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A rare cause of acute chest pain in a young adult

R Selvaratnam, U Srirangalingam, E Mclean, L Lang-Lazdunski and P Goulden

Case presentation

A 37-year-old man presented to the emergency department with a sudden onset left-sided chest pain radiating to the left arm and the epigastrium. The pain was pleuritic in nature and not exertionally related. Associated symptoms included nausea and sweating. The patient was a 15-pack/year smoker but had no other significant past medical history. On examination he was clammy, tachypnoeic, apyrexial and had an oxygen saturation of 97% on room air. Right and left arm blood pressure were 149/78 mmHg and 147/80 mmHg respectively and peripheral pulses were symmetrical and equal. Chest auscultation revealed crepitations at the right base and reduced breath sounds on the left. The remainder of the examination was entirely normal. Initial investigations included an electrocardiogram (ECG) which revealed a sinus tachycardia without other changes and a chest radiograph (Fig 1a) showed left basal shadowing and associated upper lobe diversion. Laboratory testing demonstrated a haemoglobin of 17 g/dl, a leucocytosis of 14.4 x 109/l (normal range 4-11$\times 10^9$/l), a neutrophilia of 7.13 $\times 10^9$/l (3-5$\times 10^9$/l), an elevated urea of 12.6 mmol/l, a C-reactive protein of 213 mg/l, a plasma glucose of 7.7 mmol/l and a troponin measurement in the normal range.

Differential diagnosis

The patient presented with sudden onset left-sided pleuritic chest pain and associated chest signs. He was tachycardic but normoxaemic and apyrexial. The initial differential diagnosis of the admitting team included a left basal pneumonia with associated pleural effusion, pulmonary embolus, pericarditis, myocardial infarction (MI) or an aortic dissection. While pleuritic chest pain could be compatible with a diagnosis of infection, thromboembolism or pericarditis, the clinical signs, radiology and biochemistry favoured the former diagnosis. Against this was the sudden onset of symptoms and the lack of a prodrome or fever. The patient was normoxaemic, had no evidence of deep vein thrombosis and the only risk factor for thromboembolic disease was a history of smoking. The nature of the pain did not suggest MI or aortic dissection and the ECG did not suggest myocardial ischaemia, infarction or pericarditis. The mediastinal width was difficult to assess given the quality of the initial film (Fig 1a) but there were no features on clinical examination to suggest an aortic dissection.

Initial management

Given the presumptive diagnosis, initial management consisted of treatment with antibiotics for a community-acquired pneumonia, oxygen therapy and analgesia. The CURB65 score of 2 (moderate severity pneumonia) was interpreted in the context of a young patient with marked radiographic abnormalities managing to maintain his vital signs despite significant compromise. Treatment was instigated with a view to careful assessment over the initial 24 hours. Appropriate microbiological samples were sent to identify atypical organisms. A treatment dose of a low molecular weight heparin (LMWH) was administered to cover for the differential diagnoses of an acute coronary syndrome and pulmonary embolus.

Case progression

The following day the patient's clinical condition deteriorated with a worsening tachycardia, tachypnoea and oxygen saturations of 86% on room air. On chest auscultation there was a further reduction of breath sounds on the left side and dullness to percussion. Arterial blood gas analysis on 15 litres of oxygen showed a pH of 7.29 (7.35–7.45), pO_2 13.6 kPa (10.7–13.3), pCO_2 7.03 kPa (4.67–6.00), HCO_3 24.9 mmol/l (22–28) consistent with a type II respiratory failure. Repeat chest X-ray demonstrated a white-out of the left lung field (Fig 1b). An urgent computed tomography (CT) thorax was requested (Fig 1c) and revealed an extensive left-sided posterior basal mass, an associated haemothorax and mediastinal shift to the right. Intubation, ventilation and ionotropes were required to correct ensuing hypoxia and hypotension caused by a loss of circulating

Key learning points

- Congenital cystic adenoid malformation/pulmonary sequestration are rare congenital lung anomalies.
- Congenital cystic adenoid malformation/pulmonary sequestration usually present in early childhood but can occasionally present in adulthood.
- Clinically these lesions often mimic other common respiratory conditions but may present acutely and without prior history.
- The administration of low molecular weight heparin should be carefully considered in atypical presentations of chest pain.

