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Case Report

Lingular intralobar pulmonary sequestration supplied by the left inferior phrenic artery arising from the left gastric artery – A case report and literature review ☆

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ABSTRACT

Pulmonary sequestration is characterized by a nonfunctional mass of lung tissue with an aberrant blood supply. Intralobar pulmonary sequestration (IPS) typically affects lower lung lobes and receives its blood supply from systemic arteries. Here, we present a unique case of a 51-year-old woman presented with recurrent nonmassive hemoptysis. Contrast-enhanced computed tomography angiography (CTA) of the chest showed uniform consolidation in the inferior lingular segment of the left upper lobe. Maximal intensity projection (MIP) and three-dimensional volume rendering (3D-VR) showed the affected area's blood supply from unusual arterial branches originating from the left inferior phrenic artery arising from the left gastric artery, consistent with IPS. A multidisciplinary approach utilized endovascular intervention (coil embolization) before successful surgical resection. Detecting IPS in unusual sites, like the lingular region, poses a diagnostic challenge. Clinicians and radiologists may not initially consider this diagnosis when evaluating patients with respiratory symptoms or incidental imaging findings. A comprehensive grasp of their anatomy and vascular variations is vital for precise diagnosis and effective treatment planning.

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Introduction

Pulmonary sequestration is an uncommon congenital anomaly defined by a nonfunctional mass of lung tissue that lacks regular communication with the tracheobronchial tree, obtaining its blood supply from the systemic circulation [1]. This condition primarily presents in 2 distinct forms: intralobar and extralobar. Intralobar pulmonary sequestration (IPS) is the more common form, accounting for about 75% of cases [2,3]. In the intralobar form, lung tissue is encapsulated within the pleura of a standard lung lobe, while extralobar pulmonary sequestration resides within its pleural enclosure, usually situated beyond the confined typical lung tissue, often between the diaphragm and lower lung lobes [3,4].

Although IPS primarily affects the lower lobes, especially the left, its occurrence in the lingula is exceptionally rare and not well-documented in medical literature. Typically, the systemic artery that supplies blood to IPS originates from the descending thoracic aorta or abdominal aorta, with less frequent contributions from the celiac, intercostal, or left gastric artery [5,6].

In the current IPS literature, there are very few reports of lingular IPS. To our knowledge, this is the first case where lingular IPS is supplied by a unique variation: the left inferior phrenic artery originating from the left gastric artery.

Case report

A 51-year-old female patient without any underlying disease presented to the internal medicine department due to recurrent episodes of nonmassive hemoptysis since 2020 with 20-30 cc per time of fresh bloody content. She denied fever, weight loss, history of contact with tuberculosis, and history of smoking. Her medical history, symptoms, laboratory findings, and clinical evaluation were thoroughly assessed. In the past 2 years, she was treated for bacterial pneumonia. Moreover, computer tomography of the chest and the bronchoscope was done and the bronchoalveolar lavage did not show any evidence of any infection. She had been lost to follow-up for 2 years and then visited our hospital due to recurrent nonmassive hemoptysis.

Initial chest radiography revealed irregular ground-glass opacities in the lower left lung area, causing a distinct silhouette effect with the left side of the heart border (Fig. 1). Consequently, the patient underwent contrast-enhanced computed tomography angiography (CTA) of the chest. This imaging revealed uniform consolidation in the inferior lingular segment of the left upper lobe. Detailed imaging techniques, such as maximal intensity projection (MIP) and three-dimensional volume rendering (3D-VR), vividly showed the affected area's blood supply from unusual arterial branches originating from the left gastric artery. Additionally, these vessels drained into the left superior pulmonary vein (Fig. 2). As a result, the imaging findings were consistent with IPS.

A multidisciplinary team carefully considered the case to determine the most suitable surgical approach. However, due to recurrent hemoptysis and the risk of bleeding during surgery, we chose endovascular intervention (coil embolization) to control bleeding and better understand vascular anatomy. The embolization procedure was performed via right femoral arterial access using a 5-French catheter. A celiac angiogram revealed the left inferior phrenic artery arising from the left gastric artery. This indicated pulmonary sequestration in the lingula segment, which was supplied by the left inferior phrenic artery and drained into the left superior pulmonary vein (Fig. 3).

Following precise navigation, we successfully catheterized the left inferior phrenic artery using a 2.4-French Maestro microcatheter. Subsequent coil embolization employed three 3×3.3 mm coils, along with a single 5×5.5 mm coil. A postembolization assessment conclusively demonstrated the occlusion of arterial flow (Fig. 3).

Three days later, the patient underwent surgical resection. During the surgery, we observed a well-defined infarction in the lingular segment, with a nonpulsatile feeding artery originating from the left inferior phrenic artery. We performed a lingular segmentectomy using video-assisted thoracoscopic surgery (VATS). The patient was discharged 4 days following the procedure, and no immediate complications were

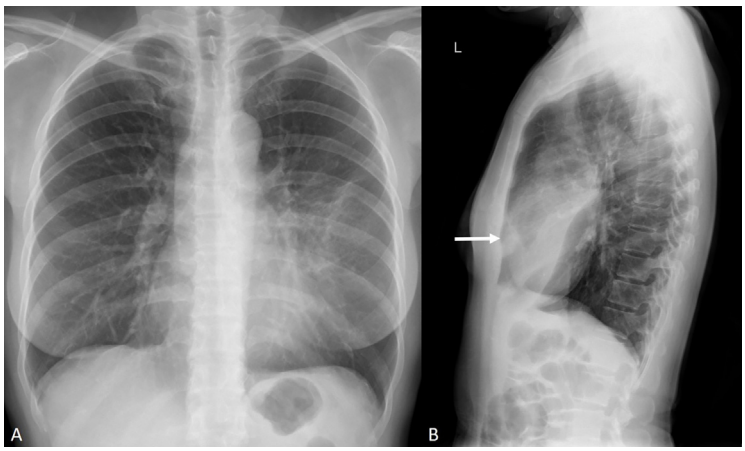

Fig. 1 – (A) Frontal and (B) lateral radiographs on the chest demonstrated focal ground-glass opacities with irregular margins in the anterior aspect of the lower left lung zone, causing a silhouette with the left side of the heart border.

