A case of sarcoidosis in a Chinese woman

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Summary
A case of sarcoidosis in a Chinese woman presenting with hilar lymphadenopathy is described. In addition, there were neurological features, a depressed tuberculintest and a positive Kveim test. The condition is rare in the Chinese.

Sarcoidosis is a systemic granulomatous disease of undetermined aetiology and pathogenesis. The condition was first recognized by Hutchinson, who published a clinical description of one case in 1875 and of two others in 1898. Boeck was the first to describe, in 1899, the histological characteristics of the skin lesions that have since come to be associated with his name as Boeck’s sarcoid (Hsing, 1964). Chapman (1955) stated that the Chinese people in Taiwan and overseas are regarded to a very large degree to be exempted from the disease. It appears that sarcoidosis is extremely rare in South East Asia and in Singapore in particular (Da Costa, 1973). Japan is the only Asian country in which a sizable number of cases of sarcoidosis have been observed.

Probably the first proved case of sarcoidosis among the Chinese was reported by Hsing (1964).

References
Khoo et al. (1964) described two cases in Indians. Three cases of pulmonary sarcoidosis were reported by Snelling from Malaysia in 1966. One of the cases was a Malay and two were Indians.

Case report

A 42-year-old Singapore-born housewife was admitted on 21 September 1973 from another hospital for investigations of a right hilar shadow on the chest X-ray. She gave a history of weakness over the right side of her face for 2 months. She had lost approximately 10 lb in weight and also experienced some difficulty with swallowing for 3 days before her admission. She was in good health until her present illness.

On clinical examination her general condition was good and she was afebrile. The blood pressure was 140/90 mmHg. The heart and lungs were clinically normal. In the abdomen the uterus was enlarged. The liver and spleen were not enlarged. She had an obvious right facial palsy of lower motor neurone type and had paralysis of the tenth nerve on the left side with weakness of the palate. The motor power was grade 4 in the lower limbs. The tendon reflexes were absent in both legs and the plantar responses were flexor. She had no sensory loss in the extremities although she complained of paraesthesiae in the legs and was unable to stand from a squatting position without help.

Her uterine enlargement was diagnosed as a uterine fibroid by the gynaecologist. Specialized nasopharyngeal examination excluded the presence of a nasopharyngeal carcinoma.

The other investigations done revealed an Hb of 10.9 g/100 ml, WBC 4500; polymorphs 80%; lymphocytes 15%; monocytes 3%; and eosinophils 2%. The ESR was 20 mm. The urinalysis was normal and the blood urea was 24 mg%. The serum uric acid was 5.4 mg%; serum albumin/globulin 3.6/3.5 g%—serum calcium 9.6 mg% and the serum phosphate 5.1 mg%. The sputum cultures and smears were negative for acid-fast bacilli. The serum protein electrophoresis was normal. A lumbar puncture was normal. The electrocardiogram was within normal limits. A chest X-ray revealed a right hilar mass and a tomogram confirmed the presence of hilar lymph node involvement. The provisional diagnosis was a carcinoma of the lung with metastases and a peripheral neuropathy. The other diagnosis considered was mediastinal Hodgkin's disease.

Bronchoscopy was normal and at thoracotomy enlarged rubbery hilar and mediastinal lymph nodes were found. Lymph nodes were also present among the pulmonary vessels, below the carina and alongside the trachea. Altogether 40 g of lymph nodes were removed. The histology showed lymph nodes filled with numerous granulomata consisting of epitheloid cells. A few Langhan giant cells were present. An occasional granuloma had central necrosis. No acid-fast bacilli were seen. The picture was highly suggestive of sarcoidosis (Fig. 1).

A Kveim test was done by injecting intradermally 0.1 ml of a saline suspension Kveim antigen into the forearm. After 5 weeks, a nodule 5 mm in diameter was removed for histology. The section showed granulomatus areas. In these areas epitheloid cells, lymphocytes and a few giant cells were seen. No caseation was present (Fig. 2). The tuberculin test was non-reactive.

Three weeks after operation she developed a left

![Fig. 1. Section showing granulomata with a Langhans giant cell.](http://pmj.bmj.com/first-published-as-10.1136/pgmj.51.594.257-on-1-april-1975)
facial palsy and she had residual signs of a right sided facial weakness which had improved markedly. Her muscle power in the legs had also improved.

Nerve conduction velocities were studied on the median, ulnar, lateral popliteal and posterior tibial nerves. There was reduction in the sensory, motor, and mixed conduction velocities in these nerves. Electromyographic and nerve conduction studies indicated the presence of a peripheral neuropathy.

Following surgery these studies were repeated and there was improvement in the nerve conduction times.