R Selvaratnam,[1] acute medicine specialist registrar; **U Srirangalingam,**[2] endocrine specialist registrar; **E Mclean,**[3] consultant histopathologist; **L Lang-Lazdunski,**[3] consultant thoracic surgeon; **P Goulden,**[1] consultant physician

[1]Maidstone General Hospital, Maidstone, Kent; [2]St Bartholomew's Hospital, London; [3]Guy's Hospital, London

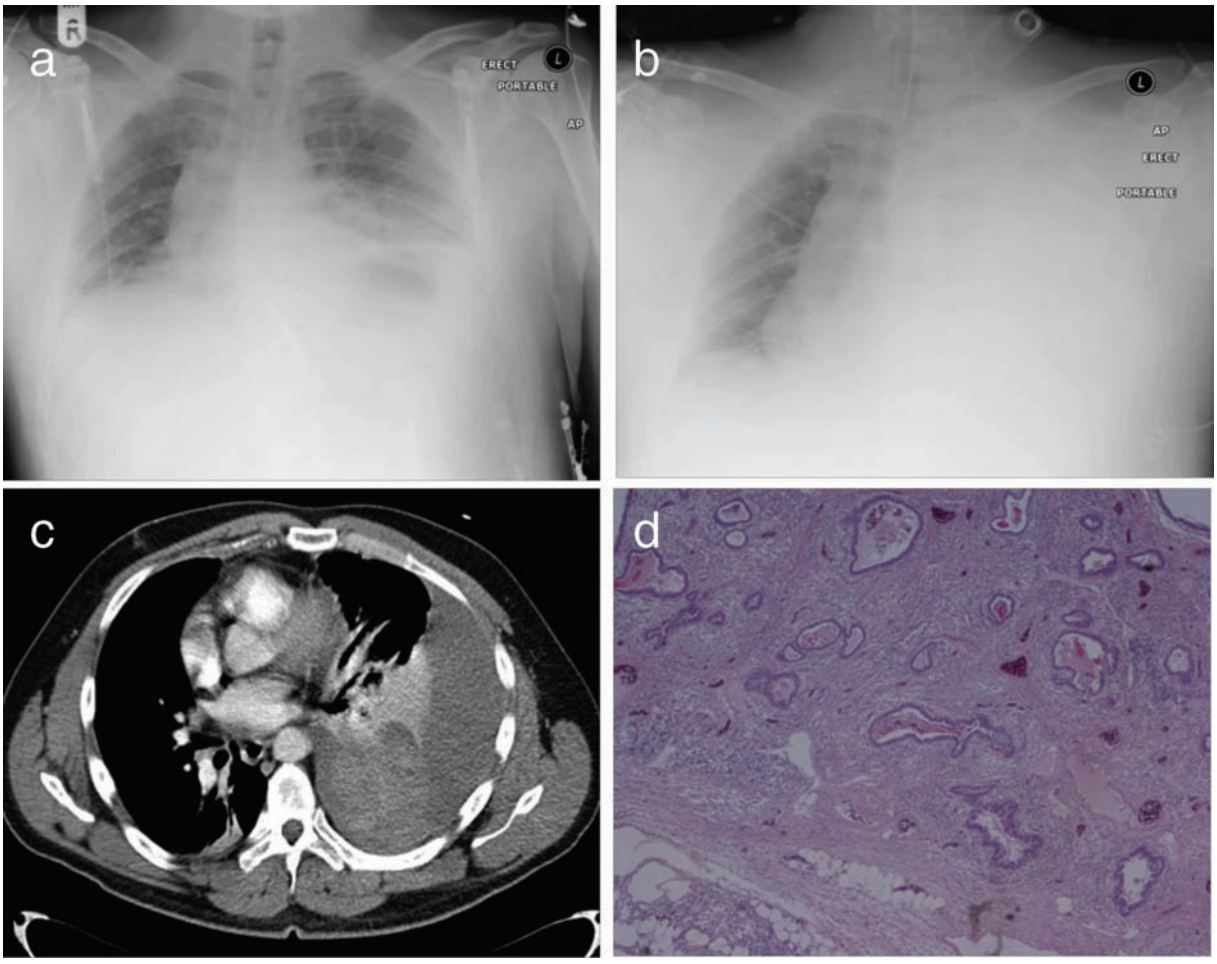

Fig 1. (a) Chest radiograph on presentation; (b) chest radiograph following clinical deterioration; (c) computed tomography thorax demonstrating left basal cystic mass and associated haemothorax. The normal lung appears anteriorly and there is mediastinal shift to the right; (d) type II congenital cystic adenoid malformation demonstrating multiple small cysts lined by ciliated, cuboidal or columnar cells with an absence of mucinous and cartilaginous elements.

volume and cardiac tamponade. His haemoglobin dropped to 10 g/dl and a pleural drain insert under ultrasound guidance confirmed the presence of a haemothorax. He received a blood transfusion and was transferred to a tertiary cardiothoracic centre for definitive management. Prior to surgery the patient's haemoglobin had dropped further to 6.9 g/dl. An emergency thoracotomy was carried out in which a left-sided cystic mass attached to the diaphragm, was excised, and a large feeding pedicle arising directly from the throraco-abdominal aorta was ligated. Approximately three litres of blood and clot was evacuated from the chest cavity. Following surgery, the patient made a full recovery. Histological examination of the excised tissue revealed an extra-lobular pulmonary sequestration (PS) with features of type II congenital cystic adenoid malformation (CCAM) (Fig 1d).

Discussion

Pulmonary sequestrations and congenital cystic adenoid malformation are congenital cystic lung lesions which may occasionally present in adulthood. Clinical manifestations of disease are often varied and may mimic other common conditions. This article reports the case of an adult patient initially presenting on the general medical take with a history of chest pain who rapidly progressed to respiratory failure and circulatory collapse as a result of congenital cystic lung disease without a prior history.

