Lung Sequestration

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Erich Stoelben

Introduction

The first description of what is now known as lung sequestration is attributed to J. J. Huber in a 1777 issue of *Acta Helvetica*. The term *sequestration* was introduced by D. M. Pryce in the *Journal of Pathology* in 1946 (Pryce 1946). Pulmonary sequestration may be defined as a segment of the lung that has no macroscopically identifiable communication with the tracheobronchial tree. The most widely used concept explains pulmonary sequestration as a formation of an accessory lung bud caudal to the normal lung buds between the fourth and eighth weeks of gestation (Langston 2003). Normally, the sequestration is perfused by a systemic artery arising from the descending aorta. The intrapulmonary sequestration (Fig. 21.1) is integrated in the normal lung (nearly always in the lower lobes, with a predominance in the left lower lobe) and drains into the pulmonary vein. Alternatively, the extralobar sequestration (Fig. 21.2) is isolated from the normal lung, located between the lung and diaphragm, in the mediastinum, or intra-abdominally. The extralobar sequestration drains into systemic veins and is mostly combined with other malformations, such as diaphragmatic hernia. With an incidence of 0.15–1.8 %, pulmonary sequestration is one of the most common congenital lung anomalies, along with congenital lobar emphysema, cystic adenomatoid malformation, and bronchial cysts.

E. Stoelben
Lung Clinic, Metropolitan Hospital of Cologne,
Ostemerheimerstr 200, 50933 Cologne, Germany
e-mail: stoelbene@kliniken-koeln.de

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Figure 21.1

Intralobar sequestration

Figure 21.2

Extralobar sequestration

Figure 21.1

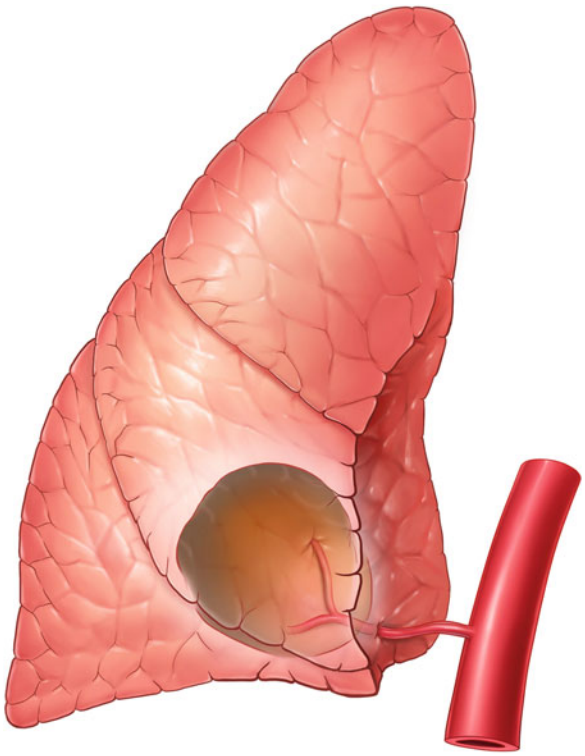
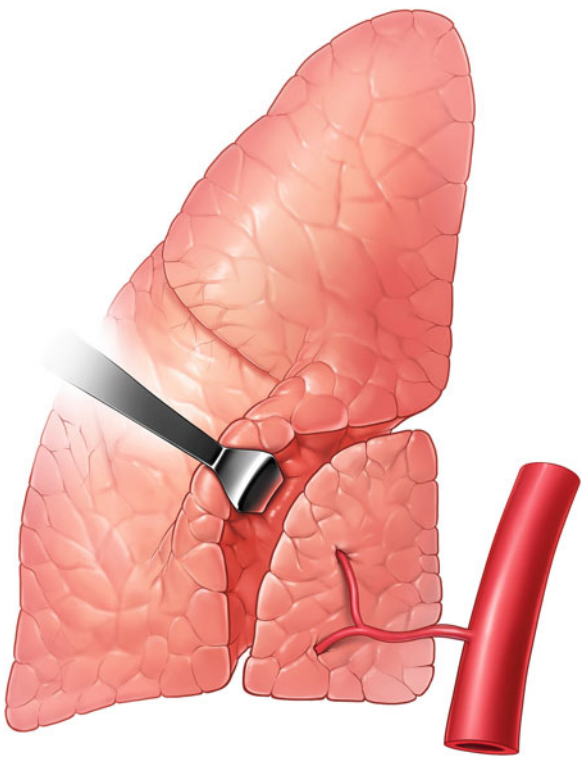


