

## Unilateral Coexisting Intralobar and Extralobar Pulmonary Sequestrations

S. Pinar Karakas, M.D., Stuart Morrison, M.D., and Oliver Soldes, M.D.

Pulmonary sequestration is a focal area of nonfunctioning, dysplastic lung parenchyma that lacks communication with the normal tracheobronchial tree. It is supplied by the systemic arterial circulation and has two types, intralobar and extralobar, that can be differentiated from each other by the pleural covering and the venous drainage. Their coexistence is extremely rare. We report the imaging findings of a patient who had coexisting but completely separate intralobar and extralobar sequestrations at the left lower lung. We elucidated the complex vascular anatomy using three dimensional volume rendering and multiplanar reconstructions from a 64-detector helical CT scanner.

### Introduction

Pulmonary sequestration is a focal area of nonfunctioning, dysplastic lung parenchyma that is supplied by a systemic artery [1]. It has neither communication with the normal tracheobronchial tree nor the pulmonary arteries. The sequestration has two types intralobar and extralobar sequestrations, that can be differentiated from each other by the nature of its pleural covering and the venous drainage. Their coexistence is extremely rare though a few case reports mention such an occur-

rence. Most case reports are about single lesions having both intralobar and extralobar sequestrations [2]. It is extremely rare to find a report of a coexisting but completely separate intralobar and extralobar sequestration [3].

### Case Report

A 7-month-old female was referred to our hospital pediatric surgery department for consultation of a lung mass identified on prenatal ultrasound and postnatal chest CT. Due to advanced maternal age a prenatal ultrasound was performed at 19 weeks of gestation which visualized a multilocular cystic mass in the left lower lobe thought to represent a congenital cystic adenomatoid malformation. A CT exam was done in the neonatal period which confirmed the lung mass in the left lower lobe. No images or further details were available regarding these imaging studies since they were done at an outside institution. The patient had a normal physical exam and laboratory values at presentation to our hospital. Imaging was performed in the arterial phase with a 64 detector computerized tomography

Citation: Karakas SP, Morrison S, Soldes O. Unilateral coexisting intralobar and extralobar pulmonary sequestrations. *Radiology Case Reports*. [Online] 2008;3:171.

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

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Published: August 9, 2008

DOI: 10.2484/rcr.v3i3.171

**RCR** Radiology Case Reports | radiology.casereports.net

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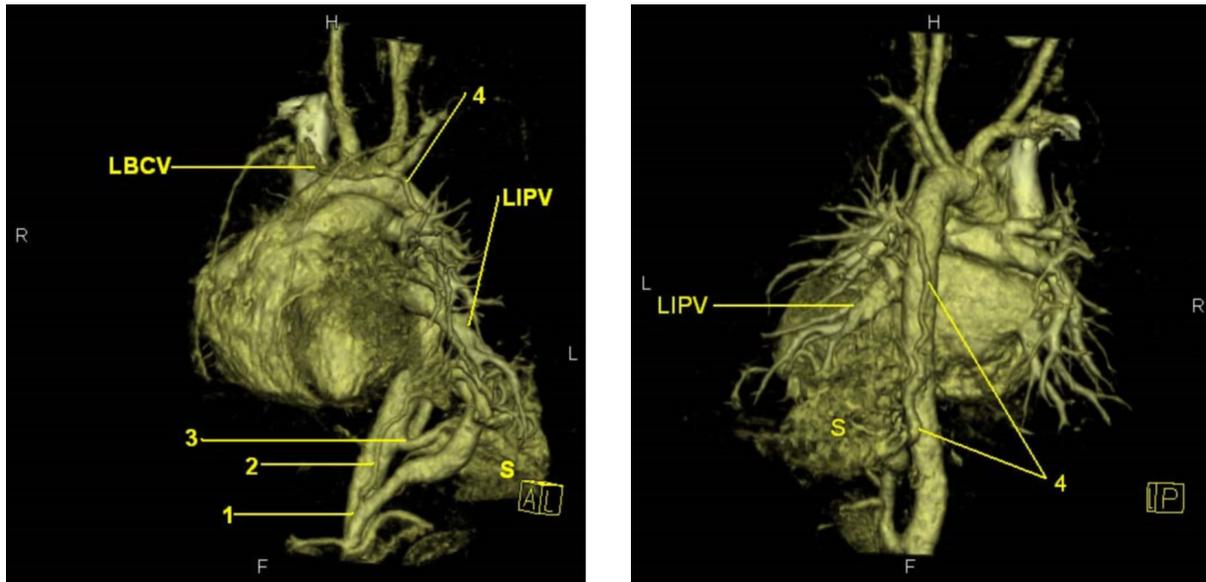


Figure 1A-B. Three-dimensional volume-rendered images with surface shading in A) sagittal and B) posterior views. The celiac artery origin of the large caliber artery (1) is clearly seen as well its smaller caliber branch (2). An anomalous artery originates directly from the thoracic aorta (3). The anomalous systemic vein (4) associated with the extralobar sequestration is seen originating from the sequestration (s) and draining into the incompletely filled left brachiocephalic vein (LBCV). Left inferior pulmonary vein (LIPV) is draining the intralobar sequestration.

