Exertional dyspnoea and nonproductive cough in a 22-year-old man

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A 22-year-old man presented with a 2-year history of nonproductive cough and exertional dyspnoea. He had never smoked. Because of a diagnosis of presumed bronchial asthma about 2 years ago, he was treated with multiple bronchodilators and inhaled corticosteroids for several months but no improvement in his symptoms resulted. One year prior to admission, he had been admitted to another hospital with same complaints and pulmonary tuberculosis had been suspected. Therapy with isoniazid (300 mg/d), rifampicin (600 mg/d), and ethambutol (25 mg/kg/d) for 9 months resulted in no improvement.

Physical examination revealed a blood pressure of 130/80 mmHg, heart rate of 100 beats/min, temperature of 37°C, and respiratory rate of 28 breaths/min. Breath sounds were normal on quiet breathing, but there were inspiratory and expiratory wheezes and coarse rales over the right upper lung on forced inspiration and expiration. Findings from the rest of the physical examination were within normal limits.

Serum electrolytes, renal function, and urinalysis results were normal. Chest X-ray showed right hilar and paratracheal lymph node enlargement with right upper lobe atelectasis (figure 1). His peak flow was 100 l/min and did not improve following the inhalation of nebulized salbutamol. His forced expiratory volume in 1 s (FEV_1) was 1.8 l at best, vital capacity was 4.2 l and flow volume loop showed flat inspiratory and expiratory phases indicative of large intrathoracic airway obstruction. The patient was anergic to skin test with purified protein derivative. Bronchoscopy revealed a regular bluish nodular lesion with slightly increased vascularity and nearly complete obliteration of the orifice of the anterior segmental bronchus of the right upper lobe. Multiple biopsies taken from this endobronchial mass lesion were reported as non-caseating infiltration of the mucosa by epithelioid cell granulomas.

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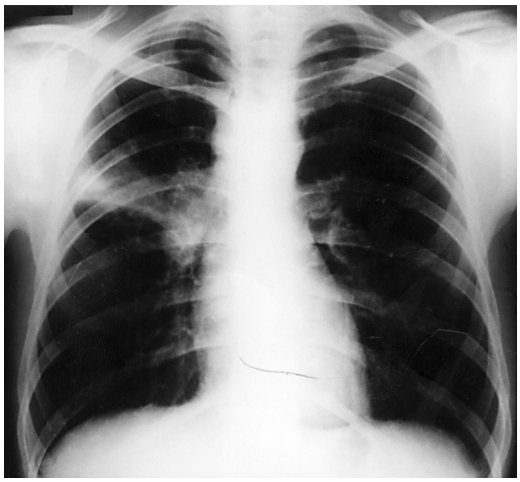

Figure 1 Chest X-ray showing right hilar and paratracheal lymph node enlargement with right upper lobe atelectasis

Questions

1 What is your diagnosis?
2 What other possible diagnoses are compatible with the patient's clinical presentation?
3 How would you manage this patient?

Answers

QUESTION 1
The diagnosis is sarcoidosis.

QUESTION 2
Other possible diagnoses include neoplasm (bronchogenic carcinoma or lymphoma), tuberculosis, fungal disorders (aspergillosis, nocardiosis), other granulomatous disease (syphilis), HIV, and bronchial asthma.

QUESTION 3
The patient should receive corticosteroid therapy.

Outcome

Prednisolone 40 mg daily was started and gradually tapered to 10 mg on alternate days. The patient's clinical condition improved within 2 months, and 3 months after the initiation of therapy, a control chest X-ray showed complete resolution of the previous right-upper lobe atelectasis, and also right hilar lymph node enlargement (figure 2). Control bronchoscopy revealed partial resolution of the previous nodular endobronchial lesion.

Discussion

Sarcoidosis is a multisystem granulomatous disorder of unknown aetiology, characterised histologically by epithelioid tubercles involving various organs or tissue, with symptoms dependent on the site and degree of involvement. It occurs predominantly between ages 20 and 40. The most common sites are the lungs, lymph nodes, liver, eyes, and skin.[1] The lungs are the most frequently involved organ, and pulmonary symptoms include dyspnoea on exertion, nonproductive cough, and wheezing. Because the lung is involved so commonly, the routine chest film is usually abnormal (box 1).[1]

Classic X-ray patterns of pulmonary sarcoidosis

Grade I: lymph node enlargement without pulmonary parenchymal abnormalities
Grade IIA: combination of lymph node and diffuse pulmonary parenchymal disease
Grade IIB: diffuse parenchymal disease without lymph enlargement
Grade III: pulmonary parenchymal fibrosis

Box 1

Much attention has been focused on the restrictive ventilatory defect seen in pulmonary sarcoidosis. However, a number of reports have indicated that sarcoidosis may cause varying degrees of upper and lower airway obstruction.[2 3] Initial reports suggested that obstructive ventilatory defects occur only in the late stages of the disease, associated with pulmonary fibrosis. However, occlusion of a lobar bronchus and lobar atelectasis may occur without pulmonary fibrosis due to extrinsic compression by enlarged lymph nodes or, rarely, endobronchial disease.[4] Moreover, isolated endobronchial lesions, which are extremely rare in pulmonary sarcoidosis,[5] may also cause obstructive ventilatory defects, as in our patient.

