



What is the rare but well-documented cause of this recurrent chest infection?

Case history

A 61-year-old female presented with a history of recurrent chest infections over several years. Episodes were characterised by cough and green sputum without haemoptysis. There was no history of chest pain, shortness of breath, fever, night sweats or weight loss. She had received several courses of antibiotics for this from her general practitioner. Her past medical history included whooping cough as a child and hysterectomy at age 40 years, for reasons unknown. She had smoked 10 cigarettes per week since her teenage years and denied any alcohol intake. There were no pets at home or any history of recent travel.

Examination

The patient was comfortable at rest, with no cyanosis, clubbing, leg oedema or lymphadenopathy. Pulse rate was 88 beats per min and regular, blood pressure 155/85 mmHg and her jugular venous pressure was normal. Chest examination was normal except for an occasional wheeze. Cardiovascular examination revealed a soft systolic murmur at the apex with normal heart sounds. The rest of her examination was normal.

Investigations

Initial blood results showed: white blood cell count 4.1×10^9 per L, Hb 13 g per dL, platelets 319×10^9 per L, glucose 5 mmol per L, thyroid-stimulating hormone 1.76 U per mL and normal liver and renal functions. Immunoglobulin (Ig) levels were as follows: IgG 11.6 (7–14) U per mL, IgA 3.13 (0.75–4) U per mL and IgM 0.4 (0.3–1.7) U per mL.

Sputum culture showed no growth. Electrocardiogram (ECG) showed sinus rhythm.

Spirometry results were as follows: forced expiratory volume in one second (FEV1) 1.26 L (60% predicted); forced vital capacity 1.75 L (65% pred); and FEV1/FVC 72%.

Figure 1 shows chest radiography.

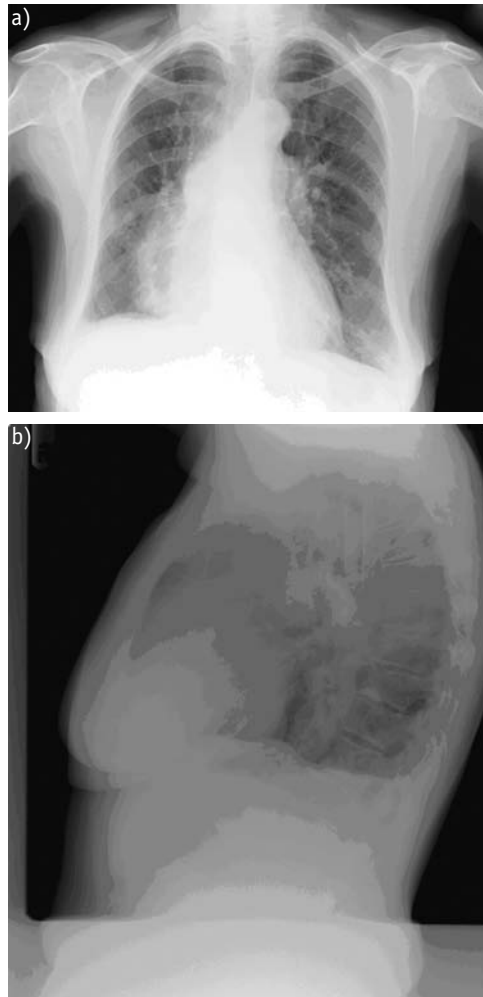


Figure 1
Chest radiography.

Task 1

Interpret the chest radiographs.

Task 2

What is your clinical diagnosis?

a. Pneumonia in view of recurrent infection.

b. Tumour in view of smoking history.

c. Developmental anomalies in view of peculiar chest radiography findings.

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Answer 1

Prominent right hilum. Ill-defined curvilinear shadow in the right paracardiac region.

Answer 2

c.