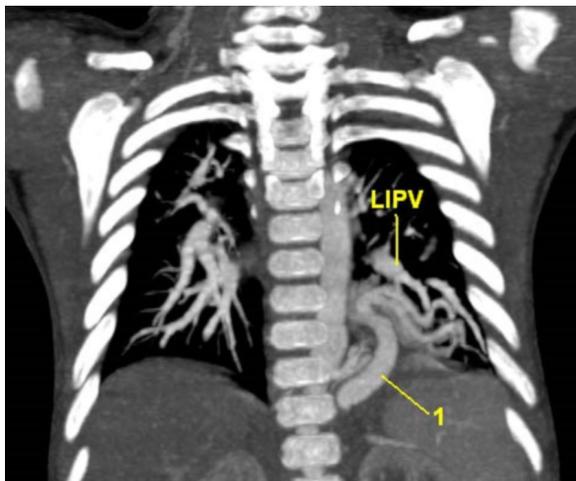


Figure 2. Coronal maximum intensity projection (MIP) image in soft tissue window. The sequestration is seen as an abnormal enhancing soft tissue mass at the left lower lobe. The large caliber artery (1) is arising from the abdomen. The left inferior pulmonary vein (LIPV) is seen draining the sequestration.

(CT) (Somatom Sensation, Siemens, Germany) following uneventful administration of 15 cc of Ultravist 240. The scan parameters were 100 kVp, 40 mAs and 1 mm collimation. Three dimensional (3-D) volume-rendered images were obtained using a freestanding workstation (Leonardo, Siemens, Germany). Sagittal and coronal multiplanar reconstructions were prepared with 50% slice overlap eliminating the stepping artifact.

The CT scan showed an abnormal, heterogeneously enhancing solid lesion in the left lower lobe posteriorly with surrounding emphysematous area manifested by parenchymal architectural destruction and air trapping. The lesion had anomalous systemic arterial blood supply with both pulmonary venous return and abnormal systemic venous return (Fig. 1). The pulmonary venous return was via the left inferior pulmonary vein draining into the left atrium. The anomalous systemic venous return was via a small vessel that extended posterior to the descending thoracic aorta then crossed over the left aortic arch and drained into the left brachiocephalic vein. At least two vessels carried systemic arterial supply to the lesion. The superior and smaller vessel originated directly from the descending thoracic aorta.

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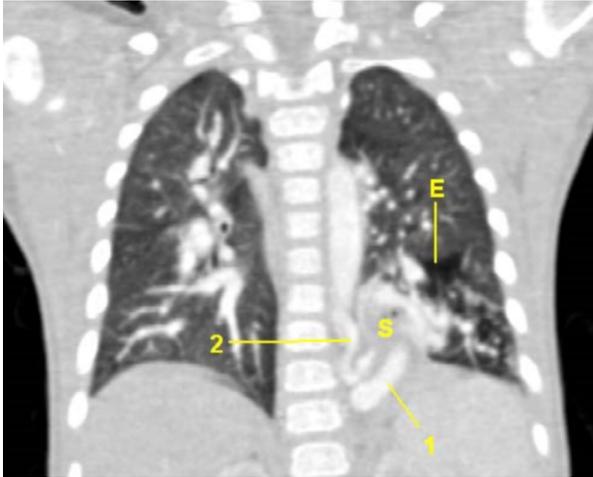


Figure 3. Coronal reformatted image with 50% overlapping slices in lung window. There is emphysematous area (E) surrounding the sequestration (s). The large caliber artery originating from the aorta (1) and anomalous systemic vein originating from the sequestration (2) are seen.

The second much larger arterial vessel originated from the celiac artery, extended superiorly and penetrated the left diaphragm before reaching the center of the lesion (Fig. 2). No normal pulmonary arterial supply to the lesion was seen. The lesion was identified as an unusual sequestration with two separate systemic arterial supplies and both pulmonary and systemic venous returns. The surrounding emphysematous area raised the possibility of concomitant congenital cystic adenomatoid malformation (Fig. 3). In light of these findings a unifying diagnosis of a bronchopulmonary foregut malformation was given.

At surgery, two completely separate masses were identified: an intralobar sequestration within the left lower lobe and a completely separate extralobar sequestration in the left lateral sulcus above the diaphragm. The intralobar sequestration was supplied by the large systemic blood vessel coming up through the left diaphragm and drained into the left inferior pulmonary vein. The extralobar sequestration was supplied by the smaller arterial vessel arising from the thoracic aorta and drained into the anomalous systemic vein.

The pathology examination defined the specimens as intralobar and extralobar sequestration based on their arterial supply and pleural covering. The left lower lobe intralobar sequestration was noted to contain a dilated

bronchus containing inspissated mucous with histologic features of a bronchial atresia that was associated with the emphysematous area. A congenital cystic adenomatoid malformation was not identified by pathology.

