

A Rare Case of Right Lower Lobe Intralobar Pulmonary Sequestration in an Adult

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ABSTRACT

Pulmonary sequestration is a rare congenital abnormality that is often diagnosed in childhood. There are two types of sequestration which are differentiated by the presence, or lack of, pleural lining dividing the aberrant lung tissue from the functional lung. Data for the management of this pathology in adults remains scarce. Here we present a case of intralobar pulmonary sequestration in an adult. The management of this patient comprised of surgical treatment involving lobectomy and ligation of the feeding vessel. The patient had an uneventful recovery, and the final pathology revealed a chronic infection in the sequestration.

KEYWORDS

Pulmonary sequestration, Right lower lobe lobectomy

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INTRODUCTION

Pulmonary sequestration is a rare congenital abnormality of the lung that has been known for over 100 years. It was first described by Rektorzik in 1861 but was further elucidated as a clinical and pathological condition by Pryce in 1946.¹ The abnormality is one in which a non-functional segment or lobe of the lung is separated from the normal bronchopulmonary tree and receives an aberrant arterial supply and venous drainage. The aberrant blood supply most often arises from the thoracic (75%) or abdominal (20%) aorta but may also arise from smaller thoracic or abdominal arteries.² There are two types of pulmonary sequestration. Intralobar sequestration is more common and is when the abnormal lung tissue shares a common pleura with the lung. Extralobar sequestration is when the aberrant tissue is separated from the remaining lung tissue by its own pleural investment.³ Most pulmonary sequestrations are diagnosed in early childhood. The more common intralobar sequestrations are usually in the lower lobes and may present with recurrent pneumonia, with radiographic

evidence of a consolidative mass in the lower lobe. Here we present a case of intralobar pulmonary sequestration in an adult, which presented with an insidious onset but classical signs on CT scan, and the accompanying successful surgical treatment.

CASE PRESENTATION

The patient was a 42-year-old female that was referred to thoracic surgery due to a long-standing diagnosis of pulmonary sequestration that was found incidentally on CT scan in 2009. CT scan of the chest showed pneumonia with findings of right lower lobe sequestration. At that time, she was experiencing fatigue with some shortness of breath during light activity. As a school teacher, she experienced further fatigue over the ensuing years with some mild chest discomfort. She presented again in 2020 to the thoracic surgery clinic. CT scan revealed a systemic artery arising from the right lateral aspect of the aorta, which terminated in an intralobar pulmonary sequestration (Figure 1). The sequestration drained back into the inferior pulmonary vein. The sequestration involved the posterior basal, lateral



