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Extralobar pulmonary sequestration presenting with torsion: a case report and review of literature

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Abstract Posterior mediastinal masses have widely variant presentations and a broad differential diagnosis. We describe an adolescent boy presenting with a posterior mediastinal mass, associated with abdominal pain and pleural effusion. Radiologic, operative, and pathologic findings are reviewed. This patient was found to have torsion of an extralobar pulmonary sequestration, of which only 2 other cases have been reported in the literature. Although radiologic findings of a systemic arterial supply to the mass may be absent, with patients presenting with this constellation of symptoms, a high index of suspicion of a sequestration should be maintained.

Posterior mediastinal masses have widely varying presentations—being symptomatic or asymptomatic—as well as a broad differential diagnosis, including tumors, infections, abscesses, aneurysms, pulmonary or enteric cysts (esophageal diverticula, tumors), and diseases of the thoracic spinal cord and vertebrae [1]. Congenital pulmonary malformations are rare, occurring in 1 per 8300 to 35,000 live births [2], and constitute a small minority of posterior mediastinal masses. We describe an unusual case of a 13-year-old boy presenting with a posterior mediastinal mass and abdominal pain as a result of torsion of a pulmonary sequestration.

1. Case report

A 13-year-old adolescent boy presented to the emergency department with a 2-day history of vomiting and severe, sharp abdominal pain. The patient's medical and family histories were unremarkable. Physical examination was notable for tachycardia, inspiratory pain, and guarding on abdominal examination. Over the next few days, his abdominal pain worsened, and vomiting continued without any other systemic signs.

Electrocardiogram and chest radiograph were interpreted as normal findings. Computed tomography (CT) enterography was performed to rule out inflammatory bowel disease. This CT incidentally revealed a well-defined posterior mediastinal soft tissue mass (1.7 × 2.4 × 2.9 cm) without calcification or feeding vessel, with characteristics

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suggestive of a possible neurogenic tumor. Chest CT, urinary vanillylmandelic acid, and homovanillic acid were obtained to rule out neuroblastoma. Further imaging revealed a mass that was adherent to the chest wall and did not move with respiration. Whole-body positron emission tomographic (PET) scan using 18-FDG (Fig. 1) showed increased uptake with a standardized uptake value (SUV) of 2.1 in the posterior mediastinal mass. Metaiodobenzylguanidine scan was obtained to further rule out neuroblastoma and was negative. A moderate left-sided pleural effusion developed, and the pleural fluid was aspirated. Cytology revealed no evidence of malignancy but revealed inflammatory cells. Peripheral blood studies were notable for elevated segmented neutrophils, monocytes, and eosinophils as well as elevated erythrocyte sedimentation rate and serum C-reactive protein, suggestive of an inflammatory state.

Thoracoscopy was performed, and at operation, the mass was densely adherent to the aortic wall, was dark and hemorrhagic in appearance, and arose from a stalk. No abscess was observed (Fig. 2). The mass was resected. Grossly,

