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Familial sarcoidosis presenting with stridor

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Sarcoidosis uncommonly presents with bronchial narrowing, produced by mucosal lesions or external compression by enlarged glands. We report two sisters with sarcoidosis who presented with severe inspiratory stridor as a result of obstruction of the major airways. An unusual feature of the second case was unilateral glandular enlargement. This produced such severe obstruction that mechanical dilatation was necessary to relieve the dyspnoea.

Case reports

PATIENT 1

A 26-year-old accounts clerk presented with a three-month history of increasing dyspnoea, non-productive cough, and lassitude. Although she smoked 20 cigarettes per day, there was no history of previous respiratory disease. On examination, she was severely breathless at rest, with a marked inspiratory stridor. The chest radiograph showed bilateral hilar lymphadenopathy and pulmonary infiltrates. Tomograms confirmed the hilar lymphadenopathy. Sarcoidosis was diagnosed and, in view of the severe dyspnoea, treatment was started with prednisolone, 40 mg daily. However, after a week's treatment the severe dyspnoea was unchanged. A bronchoscopy was done to determine the degree of the airway narrowing and to confirm the diagnosis. The carina was greatly widened and the right and left main bronchi were markedly narrowed by external compression. A bronchial wall biopsy confirmed the diagnosis of sarcoidosis. Soon afterwards, she began to improve. The prednisolone was gradually withdrawn over the next 12 months by which time the hilar enlargement was no longer present but there was persistent infiltration at the base of the left lung.

PATIENT 2

The 30-year-old sister of the previous patient presented two years later with a 12-month history of increasing breathlessness and a non-productive cough. Examination at this stage revealed persistent wheezing over the left lung. A chest radiograph showed left hilar enlargement and de-aeration of the left upper lobe. Tomograms confirmed marked glandular enlargement at the left hilum and a grossly narrowed left main bronchus (figure). A bronchoscopy was performed to confirm the diagnosis and exclude tumour. It revealed a greatly broadened carina; the right main bronchus was normal but the left main bronchus was markedly narrowed by external compression. Bronchial biopsy confirmed the diagnosis of sarcoidosis.

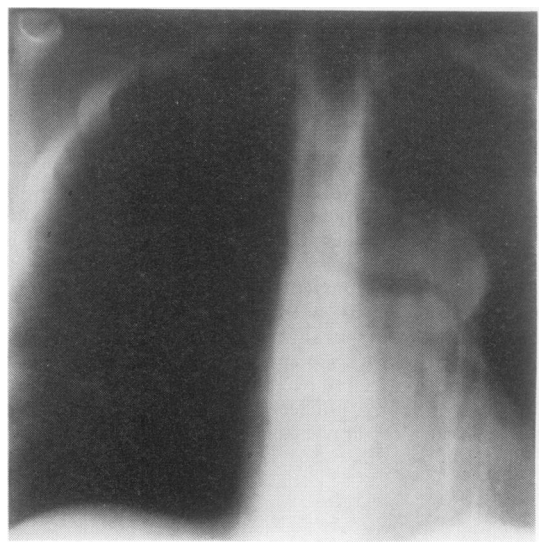

Figure *Patient 2. Tomogram at 12 centimetres showing enlarged left hilar glands and grossly narrowed left main bronchus.*

Treatment with prednisolone produced temporary relief only. Within 12 months she was again severely dyspnoeic with a marked stridor and high pitched wheezes over the whole of the left lung. Bronchoscopy, under general anaesthetic, revealed rigidity and narrowing of the left main bronchus. The lumen of the left upper lobe bronchus could not be clearly identified and the diameter of the left lower lobe bronchus was narrowed to about 4 mm. The left main and lower lobe bronchi were then dilated using Eder-Puestow dilators. There was an immediate, marked improvement in her condition. Within three months the stridor had returned but repeat dilatation again produced good symptomatic relief. She remains well at follow-up four months later.

Discussion

Pulmonary sarcoidosis commonly presents with bilateral lymphadenopathy or pulmonary infiltration and is

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usually asymptomatic. Unilateral hilar lymphadenopathy is unusual, occurring in about one per cent of cases.[1] Normally, it has the same good prognosis and tendency to resolution as the bilateral disease. However, bronchoscopy is indicated to exclude malignant disease which, of course, is far more common and may even coexist with sarcoidosis.[2]

The presentation with stridor, itself rare, is especially noteworthy in that it occurred in two sisters. These two patients constitute another example of like-sex pairings among familial examples of sarcoidosis. Like-sex pairings are significantly more common than unlike-sex pairings even when the preponderance of females with the disease is taken into account.[3] There is no adequate genetic explanation for these findings, although it has been suggested that there is a recessive mode of inheritance for susceptibility to sarcoidosis.[4]

Stenosing sarcoidosis of the bronchi is a clear indication for corticosteroid therapy. Although treatment with prednisolone achieved a satisfactory response in the first patient, it produced only temporary relief of her sister's dyspnoea and the disease progressed to produce marked stridor. Mechanical dilatation was then necessary to achieve an improvement. It would appear that this is an effective way of overcoming localised obstruction caused by sarcoidosis[5] and, when corticosteroid therapy fails to relieve bronchostenosis, repeated mechanical dilatations may be necessary to alleviate severe dyspnoea.

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