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Short reports

Superior vena caval obstruction caused by sarcoidosis

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In contrast to the early decades of this century a benign cause for the superior vena caval syndrome is nowadays uncommon.[1] We describe a case of superior vena caval obstruction caused by sarcoidosis, the second such case to be reported.

Case report

A 59-year-old widow presented with a three-year history of increasing dyspnoea. She had had rheumatic fever when a teenager and a heart murmur had been noted during her fifth pregnancy. Examination revealed an overweight lady with congestion, swelling and cyanosis of the head, neck, and upper limbs, mild proptosis, and numerous dilated superficial veins on the upper chest. The cervical lymph nodes were firm and moderately enlarged. The right lung base was dull to percussion and breath sounds were absent. Crackles were audible throughout the lung fields. The pulse was irregular and of low amplitude and precordial auscultation confirmed the presence of mitral stenosis.

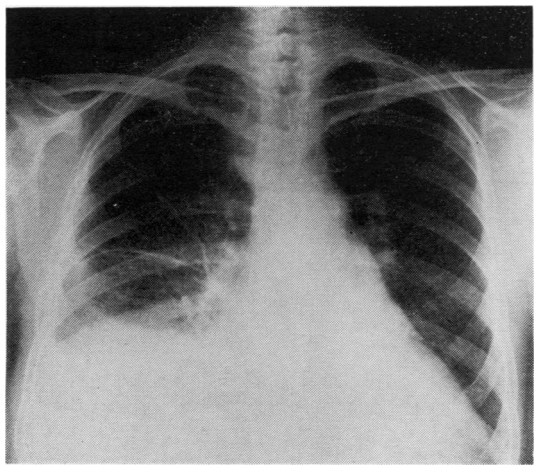

Fig 1 *Chest radiograph showing right pleural effusion, right mediastinal mass, and enlarged left atrium.*

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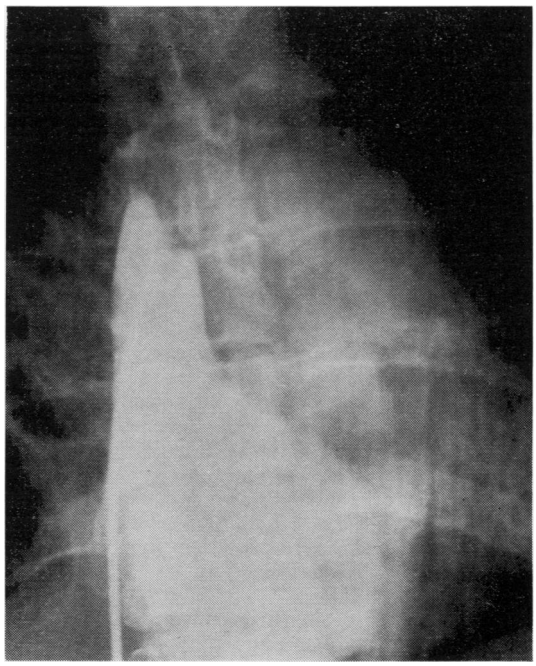

Fig 2 *Superior vena caval compression. The contrast material was injected from below via a femoral vein catheter.*

The ESR was 48 mm/hr (Westergen). The Mantoux test (1 in 10 000) was negative and a Kveim test was positive. Chest radiographs (fig 1) showed a right pleural effusion, pulmonary venous congestion, left atrial enlargement, and a right mediastinal mass. Tomography confirmed enlargement of the hilar and mediastinal lymph nodes, especially the right paratracheal group. A cavogram showed severe narrowing of the superior vena cava (fig 2). The contrast material was injected from below via a femoral vein catheter. Electrocardiography, echocardiography, and cardiac catheterisation confirmed the presence of mitral stenosis, a mobile valve and rapid atrial fibrillation. Biopsy of the supraclavicular nodes showed noncaseating granulomas typical of sarcoidosis. At mediastinotomy through the bed of the third right costal cartilage, a mass of firm enlarged nodes was

found encircling the superior vena cava. Biopsy again demonstrated sarcoidosis. Repeated microscopy (including special stains) and culture of sputum, pleural fluid, and lymph nodes for fungi and mycobacteria were negative.

The patient was given digoxin, diuretics, and steroids with slight improvement. At closed mitral valvotomy a tightly stenosed valve (0·6 cm^2) was split. Postoperative recovery was slow, and three weeks later she developed a urinary infection which was treated with sulphonamides. Unfortunately within one week she suffered profound bone marrow depression (attributed to the sulphonamide therapy) and died of septicaemia on the twenty-ninth day after operation. Necropsy revealed a mass of firm nodes obstructing the superior vena cava. The heart and lungs showed changes typical of rheumatic mitral stenosis. Postmortem histology confirmed sarcoidosis of the lymph nodes; there was no evidence of sarcoidosis in any other tissue examined.

Discussion

Only one previous case of superior vena caval obstruction caused by sarcoidosis has been recorded.[2] In that case we note that no Kveim test was performed and that regression of the obstructing mediastinal mass occurred during treatment which included antituberculous therapy. Diagnosis of sarcoidosis and in particular differentiation from fungal or tuberculous granuloma can be difficult. Reversal of tuberculous superior vena caval obstruction during antimycobacterial therapy has recently been described.[3] In our case we feel that the combination of a negative Mantoux test, a positive Kveim test, repeatedly negative cultures and microscopy for tuberculosis and fungi and detailed necropsy studies, all support a diagnosis of sarcoidosis.

In view of the prominence of lymphadenopathy in sarcoidosis, it is perhaps surprising that compression effects are not more common. Compression of the pulmonary artery by enlarged hilar nodes was reported by Westcott and DeGraff[4] and bronchial compression by Talbot *et al.*[5]

In the presence of severe superior vena caval obstruction, investigations are not without risk. In cases where there is an obvious underlying malignancy and severe obstruction, immediate antitumour therapy is indicated, without resorting to hazardous investigations. In other cases, however, a fuller investigation and accurate diagnosis are desirable. The injection of contrast material into an obstructed high pressure venous system can be distressing and may cause haemorrhage. Injection of contrast material below the block via a femoral vein catheter was used in our patient. Right mediastinotomy was used in preference to cervical mediastinoscopy as it gives a more direct access to the area of the superior vena cava and the risk of haemorrhage is probably less. Since the overwhelming majority of cases of superior vena caval obstruction are caused by advanced intrathoracic malignancy, treatment is palliative in the form of radiotherapy or chemotherapy. Diuretics and steroids usually provide some symptomatic relief. In benign superior vena caval obstruction, conservative management is indicated, the obstruction being well tolerated and likely to improve with time. Successful surgical relief of superior vena caval obstruction awaits the development of a non-thrombogenic graft and recent attempts are encouraging.

We thank Mr B Moore for allowing us to publish details of this patient.

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