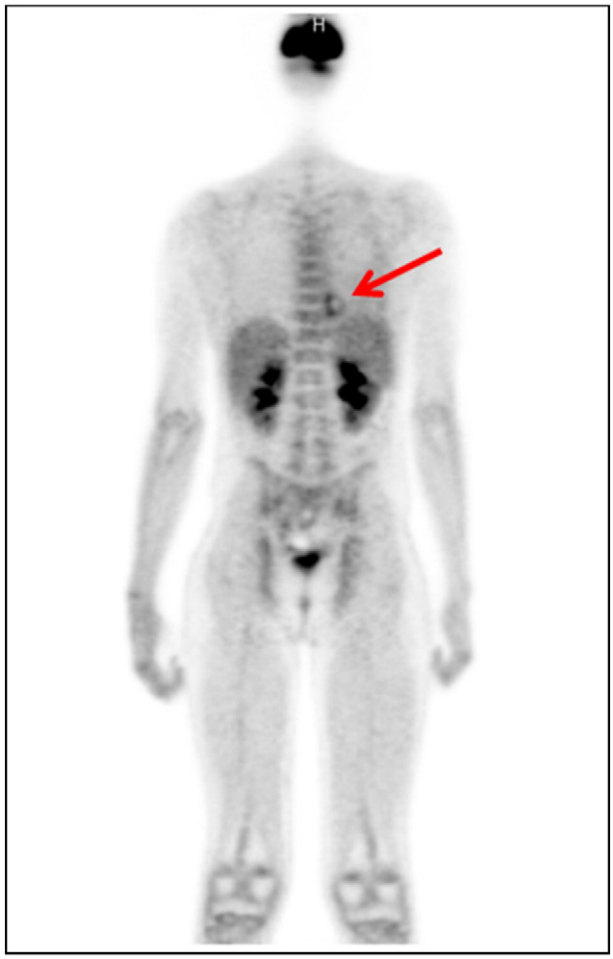

Fig. 1 PET scan, whole-body, increased uptake (SUV of 2.1) in the posterior mediastinal mass (arrow).

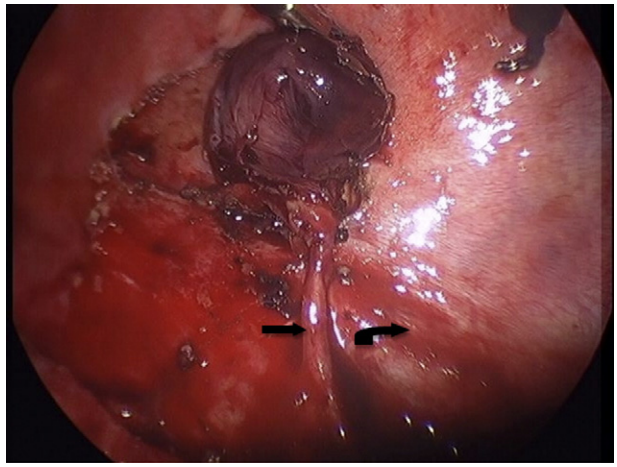

Fig. 2 Mass with unusual vascular stalk (arrow) continuing inferiorly below the diaphragm. Aorta indicated (curved arrow).

pathologic examination of the mass revealed a red-to-brown, well-circumscribed lobular mass with an attached vessel (0.9 cm). The cut surface of the specimen was hemorrhagic with cystic degeneration consistent with ischemia owing to torsion. Microscopically, the artery extended into diffusely hemorrhagic tissue, which had "ghost" outlines of alveoli. Adjacent to the vascular supply was bronchial epithelium surrounded by islands of cartilage. A single metaplastic bronchiole was identified. The reticulin stain highlighted the parenchymal architecture (Fig. 3). The final diagnosis was an infarcted extrapulmonary sequestration owing to torsion. Postoperative imaging showed normal pulmonary vascularity and cardiac silhouette. The patient was discharged home on postoperative day 2 in good condition. Follow-up of more than 15 months has shown no complications.

2. Discussion

In the pediatric population, the most common cause of posterior mediastinal mass is a neurogenic tumor, such as a neuroblastoma, ganglioneuroma, neurofibroma, or schwannoma [3]. As a result, rare entities such as pulmonary sequestration, with an estimated incidence of only 0.225% to 0.425%, are often not considered in the initial evaluation of these patients [4,5].

Pulmonary sequestrations are rare, with an estimated incidence of 0.15% to 1.7% of patients [5]. Less common than its intralobar counterpart, extralobar sequestration (ELS) is hypothesized to arise embryologically as an extra lung bud from the foregut [4] that migrates caudally with its parent structure. Eighty percent of ELS cases occur between the lower lobe and the diaphragm. Sixty percent of patients with ELS also have associated congenital abnormalities, such as congenital cystic adenomatoid malformation, congenital diaphragmatic hernia, lung and chest wall deformities, vertebral deformities, hindgut duplications,