Pseudodextrocardia in bronchiectasis

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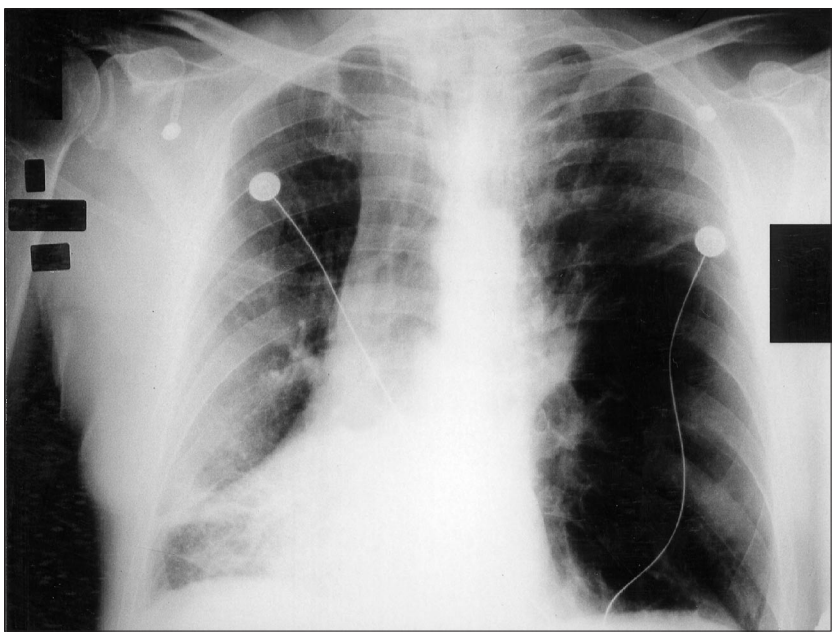
INTRODUCTION

Bronchiectasis is a chronic inflammatory pulmonary disorder characterized by chronic productive cough, recurrent bronchial sepsis and haemoptysis, with airflow obstruction and progressive tissue destruction. Kartagener's syndrome, associated with dysfunctional cilia, sinusitis and situs inversus, is a rare cause of bronchiectasis (Marwah and Sharman, 1995). This article reports two cases where patients had been erroneously diagnosed as having Kartagener's syndrome based on misinterpretations of chest radiographs.

Figure 1. Posteroanterior chest radiograph on admission showing right basal acute airspace shadowing, left basal hyperlucency consistent with bulla formation and apparent dextrocardia.



CASE REPORT 1

A 61-year-old man, known to have bronchiectasis, was admitted with breathlessness, wheeze, cough and copious yellow sputum. Examination revealed poor air entry and bibasal, coarse, pan-inspiratory crackles. Investigations showed an elevated white cell count (11.7 x 10^9 cells/ml), C-reactive protein level (100 mg/litre) and type 1 respiratory failure (partial pressure of oxygen 6.9 kPa). He had stopped smoking 6 years previously. The chest radiograph is seen in *Figure 1*. The electrocardiograph was normal. Forced expiratory volume in 1 second (FEV_1) was 0.72 litres (24%), and ratio of FEV_1 to forced vital capacity was 45%. Sputum culture isolated coliforms. He responded well to intravenous cephalosporin, supplemental oxygen and oral prednisolone.

A chart review indicated previous admissions with exacerbations of bronchiectasis. He had been diagnosed as having Kartagener's syndrome on the basis of previous chest radiographs, interpreted as showing dextrocardia.

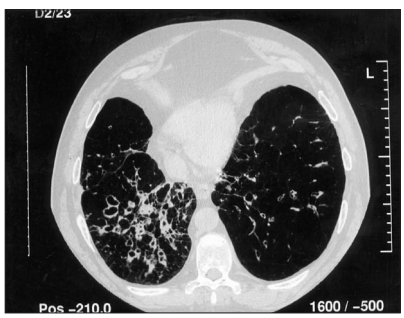
Bronchiectasis was diagnosed by bronchography in 1974. Alpha-1-antitrypsin status was normal. He had been treated for pulmonary *Mycobacterium kansasii* infection in 1992.

