

CASE FOR DIAGNOSIS

Chest pain in a patient with cystic fibrosis

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Case history

A 38 yr old female with a history of cystic fibrosis (CF) presented to an outside emergency department with dyspnoea and right-sided chest pain of approximately 12 h duration. Her history was significant for recurrent pneumothoraces and a recent respiratory exacerbation of CF requiring prolonged antibiotic therapy for *Pseudomonas aeruginosa*. She described the pain as relatively acute in onset, sharp, increased with deep inspiration, without outward radiation, and progressive in intensity. There was no history of travel, worsening cough, fever, chills or increase in her chronic expectoration of blood-streaked sputum. An outside chest radiograph was interpreted as demonstrating a "rounded" right lower lobe pneumonia and she was transferred to our institution for further evaluation.

On examination, she was thin, afebrile and in minimal respiratory distress. There were decreased breath sounds throughout all lung fields, symmetric chest wall excursion with inspiration and bilateral basilar crackles. Subjective right lateral chest discomfort was reported with deep inspiratory manoeuvres. The remaining physical examination was normal except for clubbing of the upper extremities.

Laboratory data revealed an elevated white blood cell count of 19.2×10^9 cells·L⁻¹ with a normal differential. Haemoglobin and haematocrit were normal, as were prothro-

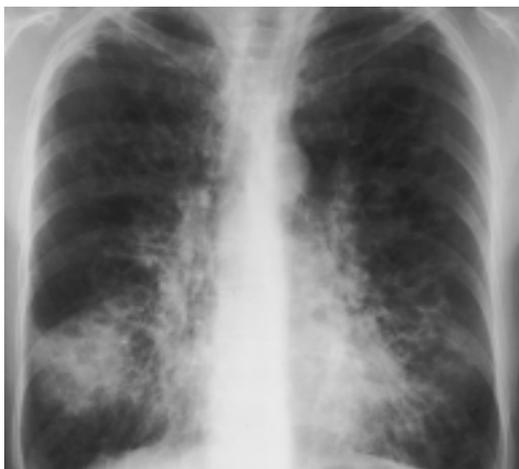


Fig. 1. - Admission posteroanterior chest radiograph.

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Fig. 2. - Computed tomography scan of the chest. Open arrow: multiple cysts; closed white arrow: 3.8x5 cm round mass,

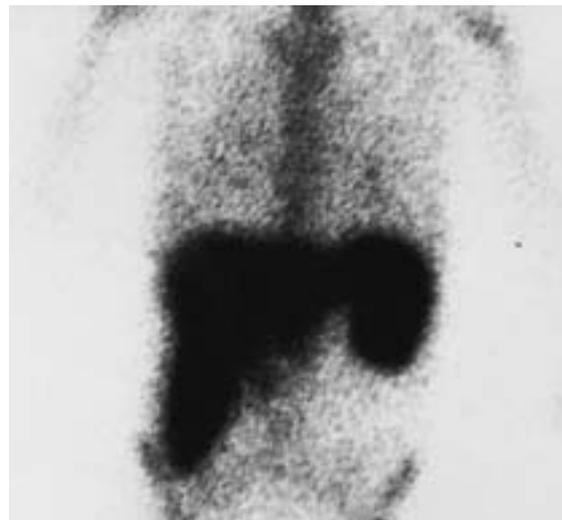


Fig. 3. - Indium-111-labelled leukocyte scan.

mbin time and partial thromboplastin time. The admission chest radiograph and corresponding chest computed tomography (CT) scan are shown in figures 1 and 2, respectively.

The patient was begun on broad-spectrum antibiotics but continued to have severe right-sided chest pain. The possibility of intraparenchymal haemorrhage was raised and further history revealed an increased volume of haemoptysis the day before admission. An indium-111 (¹¹¹In) tagged white blood cell scan was obtained to help to differentiate localized abscess from haemorrhage and is shown in figure 3.

BEFORE TURNING THE PAGE, INTERPRET THE RADIOGRAPH, CT SCAN AND INDIUM-111 SCAN, AND SUGGEST DIAGNOSIS AND TREATMENT

Interpretation

On the admission chest radiograph (fig. 1), the patient had large lung volumes with multiple parenchymal cysts predominantly central in distribution, consistent with central bronchiectasis. A focal rounded opacity was present in the right lower lobe.

The CT scan of the thorax (fig. 2) demonstrated extensive bronchiectasis manifested by multiple cysts (open arrow) which communicated with the central airways on serial images. The 3.8×5cm round mass in the right lower lobe (closed white arrow) had an air/fluid level which was interpreted as a pulmonary abscess.

The nuclear medicine study (fig. 3) demonstrated diffuse increased activity throughout the lungs which was consistent with the chronic inflammation of bronchiectasis. There was no evidence of focal increased uptake in the right lower lobe to indicate an infection.

Based on the findings on the ¹¹¹In-scan and the patient's afebrile state, the mass with the air/fluid level was re-interpreted as a haemorrhage into a large parenchymal cyst.