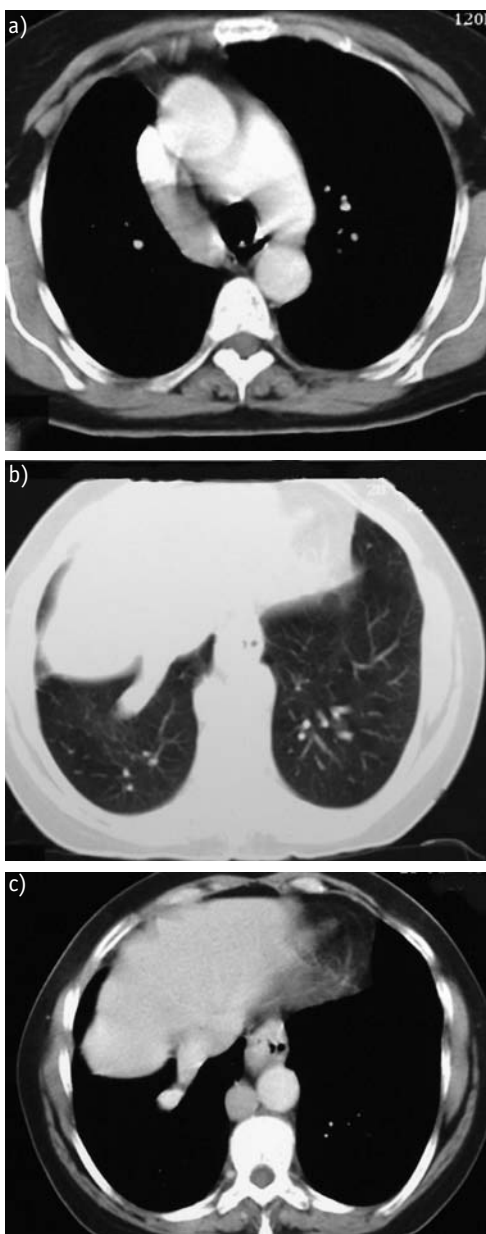
Bronchoscopy showed no upper lobe bronchus. There were three openings from the bronchus intermedius and no apical segment division. No endobronchial lesion was seen.

Figure 2 shows the results of computed tomography (CT) scanning.

Answer 3

The right lung is smaller with a slightly displaced mediastinum to the right. A large pulmonary vein can be seen in the right lung, draining into the venous system below the diaphragm forming the inferior vena cava (IVC), which is consistent with **Scimitar syndrome**. In addition, the right upper lobe bronchus is absent and there is trifurcation of the right bronchus intermedius (not visualised in these images).

Figure 2
CT scans.

**Task 3**

Interpret the CT scans.

A transoesophageal echocardiogram revealed morphologically normal valves with mild mitral regurgitation and trivial tricuspid regurgitation. The interatrial septum was intact. The right heart was not dilated and there was no evidence of intracardiac shunt.

Discussion

Scimitar syndrome is a rare but well-documented congenital anomaly, first described in 1836 by CHASSINAT [1] and COOPER independently [2]. The term "scimitar" was not used until 1956, when HASALAZ *et al.* [3] described the appearance of an anomalous vein whose curvilinear opacity on radiography resembled a Turkish sword. However, it was NEILL *et al.* [4], in 1960, who coined the term Scimitar syndrome [5].

Scimitar syndrome is also known as congenital venolobar syndrome and it consists of partial anomalous pulmonary venous drainage from the right lung into the IVC, partial agenesis or hypoplasia of the right lung, dextrocardia and anomalous systemic arterial supply especially to the right lower lobe. Other features include hypoplastic or absent pulmonary artery, broncho-pulmonary sequestration and cardiac anomalies (atrial or ventricular septal defect, coarctation of aorta, pulmonary stenosis, tetralogy of fallot) [6, 7].

This anomaly occurs as the result of abnormal development of the right lung bud early in embryogenesis. It tends to most frequently involve the right lung, for unknown reasons, with rare reports of left lung involvement. It occurs

more commonly in females, with occasional familial occurrence. The clinical presentation is quite variable, being asymptomatic to florid heart failure. DUPUIS *et al.* [8], in their review of 122 adult patients, showed recurrent pneumonia as the most common presentation, occurring in 38 patients, exertional breathlessness in 23 patients, deformity of thorax in eight and haemoptysis in seven.

In the adult form, which is defined as those patients in whom the syndrome is detected after the first year of life, clinical examination may be entirely normal. ECG may show right ventricular strain. Echocardiography, apart from identifying associated anomalies, may also be helpful in establishing the diagnosis. However, this can miss about 33% of patients [9]. CT, cardiac catheterisation, and magnetic resonance imaging with three-dimensional magnetic resonance angiography are more rewarding [10, 11].

One-quarter of cases are symptomatic in the newborn, presenting as respiratory or cardiac failure needing corrective surgery. Although repair of the anomalous venous return or ligation of collaterals is the procedure of choice, pneumonectomy has also shown similar early and late results. Surgery is indicated when the right-to-left shunt is >2:1 [12, 13]. Sometimes patients present with an incidental radiological finding with no symptoms. They require no intervention.

The current patient presented with recurrent chest infections and since there was no significant shunt, she was managed conservatively with antibiotics.

References

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