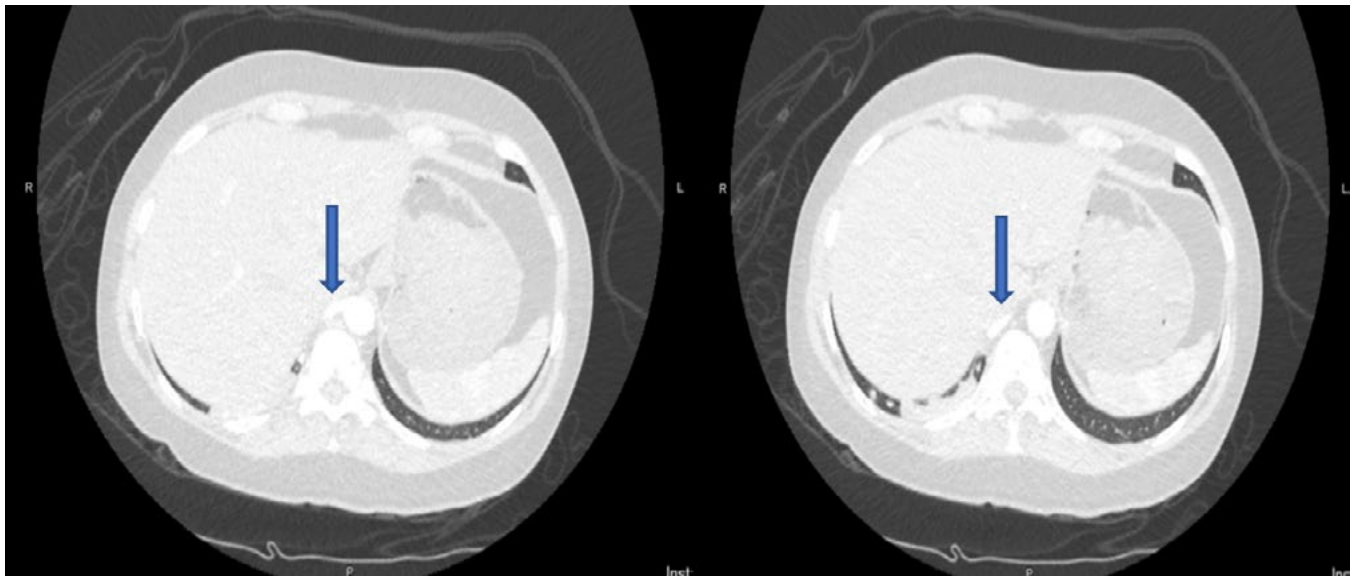


FIGURE 1. Axial CT scan of chest with IV contrast demonstrating take off of feeding vessel of the sequestration coming off the aorta.

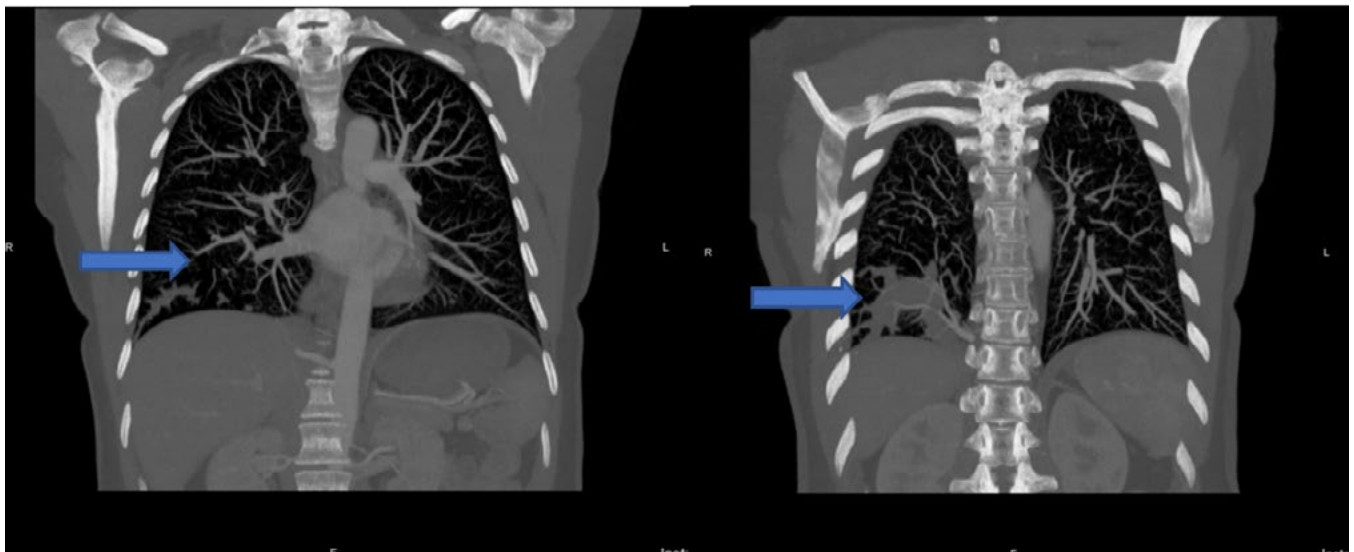


FIGURE 2. Axial CT scan of chest with IV contrast further demonstrating the separate blood supply of the sequestration.

basal, and some of the medial basal segments of the right lower lobe of her lung (Figure 2).

A pre-operative workup found her pulmonary function tests were within normal limits. She underwent a ventilation-perfusion scan which demonstrated no segmental or subsegmental defects. She also underwent a bronchoscopy pre-operatively and was noted to have no airway abnormalities, no congenital abnormalities, nor any areas of bleeding nor telangiectasia, specifically

in the right lower lobe. Washings were taken and aspirated and sent off for tuberculosis, fungal, and routine cultures which showed no growth. Clinical examination was within normal limits.

