reveal any incriminating factors.

It appears that although sarcoidosis is a rarity in our geographical region, there is a greater predilection among the Indian ethnic group which may be related to genetic factors.

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