A diagnosis of seclusion

Case history

A 40-yrold female was seen in the rapid-access chest clinic with a 4-month history of dry cough, mild dyspnoea and left chest discomfort, which seemed to have started during a holiday to a sunny European country. Antibiotics given by her primary care physician had not helped. The patient was an ex-smoker (10 pack-yrs, having given up 14 yrs previously). There were no significant comorbidities A chest radiograph (CXR) was performed (fig. 1).



Figure 1 Chest radiograph

Task 1

What does the CXR demonstrate? a) Cardiomegaly and Kerley-B

- lines.
- b) A right pleural effusion.
- c) A density overlying the left hilum.
- d) A nodule in the left upper lobe.

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Answer 1

The correct answer is c).

The CXR demonstrated a density overlying the left hilum. The patient underwent a computed tomography (CT) scan, which showed a softtissue mass in the lower left lobe without any effusion, lymphadenopathy or other distant disease (fig. 2). A CT-guided biopsy was undertaken, which did not reveal any evidence of malignancy. Following the biopsy, the patient developed increased left pleuritic chest pain and CXR showed a small left pleural effusion. She then underwent a positron emission tomography (PET)-CT study, which showed that the index lesion had an standardised uptake value of \sim 7 (fig. 2), and there was some increased uptake at the left hilum; the small effusion was also noted.



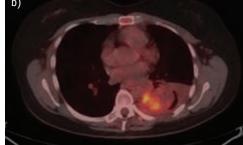


Figure 2

a) Computed tomography (CT) and b) positron emission tomography-CT scans

Task 2

What diagnosis is suggested by the PET-CT scan?

- a) Pulmonary vasculitis.
- b) Bronchial carcinoma.
- c) Pulmonary sequestration.
- d) Mesothelioma.

Answer 2

The correct answer is c).

A provisional diagnosis of nonsmall cell lung cancer was made, although a pulmonary sequestration was considered as a differential diagnosis. The patient was referred to a surgeon with a view to obtaining a diagnostic videoassisted thorascopic surgery (VATS) biopsy, proceeding to resection if appropriate.

A left VATS was undertaken and a subsequent decision was made to proceed to a left minithoracotomy. The lower left lobe was freed from the chest wall with an extra pleural dissection. Further dissection revealed a plane between a consolidated area and the remainder of the lower lobe and it became apparent that this was an intrapulmonary sequestration. A sequestrectomy was performed, dividing two large supplying vessels from the aorta. The patient has made a complete recovery.

The word "sequestration" implies separateness or seclusion. The term was coined by PRYCE [1] in 1946 to describe a disconnected bronchopulmonary segment or cyst with an anomalous systemic artery supplying the involved lung. It is a congenital malformation, thought to develop from an accessory lung bud from the ventral aspect of the primitive foregut. This lung bud may be formed within normal lung tissue and become encased with the same pleural covering (intrapulmonary sequestration) or may become completely enclosed within its own pleural sac (extrapulmonary sequestration). Nearly two-thirds of pulmonary sequestrations appear in the left lung.

>50% of patients with intralobar sequestration become symptomatic after the age of 20 yrs [2]. The common symptoms include cough, sputum production and recurrent pulmonary infections. Haemoptysis is also a common presenting sign. Chest pain, asthma and pleuritic pain are less common presentations. A small number (15%) of patients may be asymptomatic when the lesion is discovered.

Task 3

Which imaging technique has highest sensitivity and specificity for diagnosing pulmonary sequestration?

- a) High-resolution, noncontrast CT scanning. ¹⁸F-2-fluoro-2-deoxy-D-glucose
- b) PET with CT fusion imaging.
- c) Volumetric contrast CT with three-dimensional (3D) reconstruction.
- d) Diffusion-weighted magnetic resonance imaging scanning.

Answer 3

The correct answer is c).

CT scans are reported to have an accuracy of 90% in the diagnosis of pulmonary sequestration, usually by demonstration of the systemic arterial supply. However, this is based upon optimal CT technique by using state-of-the-art volumetric scanning, with rapid i.v. contrast injection rate and appropriate volume and delay, sometimes with 3D reconstructions. The most common appearance is a solid mass that may be homogeneous or heterogeneous, sometimes with cystic changes. Less frequent findings include a large cavitary lesion with an air-fluid level, a collection of many small cystic lesions containing air or fluid, or a well-defined cystic mass.

PET scanning has a role in the diagnosis of lung cancer [3], but there is a significant falsepositive and false-negative rate, which depends, amongst other things, on the pre-test probability of lung cancer. This case report is a timely reminder of the fallibility of PET scanning, as well as of this uncommon condition.

References

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