### Discussion

Interruption of the normal lung development at different stages of development results in varying abnormalities in the affected area of lung, now universally termed as bronchopulmonary foregut malformation. To understand the pathophysiology of sequestration it is important to understand the lung embryology. The lung bud originates as a diverticulum from the foregut endoderm on day 22. Starting on day 26 it undergoes multiple divisions to generate the respiratory tree of the lungs. At week 16 the blind ending terminal bronchioles form which later divide between 16 and 28 weeks to form the respiratory bronchioles. The primitive alveoli begin to form between weeks 28 and 36 not reaching maturity until about 8 years of age. Sequestration has been linked to other congenital lung anomalies such as congenital cystic adenomatoid malformation and bronchial atresia, as seen in our case, with an incidence reaching up to 80% [7]. The high incidence of associated bronchial atresia is a significant finding that supports the theory of fetal airway obstruction in the pathogenesis of sequestration [7]. The tissue surrounding the respiratory bronchioles becomes highly vascularized by capillaries from the primitive systemic circulation, a vascular plexus that has anastomotic connections to the primitive ventral and dorsal aortas. As the lung grows these vessels regress to form the bronchial arteries. Concurrently, the pulmonary artery arises from the sixth branchial arch to become dominant [5]. Sequestration may result from an early interruption of the pulmonary arterial tree development while the primitive systemic capillary supply continues to mature [7]. The early venous drainage of the lungs is to the cardinal veins, primarily to the anterior cardinal vein. Later, the pulmonary veins develop from the region of the left atrium and become the main drainage route.

Clinical suspicion of pulmonary sequestration usually begins with a solid and/or cystic lung mass on prenatal sonography, recurrent pneumonia (almost always intralobar sequestration), or nonresolving incidental lung opacity on sequential chest radiographs. Extralobar sequestration occasionally presents with congestive

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heart failure and pulmonary overcirculation which may manifest as polyhydramnios in utero and as anasarca in infancy. The lower lobes, are the most common location for sequestration being most common in the left lower lobe. The intralobar sequestration (85%) is more common than the extralobar sequestration (15%) [3]. The extralobar sequestration has its own distinct pleural covering that separates it from the adjacent normal lung. The intralobar sequestration is embedded within the normal lung and contained within the normal visceral pleura. The extralobar sequestration is almost exclusively identified in infancy and it is more commonly located on the left side particularly at the left posterior costophrenic groove. The extralobar sequestration can be located below the diaphragm with a higher chance of coexisting gastrointestinal abnormalities, diaphragmatic hernia, and congenital cystic adenomatoid malformation [7]. The intralobar sequestration almost always involves the medial and posterior basal segments of the lower lobes of the lungs, also more common on the left [7].

The diagnostic imaging for sequestration is based on recognition of the presence and location of an aberrant systemic artery. In either type the aberrant arterial supply most commonly arises directly from the descending aorta (78-80%). In remaining cases it may arise from other systemic arteries, most commonly the celiac axis [7]. Multiple supplying arteries are seen in about 16% of cases [7]. In 5% of extralobar sequestration cases, the blood supply may be pulmonary or both pulmonary and systemic in origin [7]. In 95% of intralobar sequestration cases, the venous drainage is to the left atrium via pulmonary veins. In less than 5% of intralobar sequestration cases, the venous drainage is to the systemic circulation. In the majority of extralobar sequestration cases, the venous drainage is typically via systemic veins. In both types the venous drainage occurs commonly to the azygous and hemiazygous veins but drainage to the other systemic veins has been reported. The venous drainage of an extralobar sequestration could occasionally be to the pulmonary veins or less often to both the systemic and pulmonary circulation. The imaging based differentiation of supradiaphragmatic intralobar versus extralobar sequestration depends on identification of the anomalous venous drainage although this is not always reliable due to variability of the anatomy.

In the absence of indications for immediate resection such as respiratory difficulty or recurrent infections, the lesion is usually removed approximately at age 6-7

months when the child's tolerance to elective surgery and anesthesia is better. Early resection may facilitate compensatory lung growth and eliminate the need for surveillance with radiation exposure.

Correct presurgical identification of the lesion and description of the anatomy are important for the operative planning. An infradiaphragmatic lesion requires a transabdominal approach. As an intralobar sequestration requires lobectomy, an extralobar sequestration can usually be removed sparing the adjacent lung tissue. Recognition of aberrant vessels will prevent inadvertent intraoperative bleeding.

The detailed anatomy of sequestration is best recognized with cross sectional imaging modalities such as CT and magnetic resonance imaging (MRI). MRI requires anesthesia for infants and younger children and is limited in evaluation of lung parenchyma. Currently, CT is the optimum imaging modality especially with the utilization of low dose techniques with weight based milliampere per second method [8]. Volume acquisition with multidetector helical CT nearly eliminates the need for sedation and allows exquisite demonstration of the vascular anatomy and lung parenchyma with overlapping slice reconstruction, multiplanar display and 3D volume rendering. Our evaluation of the angioarchitecture was limited with axial images alone especially for the small caliber, obliquely oriented anomalous arteries and veins. Their course was easily recognized with volume rendering and multiplanar reconstruction images allowing recognition of the true nature of the extralobar sequestration and intralobar sequestration vascular components. In real time the 3-D images can be easily rotated so that the anomalous vessels can be clearly followed from the origin to the site of drainage.

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