Diagnosis: "Haemorrhage into a large parenchymal cyst"

Treatment and clinical course

The patient was observed for 3 days and continued to receive intravenous antibiotics. Subsequently, she deve-

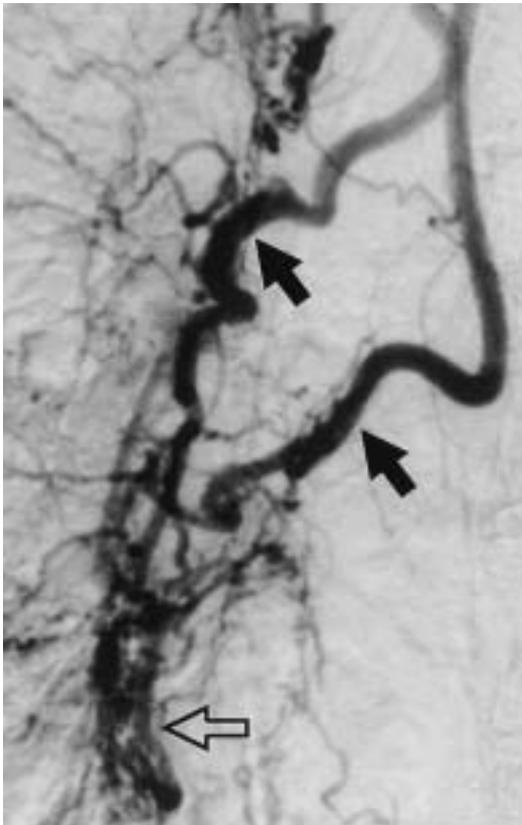


Fig. 4. – Right bronchial artery angiogram. Closed arrows: dilated right bronchial artery branches; open arrow: area of abnormality in the right lower lobe.

loped episodic haemoptysis of approximately 250–300 mL blood. An emergent arteriogram of the right bronchial circulation was performed. The arteriogram (fig. 4) demonstrated dilated right bronchial artery branches (closed arrows) supplying the area of abnormality in the right lower lobe (open arrow). The bronchial arteries to the right lower lobe were embolized with polyvinyl alcohol particles and repeat arteriogram confirmed occlusion of the embolized vessels. The patient reported an improvement in chest discomfort within 24 h and had no further haemoptysis at a two month follow-up evaluation.

Discussion

Minor haemoptysis is a common pulmonary complication in patients with CF and generally requires no specific intervention except for observation, bed rest, discontinuation of chest physiotherapy, correction of abnormal coagulation and initiation of antibiotics. Major haemoptysis (as present in this patient) is defined as >240 mL bleeding in 24 h. This only occurs in about 1% of CF patients per year and is usually seen in patients older than 16 yrs [1]. Significant bleeding almost always arises from the systemic circulation and results from bronchial artery dilation (and abnormality) within areas of chronic infection, bronchial wall destruction, and bronchiectasis. Chest radiograph findings in patients with pulmonary haemorrhage are generally nonspecific, although with massive haemorrhage, there can sometimes be an increasing focal infiltrate on the chest radiograph or CT scan [2]. When conservative measures fail to arrest massive haemoptysis, bronchial artery angiography and subsequent embolization are used to identify and interrupt bleeding vessels [3, 4].

Large intraparenchymal abscess formation is exceedingly uncommon in CF patients [5]. However, small areas of focal pneumonia can occur around small bronchi and bronchioles resulting in the formation of inflammatory nodules with pathological findings similar to micro-abscesses [6]. In patients with suspected lung abscess, ¹¹¹In-leukocyte scanning is sensitive in identifying such abnormalities [7, 8]. Using this technique, patients with pulmonary abscesses exhibit dense recruitment of labelled leukocytes within 4 h of injection. Similar findings are not present in patients with pneumonia. Despite a high sensitivity for localized pulmonary abscesses, there have been cases reported of false-positive scans in patients with CF [9, 10]. In such patients, diffuse pulmonary uptake is present during scanning and is hypothesized to occur from uptake in areas of bronchiectasis, as seen in the patient presented here.

Patients with cystic fibrosis may present with atypical symptoms and chest radiographs that are difficult to interpret. The present patient's initial complaint was of significant chest pain and her chest radiograph findings were consistent with pneumonia. It was not until repeat questioning that the history of significant haemoptysis was obtained. In patients with cystic fibrosis who frequently have complicated chest radiographs, computed tomography can assist in characterizing focal abnormalities. Although not present in this patient and extremely uncommon, intrapulmonary abscesses can occur in patients with cystic fibrosis. In those patients with suspected pulmonary abscesses, indium-111 scanning can assist in the diagnosis. The pre-

sent patient had haemorrhage into a parenchymal cyst and only later developed massive haemoptysis unresponsive to conservative management. Bronchial artery angiography demonstrated abnormal bronchial vessels and embolization resulted in an improvement in both bleeding and subjective symptoms.

Keywords: Bronchiectasis, cystic fibrosis, haemoptysis, indium-111 scan, pulmonary angiogram, pulmonary haemorrhage.

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