Intralobar lung sequestration as a cause of unexplained recurrent chest infections

This case report describes intralobar lung sequestration presenting as unexplained recurrent chest infections and bronchiectasis. Surgery is important to consider if there are recurrent infections, systemic symptoms or significant haemoptysis.

Discussion

Lung sequestration is a rare congenital or acquired entity characterized by an area of non-functioning abnormal lung tissue with blood supply from the systemic circulation and no connection with the tracheobronchial tree. In the intralobar form the abnormal lung tissue is contained within the visceral pleura of a pulmonary lobe (in the extralobar form, the sequestration is enveloped in its own visceral pleura). Imaging is key to diagnosis and also for surgical planning.

On the basis of the Pryce classification (Pryce, 1946) (*Table 1*), this patient had a type 3 intralobar sequestration. Intralobar sequestrations are more common on the left and in the lower lobe as in this case. The arterial supply to the sequestered segment is from the descending thoracic aorta in the majority of cases, but in 20% of cases this is from the abdominal aorta, coeliac axis or splenic artery (Felker and Tonkin, 1990).

Although the intralobar sequestration lacks normal communications with the tracheobronchial tree, they can be ventilated by collateral drift or through fistulous bronchial communications which

Table 1. The Pryce classification system for intralobar lung sequestrations

Pryce type	Features
1	Anomalous artery supplies functional lung tissue which communicates with the tracheobronchial tree
2	Artery supplies both normal lung tissue and non-functional non-communicating tissue
3	Artery supplies lung tissue isolated from the tracheobronchial tree

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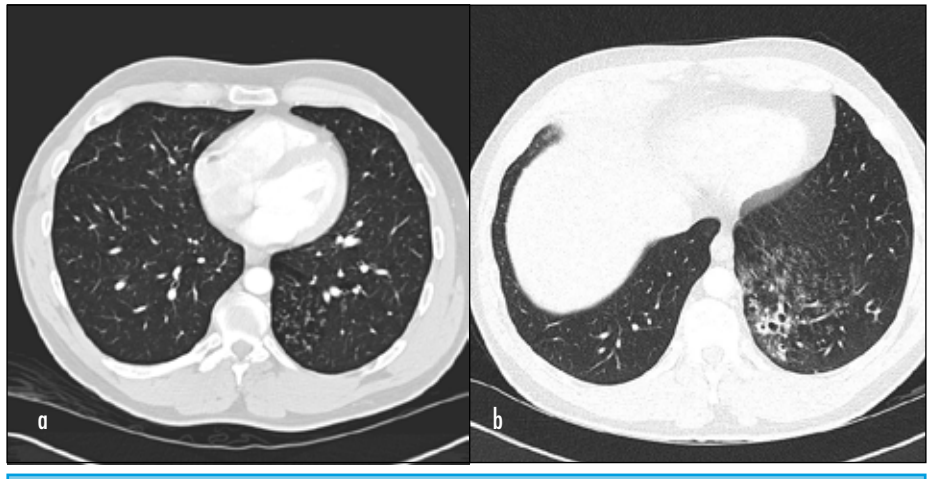
Figure 1. a. Computed tomography pulmonary angiogram with high resolution cuts. Left basal area showing bronchiectasis and air trapping, but no apparent connection of airways with remaining lung suggesting intralobar lung sequestration. There is partial visceral pleural cover to this sequestered segment. b. Lower cuts showing features of gross bronchiectasis (signet rings and bronchial wall thickening) in sequestered lung.



Case Report

A 44-year-old former paper mill worker who had childhood asthma and allergic rhinitis was referred with 12 months of increased exertional dyspnoea and frequent chest infections (four per year) with chronic mucoid sputum production. He had never smoked and there were no other occupational exposures. Physical examination was significant for polyphonic expiratory wheeze and left basal crackles. Resting saturations were 95%, dropping to 89% on exertion. Spirometry and an electrocardiogram were normal. A chest radiograph showed no infiltrates. Full lung function tests showed a minor reduction in small airway function but preserved gas transfer with no air trapping. The concern remained of chronic pulmonary embolic disease with possible localized bronchiectasis.

He underwent computed tomography pulmonary angiography with high resolution cuts of the bases. This revealed an aerated segment of lung in the left base with bronchiectasis and air-trapping, the bronchi and bronchioles of which did not connect to the main tracheo-bronchial tree (*Figures 1a* and *b*). This segment had a partial visceral pleural cover, but was otherwise encased within the lower lobar visceral pleura. The venous drainage was through the pulmonary veins to the left atrium, but the arterial supply was from the systemic circulation, although the entire course of this was not demonstrated; a formal computed tomography angiogram was performed at a later date to include the whole aorta. The appearances were consistent with an intralobar sequestration in the left lower lobe, with an arterial supply from the abdominal aorta very close and to the left of the coeliac trunk (*Figure 2*).

