

what's your diagnosis?

Diagnosis: Intralobar sequestration

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The chest CT scan with contrast (Figure 1) revealed large cystic parenchymal changes and a lesion with an aberrant aortic branch supplying the lesion. After a discussion of the risks and benefits of surgery with the patient, he elected surgical resection. At surgery, an intralobar sequestration (Figure 2) was found occupying almost all the right lower lobe with a feeding systemic arterial supply directly from the descending aorta along the inferior pulmonary ligament in the exact location shown on contrast CT (Figure 1), almost the size of the aorta. The pulmonary vein anatomy was normal, and the patient had a formal right lower lobectomy.

DISCUSSION

Pulmonary sequestration is a complex group of abnormalities involving the anomalous connection of the pulmonary parenchyma and the pulmonary and systemic vasculature. The term sequestration was first introduced by Pryce¹ in 1946. Classically, the sequestration may be intra- or extralobar, according to the relationship to the normal lung parenchyma. Although diagnosed more frequently, pulmonary sequestration remains an extremely uncommon entity. Intralobar sequestration, however, is the most common form of pulmonary sequestration, accounting for 75% of cases. Over 50% of cases, however, become symptomatic after the age of 20 years.²

Arteriography is considered the gold standard for identification of the aberrant artery. Newer, less invasive imaging techniques are equally effective and safer alternatives to angiography. These include CT angiography, Doppler ultrasound, and magnetic resonance angiography (MRA). Color-enhanced, three-dimensional MRA can identify both the arterial and venous aberrancies and define the soft tissue abnormalities associated with sequestration.^{3,4} Multiplanar three-dimensional reconstruction of the CT angiogram can demonstrate the entire route of the anomalous vessel and define the consolidation of the sequestered lobe.⁵ These less invasive imaging techniques can provide additional information over traditional angiography and they should be used routinely in the preoperative evaluation of sequestrations.

The differential diagnosis includes congenital or acquired lung cyst, tumors of the posterior mediastinum, Bochdalek hiatal hernia and congenital diaphragmatic cyst. Treatment of intralobar sequestration, whether symptomatic or not is lobectomy or segmentectomy.^{10,7} Wedge resection and control of an abnormal systemic artery is the operation of choice for extralobar sequestration.^{6,7}

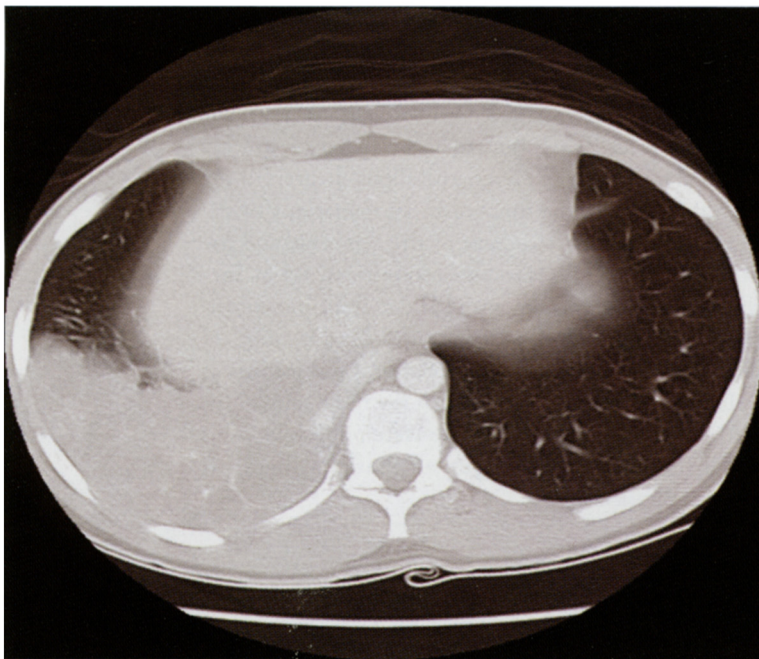


Figure 1. Contrast CT scan

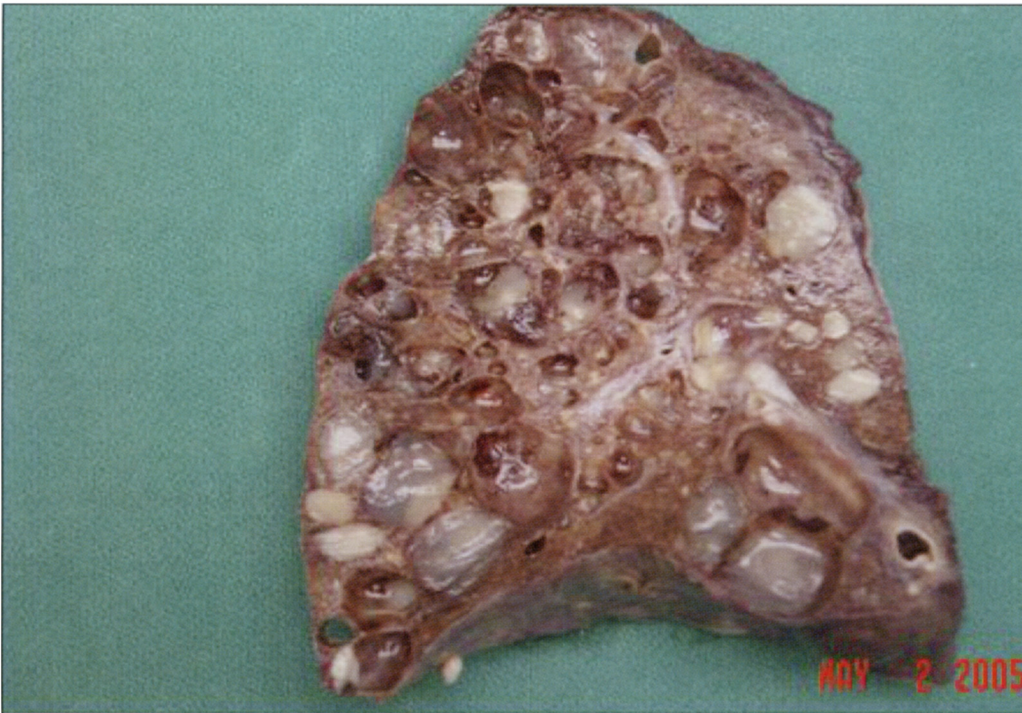


Figure 2. Gross pathology showing multicystic lesions filled with mucus.

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