Torsion of an extralobar pulmonary sequestration: A rare cause of acute chest & flank pain in an adolescent

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Abstract

Pulmonary sequestration is a rare congenital malformation with varied presentation, most commonly discovered antenatally or after evaluation for recurrent pneumonia. The current report describes the case of an adolescent female with an extralobar pulmonary sequestration (ELS) who presented with chest and flank pain due to torsion. Clinical, radiologic, operative and pathologic findings are reviewed. This entity is rare and specific radiologic findings may be absent, therefore a high index of suspicion for a sequestration should be maintained in the appropriate clinical setting.

1. Case report

A 15 year old previously healthy female presented to the emergency department with a 1 day history of severe, left sided flank pain and left lateral chest pain. She reported similar pain in the same location intermittently over the prior 3 months that was mild and treated successfully with NSAIDS. Her chest pain was pleuritic and increased with movement. She also reported 1 day of ankle pain due to torsion. Clinical, radiologic, operative and pathologic findings are reviewed. This entity is rare and specific radiologic findings may be absent, therefore a high index of suspicion for a sequestration should be maintained in the appropriate clinical setting.

Key words:
Pulmonary sequestration
Torsion
Posterior mediastinal mass

Pulmonary sequestration has been described as a congenital pulmonary malformation in which a mass of non-functioning lung tissue receives systemic arterial blood supply and does not have a demonstrable connection to the tracheobronchial tree. Sequestrations may have their own visceral pleural lining (extralobar) or may be invested by the visceral pleura of the functioning lung (intra-lobar). Pulmonary sequestrations are rare and are typically found in utero or present as recurrent pneumonias. We describe an unusual case of a 15 year old girl presenting with severe left chest and back pain due to a posterior mediastinal mass later found to be an extralobar pulmonary sequestration that had undergone torsion.

During her hospitalization, she was admitted to the medical unit where her workup revealed white blood cell count of 16,400 with 80% PMNs and an elevated CRP of 5.2 mg/dl. Due to the absence of localizing findings, magnetic resonance imaging (MRI) of the chest and abdomen was obtained (Fig. 2) and revealed a solid mass in the left posterior mediastinum in a paraspinous location measuring 1.7 x 3.7 cm. There was no enhancement post-contrast. A small, enhancing left pleural effusion was also identified. Initial laboratory studies revealed a WBC count of 16,400 with 80% PMNs and an elevated CRP of 5.2 mg/dl. The patient was admitted to Rady Children’s Hospital, San Diego and surgical consultation was obtained. A preoperative diagnosis of posterior mediastinal mass was made and the patient underwent left thoracotomy. Intraoperatively, a moderate pleural effusion was drained and the mass had an appearance suggestive of pulmonary sequestration that had undergone torsion and the lesion was subsequently resected.

A dominant feeding vessel was identified. The mass measured 3.7 cm in greatest dimension and was well demarcated from the adjacent pulmonary parenchyma. Pathologic examination (Fig. 4) revealed a well-circumscribed hemorrhagic infarction, focal cartilaginous tissue and a mucin-filled cystic structure containing foamy macrophages and inflammatory cells. Reactive fibroblastic proliferation and chronic inflammatory infiltrates with occasional eosinophils were seen.

Thick-walled vessels with infarction were noted at the surgical margin. Well-preserved dilated bronchioles, alveolar ducts and alveoli, which are usually seen in extralobar sequestration, were

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http://dx.doi.org/10.1016/j.epsc.2014.06.006
not observed due to severe infarction. These findings confirmed the clinical diagnosis (Fig. 4). The patient recovered without incident and was discharged 4 days later.

2. Discussion

Pulmonary sequestration is a rare congenital pulmonary malformation in which a mass of non-functioning lung tissue receives systemic arterial blood supply and does not have a demonstrable connection to the tracheobronchial tree. The extralobar variety is less common than the intralobar, accounting for only 15–25% of all sequestrations [1,2]. ELS is thought to occur as a result of an error during embryogenesis during which an extra lung bud arises from the foregut and migrates caudally [1]. Sixty percent of patients with ELS have associated anomalies such as CCAM, diaphragmatic hernia, bronchogenic cysts and cardiac anomalies [3,4]. As a result, most are discovered in the neonatal period or early childhood. Many cases are also now identified antenatally, making an adolescent presentation of an ELS rather unusual [5]. Sequestrations discovered later in life may be associated with malignant transformation or recurrent infections therefore elective resection is often recommended [6], even in asymptomatic lesions. ELS are more commonly on the left side and range in size from 0.5 to 15 cm [7].

Torsion of an ELS is exceedingly rare with only 7 prior pediatric cases reported in the literature [7–12]. Our patient presented with chest and flank pain localized to the area of the infarcted sequestration. She had no history of previous lower respiratory tract infections. We hypothesize that her 3 month history of pain may have been due to intermittent torsion, and a very similar case was reported previously in a 13 year old girl who had pain for 2 months prior to her torsion being discovered [8]. Although chest pain has been reported as a presenting symptom, abdominal pain has been reported in all prior patients as a hallmark symptom, which our patient did not have. Leukocytosis [10] and elevation of the CRP [7,12] have been reported,
although our patient is the first to have fever reported at presentation.

Chest radiography is typically non-specific in sequestration that has undergone torsion, as was the case in our patient. Prior cases have reported lower lobe opacities, small pleural effusions and normal plain films [5,7,9,10,12]. Cross-sectional imaging with computed tomography or MRI is frequently used to further evaluate such cases and usually reveals a homogenous, well circumscribed, posterior mediastinal mass. The dominant blood vessel may or may not be identified. In ELS not associated with torsion, the dominant vessel is more commonly seen, however when the lesion has undergone torsion, this may prevent adequate visualization [7,10,12]. The absence of this classic finding (dominant feeding vessel) may lead to delay in diagnosis. In our patient, imaging did not reveal a vascular stalk and there was no enhancement with contrast, therefore a diagnosis of pulmonary sequestration was felt to be less likely than infection or neoplasm. Therefore, a high index of suspicion for pulmonary sequestration must be maintained during the evaluation of a posterior mediastinal mass, even when a dominant feeding vessel is not identified on cross-sectional imaging.

3. Conclusion

Clinical symptoms of a pulmonary sequestration that has undergone torsion are nonspecific and imaging may be suggestive of more ominous masses in the posterior mediastinum such as malignant tumors [13]. This may lead to unnecessary work-up and ultimately delay definitive treatment. Thus, clinicians should consider torsion of a pulmonary sequestration in their differential diagnosis for posterior mediastinal mass in pediatric patients, particularly in the setting of severe acute chest or flank pain with fever and elevated inflammatory markers. This rare entity should also be considered in the differential diagnosis of pediatric patients with unexplained chest or back pain, as our patient did not present with the more typical abdominal pain associated with this condition.

Conflict of interest statement
The authors have no conflicts of interest to disclose.

References