Comment

The aetiology of sarcoidosis is still a matter of debate. Systemic sarcoidosis has been thought to be an atypical reaction to tubercle bacilli, but it seems more relevant that it is a chronic granulomatous process produced by some undiscovered agent or agents. The disease progresses and regresses irregularly in a remarkable protean pattern. It often has widespread organ localization with little or no constitutional manifestations. The chief localizations are the hilar glands and the lungs. In a few, hilar enlargement may appear unilateral and the diagnosis in these is sometimes made at exploratory thoracotomy. Erythema nodosum is frequently associated with thoracic lymphadenopathy but our patient did not have any skin lesions. The majority of those with thoracic lymphadenopathy show spontaneous regression of lymphadenopathy. Lymphadenopathy may persist without appreciable change and is usually associated with chronic and apparently irreversible lesions in other organs. There was no evidence of pulmonary involvement in this case and the lung function was normal.

The patient's eyes were examined and they were normal. The X-rays of her hands revealed no abnormality. In sarcoidosis the serum calcium may increase and hypercalciuria is not unusual. In this patient the serum calcium was normal. The serum immunoglobulins were IgG, 1960 mg/100 ml (770–1510); IgA, 304 mg/100 ml (134–297); IgM, 70 mg/100 ml (67–208).

Neural involvement is not uncommon and in our patient there was evidence of nervous system involvement. The bilateral facial paralysis, weakness of the palate, the absent reflexes in the lower limbs due to peripheral nerve involvement have all been reported previously in sarcoidosis (Colover, 1948). When the facial paralysis is bilateral, the interval between the onset of paralysis on the two sides is a matter of days, but occasionally this period may be many months. When there is paralysis of the soft palate, the latter is often more marked on one side. The pathology of the peripheral nerve lesions was described by Mazza (1908). The nerves involved were irregular in calibre. Histologically there were lesions composed of endothelial cells, lymphocytes, mononuclears, plasma cells and giant cells between the nerve fibres, many of which were disrupted. In our patient the nerve conduction times showed some improvement following surgery and before the commencement of steroid therapy. The patient was started on prednisolone 30 mg a day.

The significant findings of this case consist of cranial nerve involvement, a peripheral neuropathy, a depressed tuberculin test and a positive Kveim test, in a patient with hilar gland involvement.
Case reports

Behçet’s disease and the alimentary tract

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Summary

Two patients with perforation of the alimentary tract in Behçet’s disease are described. One of these is believed to be the first recorded case of oesophageal perforation in this condition, while the second is one of the few survivors of bowel perforation. The literature, aetiology and treatment are reviewed.

The association of inflammatory disease of the eye with oral aphthous ulceration was first reported by Adamantiades in 1931. Behçet (1937, 1938) extended the description to include genital ulceration, and this group of symptoms was generally known as Behçet’s syndrome, or Behçet’s triple symptom complex. During the 1950s the association of Behçet’s syndrome with neurological disease became recognized (Alemà and Magni, 1952; Hermann, 1953; Viane, 1957; Alemà and Bignani, 1966), and polymorphous skin manifestations are well known (Marchionini and Muller, 1966; Nazzaro, 1966). Berlin (1960) emphasized the view of Behçet’s disease as ‘a multiple symptom complex’. The most extensive and numerous recent reports have come from Japan where the disease is remarkably common (Oshima et al., 1962, 1963; Shimizu, 1970, 1971), yet reported cases of alimentary perforation remain rare. The following case reports include one of oesophageal perforation, believed to be the first on record.

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Case 1

A 52-year-old housewife first presented in July 1960 with a 3-month history of mouth ulceration, itching eyes and lesions on both arms and legs resembling erythema nodosum. She was treated with Neocortef eye drops and oral prednisone for 3 months. In August she had indigestion, and a barium meal demonstrated an ulcer on the lesser curve of the stomach.

She was then well until June 1961 when there was a recurrence of mouth ulceration, episcleritis and erythema nodosum followed by jaundice. The latter was considered to be due to ascending cholangitis and rapidly settled on antibiotics. Over the next 3 years she had repeated episodes of abdominal pain, sometimes accompanied by vomiting and diarrhoea. Altogether four barium meal and follow-through examinations were performed; one showed a duodenal ulcer and all showed strikingly coarse jejunal folds with dilatation of the upper jejunum. In September 1964 an exploratory laparotomy showed all viscer a other than the small bowel to be normal. Throughout the small bowel, from 12 cm below the duodeno-jejunal junction to 7-5 cm above the ileocaecal junction, there were multiple discrete lesions 3-5 mm in diameter with radiating folds and a thickened edge; it was thought that these were of different ages as some appeared fibrotic, some plum-coloured and some bright red. At one point there appeared to have been a sealed perforation.

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References