On the day of surgery, she underwent a paravertebral block, intubation with a double-lumen tube for ventilation, and general anesthesia for the procedure. The surgery was approached from a standard right posterolateral thoracotomy. Dissection showed that the main branch of the



pulmonary artery fed into the lower lobe of the right lung, whereas the sequestered area in the lower lobe was fed from an artery coming directly through the diaphragm into the mediastinum and the lower lobe. This aberrant artery was gently encircled using a vessel loop. With as minimal manipulation as possible, the aberrant artery was stapled off, followed by the inferior pulmonary vein and pulmonary artery. The dissection was completed around the lower right lobe fissures, and the bronchus, as well as lung tissue, were stapled. Mediastinal lymph node dissection was performed. An ON-Q pump was left for pain control at the end of the case.

Intraoperative findings were consistent with the CT scan findings of the patient having a right lower lobe pulmonary sequestration with feeding vessel from the abdominal aorta through the diaphragm into the mediastinum and the lower lobe directly with symptoms. Final pathology was consistent with the clinical impression of pulmonary sequestration. She did very well postoperatively and was discharged home after an uncomplicated hospital course.

DISCUSSION

The diagnosis of pulmonary sequestration is often made in childhood or early in adulthood as it presents with frequent infections or, more rarely, hemoptysis.^{4,5} The exact etiology is not known, but it is suggested that it is the result of an abnormal accessory tracheobronchial bud evaginating from the foregut.⁶ Intralobar pulmonary sequestration, as with our patient, shows no preference for one side of the thorax versus the other. Extralobar pulmonary sequestration is reported to be on the left side in up to 80% of cases. The exact etiology of side preference is not known, and bilateral sequestration is rare.⁷ Intralobar pulmonary sequestration accounts for 75-90% of sequestrations, with 20% occurring in the right lower lobe. The extent of sequestrations is variable.⁸

Most patients with pulmonary sequestration will remain asymptomatic which makes the true incidence difficult to discern. Of symptomatic patients, typical presenting symptoms include chronic cough, chest pain, recurrent pneumonia,

pulmonary abscess, as well as hemoptysis.⁹ For unknown reasons, but likely due to the non-functionality of the tissue, the sequestered tissue is prone to infections. Congestive heart failure has been reported in the literature, which is due to left to right shunting.

Radiologic manifestations of pulmonary sequestration include recurrent focal airspace disease, a parenchymal mass, and air or fluid-filled cystic lesions. Intralobar sequestration typically manifests radiologically as a homogenous consolidation with irregular margins. Angiography and MR imaging modalities can also help define the anomalous vascular supply for surgical planning.¹⁰

It is generally accepted that most patients with pulmonary sequestration should be treated operatively with lobectomy regardless of symptoms. This is in part due to the risk of developing recurrent infections. This, however, remains a source of debate. A recent retrospective review of pulmonary sequestration comparing resection versus observation was not able to determine the benefit of one over the other. The paper reported that various factors, including clinical manifestations related to pulmonary sequestration, risk of surgical complications, comorbidities, and individual patient preferences, need to be considered on a case-by-case basis.⁵

A more recent treatment modality for pulmonary sequestrations is transarterial embolization. Reported cases have utilized a combination of polyvinyl alcohol (PVA) particles, gel foam, and coils with good success.⁴ Embolization works by occluding the arterial supply leading to sequestration infarction and is an alternative treatment method to conventional surgical resection. Some concerns have been risen from these techniques due to pain and risk of infection of infarcted pulmonary tissue. Embolization can also be used pre-operatively to decrease the risk of intraoperative hemorrhage, but its benefit remains theoretical.¹¹

The patient in the present study was treated surgically and did well. On pathological analysis of the resected specimen, she did have evidence of continued acute on chronic infection. Her symptoms, however, were deceptively few. It could only be



supposed that the infection would eventually progress into a more serious issue should the sequestration have been treated conservatively. Discussions with the patient have confirmed our impressions, and having a good outcome has confirmed our analysis of the risk-to-reward ratio.

CONCLUSION

Pulmonary sequestration is a rare pathological finding that, when diagnosed in adulthood, is often found incidentally on imaging but should be suspected in patients with recurrent pneumonia. It is diagnosed definitively with imaging modalities and is most often treated with surgical resection. A pre-operative CT scan of the thorax with IV contrast is vitally important for surgical planning to avoid injuring the aberrant vessels during the operation. The decision to operate needs to be multifactorial with considerations of the patient's clinical manifestations, risk of surgical complications, comorbidities, and patient preference evaluated on a case-by-case basis.

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