

## CLINICAL VIGNETTE

# Extralobar Pulmonary Sequestration Presenting as Abdominal Pain

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### Case

The patient is a 32-year-old schoolteacher who presents with onset of right upper quadrant pain. She had complained of low back pain for the past 2 weeks that was attributed to poor posture and activities associated with her job. On the day of admission, she was laying on the floor doing stretching exercises when she experienced sudden onset of acute right upper abdominal pain radiating toward the back. There was no fever, chills, diarrhea or dysuria. She had vomited twice that was attributed to severe pain. The pain was exacerbated by laying supine and alleviated in part by decubitus fetal positioning. It was made worse by deep inspiration, but the patient denied dyspnea.

Past medical history was otherwise unremarkable. She exercises avidly and denied alcohol, tobacco or illicit drug use. Oral contraceptives were the only prescribed medication.

Physical examination revealed an alert, oriented female in acute distress secondary to abdominal pain, lying on her left side in fetal position.

The patient was afebrile. Vital signs revealed HR 90/min RR 16/min BP 108/56 with pulse oximetry of 98% saturation.

Auscultation of the lungs and cardiac exam were unremarkable. Abdominal examination revealed hypoactive bowel sounds. The abdomen was soft and nondistended. Palpation revealed severe right upper quadrant and periumbilical pain with equivocal rebound.

Laboratory examination revealed WBC 8.2 with normal differential, Hgb 12.5 gm/dl, and platelets 327,000. Serum electrolytes, BUN and creatinine

was normal. Hepatic panel including transaminases, alkaline phosphatase and bilirubin were normal. Serum lipase was normal. Urine pregnancy test was negative and urinalysis was unremarkable.

An abdominal ultrasound revealed findings compatible with fatty infiltration of the liver. Gallbladder, biliary tree, kidneys and visualized pancreas were all normal. CT abdomen with IV and oral contrast revealed equivocal wall thickening of the transverse colon raising the possibility of colitis. The examination was otherwise normal except for a moderate right pleural effusion.

CT scan of the chest with contrast again revealed the presence of the pleural effusion. There was a soft tissue lesion within the medial pleural space measuring 6 cm sagittal x 2.6 cm AP x 4.8 cm transversely. No intrapulmonary masses, infiltrate or adenopathy was seen (Figure 1).



Figure 1

The patient underwent percutaneous CT guided needle biopsy which was nondiagnostic.

Thoracic surgery consultation was obtained and the patient underwent video assisted thoracoscopy. A large hemothorax of 800 cc was evacuated. The mass was located along the right paravertebral gutter with some inflammatory component adhering to the parietal pleura, but without obvious communication to the lung. A small vascular communication was noted arising directly from the aorta. The mass was excised. Pathological examination was consistent with extralobar pulmonary sequestration. Post-operative course was uneventful and the patient was discharged home in good condition.

### **Discussion**

Pulmonary sequestration is defined as non-functioning lung tissue that receives its blood supply from an anomalous systemic artery, as opposed to the normal pulmonary vascular system. Drainage is through the pulmonary veins.<sup>1</sup> It does not have connection with the normal bronchial tree. There are two types, intralobar and extralobar. Extralobar is defined by being enclosed in its own pleura, while intralobar shares the pleura from the native normal lung.<sup>2</sup> Pulmonary sequestrations are fairly rare. The overall incidence of lung malformations in the general population is estimated from 2.2-6.6% with pulmonary sequestration found in 0.15-1.8% of that cohort.<sup>1</sup> Intralobar sequestrations account for 75-85% of the total, with the left side being involved in 55-60% of cases. Extralobar sequestration has a male predominance of 80%.<sup>3</sup>

Clinical presentation is most often found in early infancy and pediatric populations. Respiratory distress due to a large sequestration occupying the thoracic cavity or recurrent respiratory infections from intralobar sequestrations are the most common symptoms that lead to the discovery of this entity.<sup>3</sup> Adults are most often asymptomatic, and often present with a pulmonary nodule found in the lower lobes on routine chest radiograph.<sup>4</sup> Hemothorax like our patient, is extremely rare with the first case report documented only fairly recently.<sup>5</sup>

Physical examination is often unrevealing due to the lack of normal communication with the bronchial tree. Interestingly the acute right upper abdominal pain in our patient was hypothetically felt to be due to diaphragmatic irritation caused by the hemothorax.

Diagnosis of pulmonary sequestrations in adults is most often made during pregnancy, often as incidental findings during routine perinatal ultrasonography. In infants, plain chest radiographs are taken for respiratory distress due to a large sequestration occupying the thoracic cavity, or for recurrent respiratory infections is the most common postnatal imaging study.<sup>4</sup> CT scan usually reveal a solid mass, either homogeneous or heterogeneous, but may appear cystic or cavitary, presumably secondary to recurrent infection.<sup>4</sup>

Surgical resection is recommended for large or symptomatic pulmonary sequestrations. Extralobar sequestrations can be removed without loss of functioning lung tissue. Intralobar sequestrations are enclosed in the normal lung visceral pleura, which would require lobectomy or segmental resection.<sup>6</sup> Identification and control of the anomalous arterial blood supply is essential to avoid significant morbidity due to uncontrolled hemorrhage.<sup>7</sup>

A proposed cause for hemothorax is pulmonary infarction secondary to torsion of the anomalous blood vessels.<sup>7</sup> A case report of a 13 -year- old girl with episodes of abdominal pain due to intermittent torsion with eventual development of infarction and hemothorax was proposed.<sup>8</sup> This could explain the etiology of our patient's intermittent back pain prior to her presentation with hemothorax.

In summary, pulmonary sequestrations, although rare, should be considered in the differential diagnosis of nontraumatic hemothorax.

### **REFERENCES**

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