Because of his attributable systemic symptoms, he was referred to a thoracic surgeon to consider resection of the sequestration to improve his symptoms and prevent more serious complications such as massive haemoptysis. The patient has currently declined surgery after informed discussion but is being monitored. To date, there have no episodes of major haemoptysis.

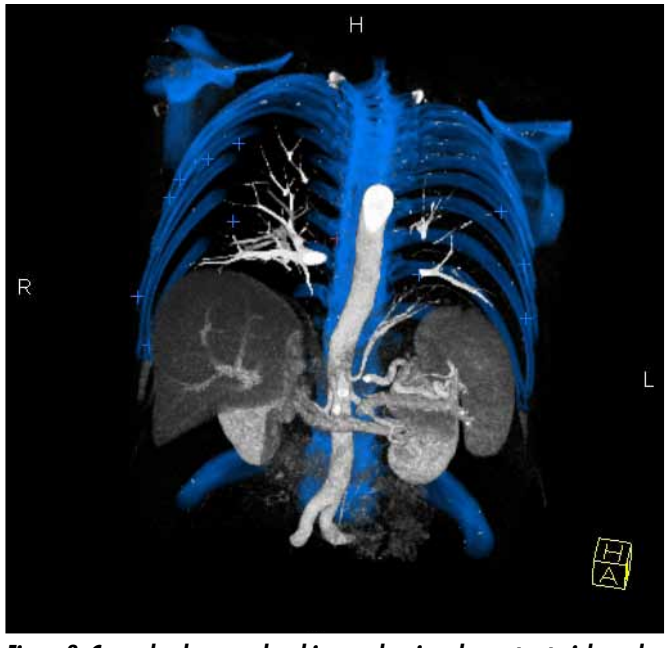


Figure 2. Coronal volume rendered image showing aberrant arterial supply to sequestration noted from abdominal aorta.

develop after infection (Stern et al, 1991). This would account for the aeration of the sequestered lung in this case.

The pathogenesis of intralobar sequestration is not fully understood but it can be acquired, as opposed to extralobar sequestration which is congenital. Patients often present as young adults. Left lower bronchial obstruction (such as by foreign body; Stocker and Malczak, 1984) may lead to post-obstructive pneumonia with partial or complete interruption of the normal pulmonary arterial supply to the affected lung tissue with parasitization of the neighbouring pulmonary ligament arteries to create an anomalous arterial supply.

Presentation is usually as recurrent infections with haemoptysis which can be severe and more likely to occur with increasing age (Rubin et al, 1994; Hirai et al, 2007). The pathophysiology involves a high pressure delivery of arterial blood from the systemic circulation to an area of lung not connected with the normal tracheobronchial tree, which is may often be extensively damaged with bronchiectasis, and then being returned via the pulmonary veins to the left atrium. Therefore, ventilation/perfusion mismatch is exaggerated accounting for exercise-induced desaturation and high cardiac output failure can ensue to a shunt from the systemic artery back to the left atrium, i.e. a left to left shunt. A murmur is sometimes heard over the area of shunting. The shunt can increase with age and can present later as gross heart failure if unrecognized (Fabre et al, 1998). The supplying artery comes from the descending thoracic aorta in 73% of cases; the coeliac artery (as in this case) is the origin in 21% of cases.

There is no effective medical treatment which can only be guided at the secondary effects of the sequestration (bronchiectasis and infection). Surgery is the only treatment of choice in most cases (embolization has rarely been used as discussed below). Computed tomography angiography is generally the most helpful imaging modality to aid surgical planning. The indications for surgery are recurrent infections and/or haemoptysis, massive haemoptysis or compressive symptoms because of volume effects of the sequestered lung. There is also a strong argument for considering surgery even in the asymptomatic patient although this is still not done universally (Laberge et al, 2005). The argument for surgery in the asymptomatic patient would be to prevent massive haemoptysis or recurrent infections, and the development of significant heart failure. In reality, many adult patients will have already had symptoms to justify their imaging which leads to the diagnosis.

Surgery is often a lower lobectomy for intralobar sequestration by thoracotomy, although it can be done by video-assisted thoracoscopic surgery which may reduce postoperative pain and length of stay compared to open thoracotomy and be of particular relevance to older patients with more co-morbidities (Tsang et al, 2006; Gonzalez et al, 2010). Embolization can be performed for type 1 sequestrations (Lee et al, 2003). BJHM

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LEARNING POINTS

- Sequestration is an important cause of unexplained bronchiectasis and recurrent chest infections and should be considered especially in unexplained left lower lobe bronchiectasis.
- The initial chest radiograph may be normal especially if the lower lobe bronchiectasis is mild.
- If there are significant systemic symptoms, resection is required to prevent more serious complications such as massive haemoptysis.
- It is important consider the possibility of resection even if there are no symptoms to prevent long-term complications.
- Computed tomography angiography is the most helpful imaging modality to assess sequestration and plan surgery.
- High output cardiac failure can develop as a result of shunting from the systemic arterial supply into the pulmonary veins.