Because sarcoidosis can occur in almost any part of the body, like tuberculosis or syphilis, it may be confused with other disorders. However, it is most commonly confused with neoplastic disease such as bronchogenic cancer and lymphoma or with disorders also characterised by a mononuclear cell granulomatous inflammatory process, such as the myobacterial and fungal disorders.[1 6] Whether or not the presentation is 'classic', biopsy evidence of a mononuclear cell granulomatous inflammatory process is mandatory in order to make a definitive diagnosis.[6] However, the histologic findings are not sufficiently specific to make the diagnosis by themselves, since noncaseating granulomas are found in a number of other diseases, including infections and malignancy. Thus, a definitive diagnosis of sarcoidosis is based on biopsy in the context of the history, physical examination, blood tests, X-ray, lung function, and, if available, gallium 67 scan and bronchoalveolar lavage (figure 3).[6]

The therapy of choice for sarcoidosis is glucocorticoids.[6] The major problem in treat-

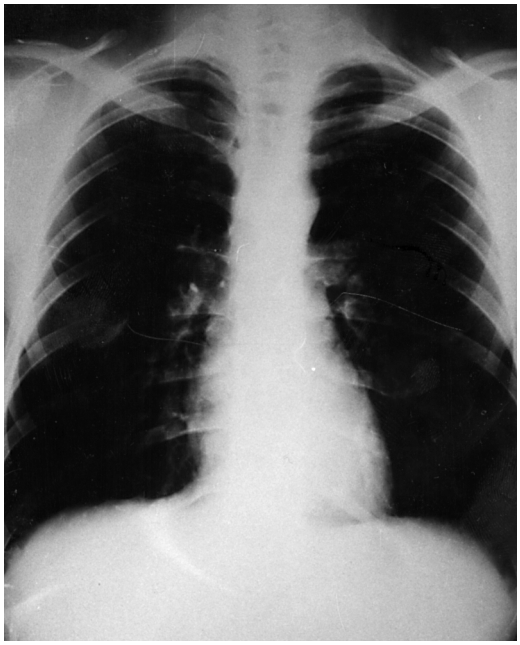

Figure 2 Control chest X-ray showing complete resolution of the previous right-upper lobe atelectasis, and also right hilar lymph node enlargement

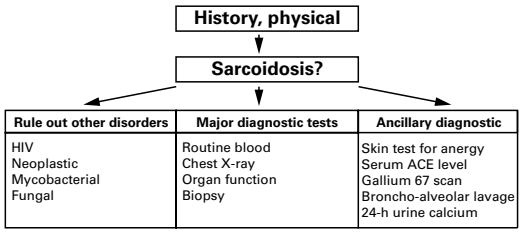

Figure 3 Diagnosis of sarcoidosis (adapted from[6])

ing sarcoidosis is in deciding when to treat. Since the disease may remit spontaneously and since steroids may cause significant side-effects, treatment is usually started only if there is an indication of interference with the function of a vital organ (lungs, kidneys, eyes, heart, or central nervous system). Because of severe dyspnoea and cough, the present case was treated with high-dose oral corticosteroids. He improved dramatically within 3 months and has been free of complications for 2 years.

Final diagnosis

Bronchial obstruction due to endobronchial sarcoidosis.

Learning points

- although obstructive ventilatory defects rarely occur in the early stage of pulmonary sarcoidosis, endobronchial sarcoidosis must be considered in a young patient presenting with clinical features of bronchial obstruction, when no evidence of other common cause of airway obstruction can be found
- corticosteroids are still an effective and safe treatment for this disorder

Box 2

Keywords: sarcoidosis; corticosteroids; pulmonary obstruction

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Masked hypercalcaemia

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A 29-year-old man presented with a 6-month history of steatorrhoea and a weight loss of 5 kg over 2 months. Endoscopy showed ulcers in the descending duodenum. Fasting serum gastrin was 427 ng/l (normal range less than 100 ng/l). Basal gastric acid secretion was 42.8 mmol/h (normal less than 15 mmol/h). No abnormality was found on abdominal computed tomography (CT) scan. The patient also complained of proximal muscle weakness and rib pain. On examination he had Harrison's sulci and lower rib tenderness. Biochemical measurements are shown in the table.

Table Serum biochemistry

	At presentation	*Post-vitamin D supplementation*	*Post- para- thyroidectomy*	*Reference range*
Ionised Ca (mmol/l)	1.24	1.49	1.25	1.19–1.35
PTH (pmol/l)	87.3	42.1	7.64	0.2–5.5
Phosphate (mmol/l)	0.5	0.79	1.14	0.8–1.4
25(OH)D (nmol/l)	<5.0	53.2	17	<50
Osteocalcin (ng/ml)	—	152	3	8.8–14.8
Ur DPD (nmol/mmol Cr)	—	39.5	5	3.1–5.6

Ur DPD = urinary free deoxypyridinoline

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Questions

1 What was the most likely diagnosis in this patient ?
2 Why did the patient have steatorrhoea ?
3 What other conditions did this patient have and can you explain the reasons for this?
4 What syndrome did the patient have ?