Figure 21.2



Clinical Presentation

The clinical presentation depends mainly on the type of sequestration. Extralobar sequestration may be detected antenatally by ultrasound. Otherwise, the malformation becomes clinically apparent through respiratory symptoms induced by concomitant anomalies or severe arteriovenous shunting. Male children are more affected, and the left lower part of the thorax is mostly involved; the diagnosis is made within the first year of life. Intralobar sequestration is not detected until recurrent bronchopulmonary infection leads to radiologic examination; therefore, most patients present in adulthood. Segmental infiltration in the dorsal part of the lower lobes in otherwise healthy patients should lead one to suspect this malformation (Corbett and Humphrey 2004). MRI with visualization of the aberrant vessels is used to confirm the radiographic and ultrasound diagnosis (Fig. 21.3).

Figure 21.3

Frontal magnetic resonance image of the thorax of a 5-month-old boy with hemoptysis and intralobar sequestration. The important systemic blood flow is drained by the lower pulmonary vein. Contrast-enhanced radiologic examinations has to be checked for atypical vessels arising from the descending aorta in all cases of infiltration of the lower lobes, especially in cases with recurrent disease. The anatomy of the pathologic vessel has to be realized to plan the surgical procedure. Since the blood is drained by the veins of the lower lobe, the sublobar resection of the sequestration should respect the venous flow of the healthy lung to avoid lung infarction

Surgical Treatment

In every case, the thoracic surgeon must be aware of the anatomic details of the aberrant vessels. In extralobar sequestration, resection of the supplementary lung is combined with correction of other anomalies, such as diaphragmatic hernia, other lung malformations, and cardiac or vascular abnormalities. Intralobar sequestration requires an atypical segmental resection or lobectomy, depending on the extent of the sequestration in the lobe (Fig. 21.4).

Identification and careful dissection of the systemic artery arising from the descending aorta is the crucial point in the treatment of lung sequestration. In extralobar sequestration the resection of the supplementary lung is combined with correction of other anomalies like diaphragmatic hernia, other lung malformations and cardiac or vascular abnormalities. Intralobar sequestration requires an atypical, segmental resection or lobectomy depending on the extent of the sequestration in the lobe. The procedure can be performed in open or video-assisted technique (Fig. 21.5). In longstanding pulmonary infection in the sequestration, the parenchyma of the surrounding otherwise healthy lung may be affected by fibrosis and lobectomy can be necessary (Adzick et al. 2003).

