Rare cause of haemoptysis: bronchopulmonary sequestration
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SUMMARY
Bronchopulmonary sequestration is a rare congenital pulmonary abnormality of the lower airways, which includes an abnormal and non-functioning lung tissue not communicating with the tracheobronchial tree and having aberrant blood supply from systemic circulation with variable venous drainage. The incidence of sequestration is around 0.15%–6.4% of all congenital lung malformations. Common presenting features are cough and expectoration. Misdiagnosed cases may present with recurrent infections and haemoptysis. CT of the chest with contrast is the imaging modality of choice. This is a case report of a 32-year-old woman who presented with cough and haemoptysis. CT of the chest showed a multiloculated mass-like lesion in the left lower lobe with a feeding artery from coeliac plexus and venous drainage via the normal left pulmonary vein. Based on CT chest findings, diagnosis of intralobar pulmonary sequestration was made. The patient was reviewed by cardiothoracic surgeons and underwent surgical resection of the sequestered lung.

BACKGROUND
Bronchopulmonary sequestration is divided into intralobar and extralobar types.1 The most common type is intralobar sequestration, and it is commonly located in the lower lobes of the normal lung and does not have its pleural covering. Intralobar subtype comprises 75% of the pulmonary sequestration, and the incidence is similar in men and women.2,3

On the contrary, extralobar sequestration is situated outside the normal lung with its pleural covering. It constitutes about 25% of bronchopulmonary sequestrations,3 is generally associated with other congenital anomalies and is often diagnosed at an early age.4

This is a case report of intralobar pulmonary sequestration in a 32-year-old woman.

CASE PRESENTATION
A 32-year-old woman was admitted with a 1-week history of moderate haemoptysis associated with vague chest pain and chest tightness. There were no associated symptoms, including fever, weight loss, loss of appetite and dyspnoea.

She underwent thyroidectomy in the past, but she never had a chest X-ray before this admission.

On examination, air entry was noticed to be decreased at the left base and oxygen saturation was 97% on room air. Initial blood results showed marginally raised white cell count with neutrophilia and mildly raised C reactive protein.

Chest X-ray on admission revealed heterogeneous opacity on the left lower zone (figure 1) Ultrasound of the abdomen and pelvis was normal. Based on the above information, an initial diagnosis of community-acquired pneumonia was made and she was started on empirical antibiotics as per local trust protocol. However, she continued to have intermittent mild haemoptysis.

On CT chest findings, a multiloculated mass-like lesion in the left lower lobe was identified (figure 2). The lesion had a feeding artery from the coeliac plexus and venous drainage via the normal left pulmonary vein. Based on these findings, the diagnosis of intralobar pulmonary sequestration was made. The patient was reviewed by cardiothoracic surgeons and underwent surgical resection of the sequestered lung.
Given the suboptimal response to the treatment and drop in haemoglobin from 129 g/L to 103 g/L, CT of the chest with contrast was done, which showed a multiloculated cystic solid mass with low-density lesions and a feeding vessel from the coeliac artery, suggestive of intralobar bronchopulmonary sequestration (figures 2–6). Ultrasound of the abdomen and pelvis revealed no abnormalities.

The diagnosis of intralobar bronchopulmonary sequestration was made based on history and imaging findings. The patient was managed conservatively with antibiotics and tranexamic acid and was subsequently reviewed by cardiothoracic surgeons who advised outpatient review after completion of antibiotics.

The patient had video-assisted thoracoscopic surgery (VATS) resection of the sequestrated lung with good postoperative recovery.

Follow-up chest X-ray showed low lung volume compatible with left lower lobe resection without the presence of the previous consolidation (figure 7).

Histopathology of the resected segment confirmed the diagnosis of intralobar pulmonary sequestration.

INVESTIGATIONS
CT of the chest with contrast and chest X-ray (preoperative and postoperative).
Ultrasound of the abdomen and pelvis.

DIFFERENTIAL DIAGNOSIS
Congenital diaphragmatic hernia.
Mediastinal tumours such as teratoma and neuroblastoma.
Bronchogenic cyst.

TREATMENT
The patient had left-sided VATS to remove the sequestrated left lower lobe of the lung.
Features consistent with intralobar sequestration were confirmed on postoperative histopathology specimen.
DISCUSSION
Intralobar pulmonary sequestration is a relatively rare congenital anomaly with few reports of initial diagnosis occurring during adulthood. The most common location of intralobar sequestration is in the posterior basal segments of the lung, with nearly two-thirds appearing in the left lung. Associated congenital anomalies are uncommon in intralobar sequestration. Some patients are asymptomatic, whereas others may present with persistent cough, expectoration and haemoptysis.

Diagnosis is usually made with imaging such as CT of the chest and in some patients may need angiography. In intralobar sequestration, the anomalous systemic arterial supply is via the descending thoracic aorta in 21% of cases, from intercostal artery in 3% of cases, and rarely from the subclavian, internal thoracic and pericardiophrenic arteries. In our case, the blood supply was from the coeliac axis which is rare.

CT of the chest effectively diagnosed the condition and also delineated the anatomy in this case, which was useful in surgical resection.

A chest X-ray can provide a diagnostic clue to lung sequestration. In the past, aortography was frequently used for diagnosis. However, the gold standard for identifying the pulmonary sequestration is CT/MR angiography as it confirms the anatomy and also identifies the anomalous systemic arterial supply and the venous drainage.

An asymptomatic patient can be observed, while those with recurrent symptoms require surgical resection of sequestrated segments.

Other treatment modalities include arterial embolisation, especially in a lesion with a single well-characterised systemic arterial supply with high-output cardiac failure.

OUTCOME AND FOLLOW-UP
No immediate postoperative complications were noted. The patient was discharged with advice to follow-up in 3 months with a repeat chest X-ray.

Learning points
- Diagnosing a case of adult congenital malformations can be challenging. Therefore, one should consider a wide differential diagnosis for evaluating a patient with unexplained haemoptysis.
- The feeding vessel from the coeliac axis can only be demonstrated through contrast-enhanced CT, and thus when in doubt consider angiography by contrast-enhanced imaging or in rare cases by invasive angiography.
- Timely diagnosis and treatment will not only minimise the patient’s distress but also decrease the burden on the healthcare system.

REFERENCES
1. Landing BH, Dixon LG. Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi, and lungs). Am Rev Respir Dis 1979;120:151.
2. Van Raemdonck D, De Boeck K, Devlieger H, et al. Pulmonary sequestration: a comparison between pediatric and adult patients. Eur J Cardiothorac Surg 2001;19:388–95.
3. Clements BS. Congenital malformations of the lungs and airways. In: Medicine PR, Taussig LM, Landau LI, eds. Pediatric respiratory. 1106. Mosby, St Louis, 1999.
4. Gezer S, Taştepe I, Sirmali M, et al. Pulmonary sequestration: a single-institutional series composed of 27 cases. J Thorac Cardiovasc Surg 2007;133:955–9.
5. Palla J, Sockrider MM. Congenital lung malformations. Pediatr Ann 2019;48:e169 doi:10.3928/19382359-20190326-02.
6. Ko SF, Ng SH, Lee TY, et al. Noninvasive imaging of bronchopulmonary sequestration. AJR Am J Roentgenol 2000;175:1005.
7. Abel RM, Bush A, Chitty LS. Congenital lung disease. In: Kendig's Disorders of the Respiratory Tract in Children. 8th ed. Philadelphia, Pa: WB Saunders, 2012: 317–57.
8. Curros F, Chigot V, Emond S, et al. Role of embolisation in the treatment of bronchopulmonary sequestration. Pediatr Radiol 2000;30:769–73.
