CASE REPORT

An atypical presentation of pulmonary sequestration

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Pulmonary sequestration (PS) is a rare congenital malformation of the lower respiratory tract and is commonly complicated by recurrent infections and presents with respiratory failure. We report an atypical clinical presentation of postprandial abdominal pain and cramps in a patient with intralobar PS.

Key words: Abdominal angina, pulmonary sequestration, steal syndrome

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INTRODUCTION

Pulmonary sequestration (PS) is a nonfunctioning lung mass that lacks normal communication with the tracheobronchial tree and is comprised of a systemic arterial blood supply. Clinical presentation is most commonly as respiratory distress, however, can remain asymptomatic.

CASE REPORT

A 28-year-old nonsmoker male with no significant medical history presented with progressive worsening of postprandial epigastric pain for 2 months. There was no nausea, vomiting, and change in bowel movements and weight. He denied fevers, chills, cough, chest pain, or recent travel. His family history was unremarkable. On physical examination, he was alert and oriented, with normal vital signs. His SpO_2 was 97% whereas breathing ambient air. There was diffuse mild tenderness on abdominal examination, although no rebound and guarding noted. His white blood cell count was 6200/L; other blood cell counts, electrolytes, renal, and liver function tests were unremarkable. Computed tomography (CT) of the abdomen and pelvis showed no abnormality, however, revealed evidence of left lower lobe PS. Esophagogastroduodenoscopy was unremarkable. CT chest with angiography noted intralobar PS in the medial aspect of the left lower lobe of the lung supplied by the celiac artery [Figure 1a and b]. There were no clinical or radiographic signs of pulmonary infection, and the CT chest finding of PS were thought to be an incidental finding. At this point of time, patients were given the choice of being conservative and follow with clinical symptoms or proceed with diagnostic and therapeutic intervention. Subsequently, the patient underwent left PS resection through left thoracotomy with the biopsy confirmed the diagnosis [Figure 2a and b]. He was noted to have remarkable improvement of postprandial epigastric pain in 3 and 6 months of clinic visits.

DISCUSSION

PS is a nonfunctional pulmonary mass that does not communicate with the tracheobronchial tree, which is supplied by an anomalous systemic artery.[1] It is a rare condition, representing between 0.15% and 6.45% of all pulmonary malformations.[2] PS is stratified into two main categories: intralobar, as in our patient, and extralobar sequestration. Intralobar sequestration involves the location of the anomalous lung segment within a normal lobe, lacking its own visceral pleura. Extralobar sequestration demonstrates a segment that has its own visceral pleura, located outside the lung, and is commonly present in the left hemithorax.[1,2] Extralobar sequestration commonly presents during

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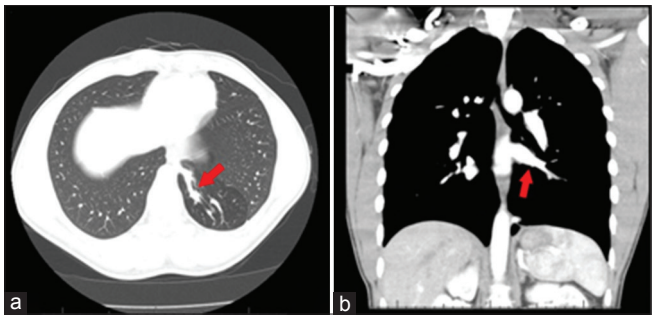

Figure 1: Computed tomography with contrast on axial (a) and coronal (b) view showing the intralobar pulmonary sequestration in the medial aspect of the left lower lobe of the lung, supplied by the celiac artery (red arrow)

infancy or childhood and intralobar sequestration manifests in adulthood, typically in the second decade of life.[3] Patients with PS have been reported to present with recurrent pulmonary bacterial infections, even fungal infections and tuberculosis.[2,4,5] Hemoptysis and hemothorax have been associated with PS, with no confirmed causal relationship.[6] PS rarely has been complicated with heart failure and ventricular tachycardia.[7] Recurrent abdominal pains in a patient with PS have been reported.[8] The unique aspect of our case involves the patient presenting with postprandial epigastric pain together with the celiac artery feeding the PS lobe, which up to best of our knowledge, has never been reported. We hypothesize the origin of postprandial pain as abdominal angina from steal syndrome.[9] Steal syndrome refers to a phenomenon where there are significant angina symptoms due to arterial insufficiency or reversal in blood flow to a particular organ system. In our case, the celiac artery is feeding the posterior basal segment of the left lower lobe leading to a deficient blood supply to the epigastric region resulting in postprandial epigastric pain, which is a very rare presentation of PS. This hypothesis has a similar overlap behind the subclavian-vertebral artery steal syndrome, a condition in which there is a reversal due to stenosis or occlusion of the proximal subclavian or innominate artery. Our hypothesis is further supported by the fact that the postprandial epigastric pain resolved after the patient underwent a resection of the PS involving the posterior segment of the left lower lobe of the lung. The most common diagnostic noninvasive imaging technique is CT with angiography for recognition of the vessel supplying the sequestered segment for surgical resection.

CONCLUSION

We recommend that PS should be considered in the diagnostic workup in patients that do not have a clear etiology for postprandial epigastric pain.

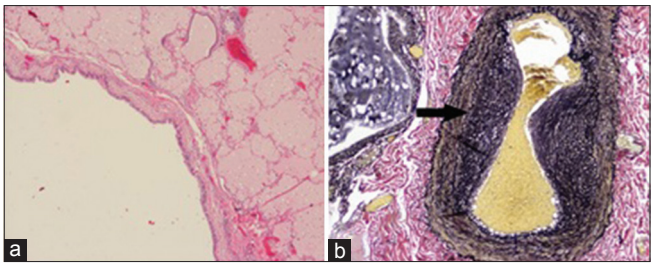

Figure 2: (a) Polycystic changes consistent with sequestered lung tissue (H and E). (b) Verhoeff-Van Gieson stain demonstrates a heavy concentration of elastic fibers (black arrow) representing a systemic artery, not a bronchial vessel

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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