Figure 21.3

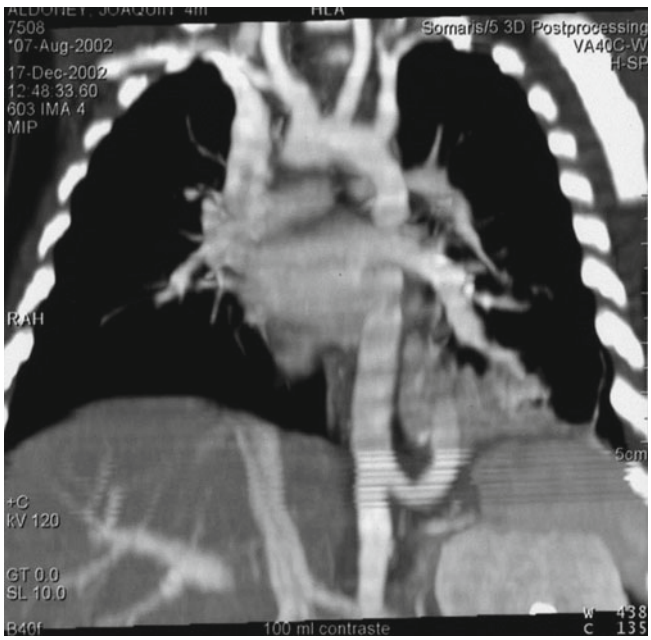


Figure 21.4

Computed tomography (CT) scan of the thorax of a 5-month-old boy with hemoptysis and intralobar sequestration. The CT scan shows a typical case of intrapulmonary sequestration with infiltration of the lung imitating pneumonia or atelectasis. Infiltration of the lung is due to intrapulmonary haemorrhage. The blood flow by the systemic artery may be important and produce a left to right shunt with cardiac strain leading to early diagnosis in life. More often the diagnosis is made by recurrent pulmonary infection in the same lower lobes. Bronchial obstruction has to be excluded by bronchoscopy

Figure 21.5

A 38-year-old female patient with recurrent bronchopulmonary infection and intralobar sequestration. This intraoperative thoracoscopic view of the left lower lobe shows a lack of anthracosis in the lung sequestration not connected to the tracheobronchial tree

Figure 21.4

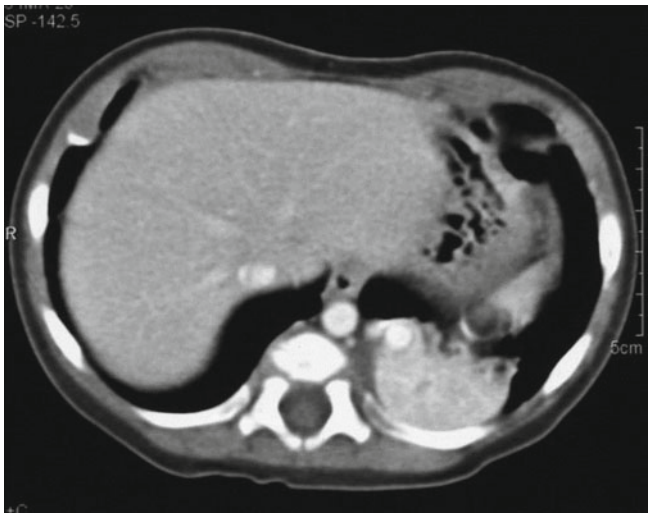
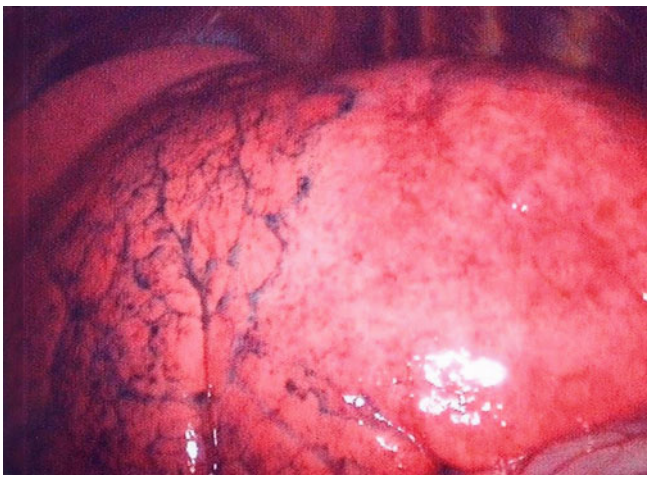


Figure 21.5



Discussion

Extralobar sequestration is diagnosed early in life by antenatal sonography or postpartal respiratory symptoms. Surgical procedures comprise resection of the malformation and correction of concomitant anomalies. Intralobar sequestration is a question of not to miss the diagnosis in patients with recurrent pulmonary infections. Otherwise the anatomical peculiarity can lead to an intraoperative challenge. Video-assisted thoracoscopic technique is adequate for vascular dissection and lung resection.

Selected Bibliography

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