A pulmonary sequestration is a segment of non-functioning lung tissue which derives its vascular supply directly from the systemic circulation and lacks communication with the remaining lung.[1] These lesions are intra-lobar or extra-lobar based on the absence or presence of a separate pleural layer surrounding the sequestration. Congenital cystic adenoid malformation is a developmental focal dysplastic lesion thought to arise from the arrest of lung development between the fourth and seventh weeks of gestation.[2]

CCAM and PS make up 25% and 6% of all congenital lung lesions respectively and are thought to share a common embryological origin. They appear to co-exist in 30% of cases.[3] The occurrence of CCAM in the fetus during pregnancy has been

estimated at one in 25,000/35,000. Seventy-six per cent of lesions are thought to spontaneously regress and up to 85% of CCAM lesions present by the age of two. Only a small percentage of subjects present in adulthood.

This case highlights an adult presentation of CCAM and PS.[4] Such lesions may be asymptomatic and identified incidentally or present with a more insidious history of recurrent chest infections. Presentations include haemoptysis and shortness of breath secondary to pleural collections (haemo-, pneumo- and pyopneumothorax). This case illustrates the acute and rapidly progressive nature in which such patients may also present.

CCAM and PS, along with other congenital cystic lung lesions, have been associated with an increased risk of malignant transformation into pulmonary blastomas, rhabdomyosarcomas and bronchioalveolar carcinomas. Therefore, once identified, the definitive management is surgical excision.

This patient received a treatment dose of LMWH as part of initial management to cover for differential diagnoses including acute coronary syndrome and pulmonary embolus. It is unclear whether this worsened the developing haemothorax or clinical deterioration. LMWH is an important aspect in the treatment of several conditions but in order to avoid potential iatrogenic complications, more consideration needs to be made as to the likelihood of a diagnosis which would require its use.

In summary, CCAM and PS are rare congenital lung lesions which may infrequently present in adults. They frequently mimic the presentation of other, more common cardiorespiratory conditions. This article illustrates the acute nature in which these cases may present and highlights the potential complication of LMWH administration.

References

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Address for correspondence: Dr R Selvaratnam, Department of Medicine, Hermitage Lane, Maidstone, Kent ME16 9QQ.
Email: rselvaratnam@doctors.org.uk