To characterize his disease more thoroughly, a high-resolution computed tomograph of the chest was performed (*Figure 2*). This demonstrated bronchiectasis and displacement of the heart towards the right hemithorax, with absence of malrotation along the sagittal axis. Subsequent review of earlier radiographs confirmed laevocardia.

DISCUSSION

Kartagener's syndrome is a rare autosomal recessive disorder, with an incidence of 1 in 15 000 to 1 in 30 000 (Marwah and Sharman, 1995), characterized by sinusitis, situs inversus and bronchiectasis (Woodring et al, 1982). Diagnosis is based on characteristic clinical features but may also be demonstrated on mucosal biopsy showing ciliary dykinesia and absence of dynein arm (a structural protein within cilia) (Woodring et al, 1982). Bronchiectasis is best diagnosed based on findings of a high-resolution computed tomograph (HRCT) of the chest (Marwah and Sharman, 1995;

Figure 2. High-resolution computed tomograph of the chest at the level of the cardiac apex showing bibasal bronchiectatic change (right>left), airspace enlargement at the left base and subsequent displacement of the heart to the right.



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CASE REPORT 2

A 29-year-old woman, diagnosed as asthmatic by her GP, was referred to the chest clinic with recurrent respiratory tract infections, night sweats, purulent sputum and nocturnal wheeze. She was admitted for further management. A chest radiograph showed displacement of the cardiac shadow to the right, with lower zone airspace shadowing. Her C-reactive protein level was 108 mg/litre, white cell count was 18.05×10^9 cells/ml and sputum cultured *Haemophilus influenzae*. She received treatment with intravenous cefotaxime with good improvement; her C-reactive protein level decreased to 20 mg/litre and white cell count to 5.61×10^9cells/ml.

Sweat test, aspergillus precipitins, immunoglobulin E, α_1-antitrypsin, immunoglobulins and autoantibody screen were normal. Forced expiratory volume in 1 second (FEV_1) was 1.53 litres (47%), and ratio of FEV_1 to forced vital capacity was 94%. The electrocardiograph was normal.

She had had a repair of oesophageal atresia and tracheo-oesophageal fistula as a baby and had frequent chest infections as a child. She had previously been admitted to another hospital with pneumococcal pneumonia. A diagnosis of dextrocardia and Kartagener's syndrome was made, although there was no evidence of sinus disease.

To characterize her disease more accurately, a high-resolution computed tomograph of the chest was performed. This revealed volume loss in the right hemithorax. The heart was appropriately orientated, although displaced to the right, confirming laevocardia. Bronchial dilatation and crowding were seen in the lingula, middle lobe and right lower lobe, consistent with bronchiectasis. The fact that the disease was much worse on the right suggests that aspiration from the fistula in childhood might have been the aetiology.

McGuinness and Naidich, 1995; Hansell, 1998), although a chest radiograph may suggest the diagnosis if the clinical history is typical.

The finding of Kartagener's syndrome as a cause of bronchiectasis is unusual (Marwah and Sharman, 1995). In an audit of bronchiectasis patients attending the authors' chest clinic, none of a cohort of 100 patients were found to have Kartagener's syndrome (Kelly and Elborn, 1999).

Tissue destruction may result in 'honeycombing' (Marwah and Sharman, 1995), alveolar destruction and overinflation of lobes (Barker, 1995). These contribute to airspace enlargement, producing radiographic features identical to emphysema. Areas of decreased attenuation on HRCT, representative of small airways disease with air trapping, may be confused with emphysema (Hansell, 1998).

These cases demonstrate a potential pitfall. An erroneous diagnosis of Kartagener's syndrome was made in case one on the basis of chest radiograph appearances and known bronchiectasis. The combination of traction from the right basal bronchiectasis and overinflation from the left-sided airspace enlargement has led to the displacement of the cardiac shadow to the right, mimicking dextrocardia. In case two, the erroneous diagnosis was again based on chest radiograph appearances which were likely to be caused by traction from the right base. Computed tomography scanning, review of previous chest radiographs and absence of dextrocardia on electrocardiography would have enabled an accurate diagnosis in both cases. **HM**

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