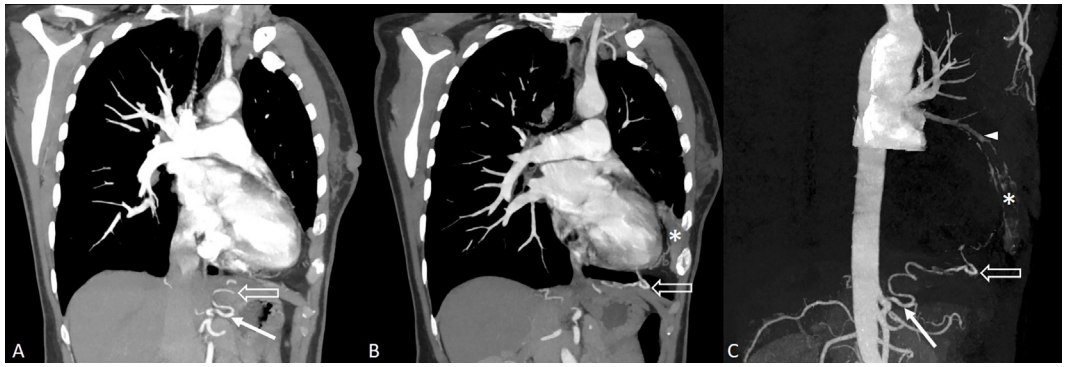

Fig. 2 – (A and B) Coronal oblique view contrast-enhanced computed tomography angiography (CTA) of the chest and (C) maximal intensity projection (MIP) with three-dimensional volume rendering (3D-VR) shows intralobar pulmonary sequestration in the inferior lingula segment of the left upper lobe (asterisk) which blood supply from a left inferior phrenic artery (open arrow) that arising from left gastric artery (arrow) and draining in to the left superior pulmonary vein (arrowhead).

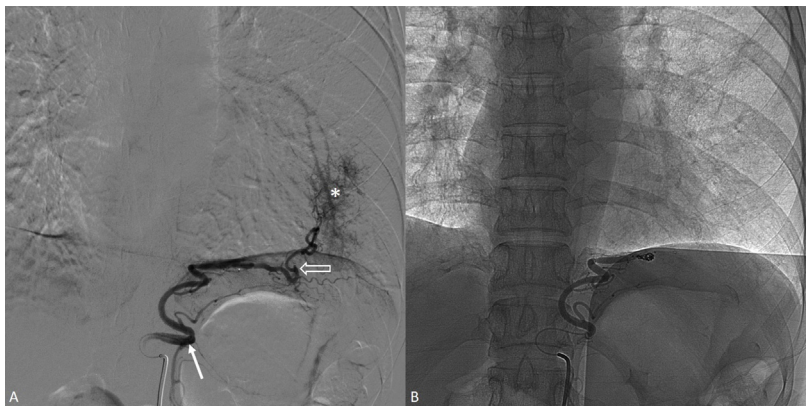

Fig. 3 – (A) Digital subtract angiogram showed the pulmonary sequestration (asterisk). The feeding artery from left inferior phrenic artery (open arrow) that arises from left gastric artery (arrow). After coil embolization (B) demonstrated the occlusion of arterial flow.

reported. A pathological examination conducted during the 6-week follow-up confirmed the presence of IPS, surrounded by diffuse alveolar hemorrhage and pulmonary interstitial fibrosis.

Having been monitored for 9 months after the surgery, the patient had not shown any symptoms of hemoptysis or surgical complication.

Discussion

Pulmonary sequestration, a rare congenital lower airway malformation, occurs when a part of the lung bud becomes separated from the developing bronchial tree, and forms its own blood supply from systemic arteries during early fetal development [7,8]. Several pathogenic mechanisms for IPS have been proposed, including the following. First, the lung bud arises before the development of the pleura [9,10]. Second, acquired origin from chronic pulmonary infection leads to the proliferation of aberrant arterial vessels [11,12].

IPS is mostly found in the left lower lobes (about 74%), less often in the right lower lobe (approximately 25%), and extremely rarely in the upper lobe, accounting for less than 0.5% of all pulmonary sequestrations [13].

Some studies have reported an unusual location of IPS in the lingular segment of the left upper lobe. In 1961, Wesley et al. reported the first case of a 62-year-old cadaveric woman with IPS in the superior lingular segment adhered to the pericardium, receiving its arterial supply from both the pericardiophrenic artery and a systemic vessel from the descending thoracic aorta [8]. Marwah et al. [14] also documented a case with a history of recurrent left-sided pneumonia and breathlessness on exertion had IPS located within the lingula, with arterial supply originating from the abdominal aorta. Furthermore, Wei et al. [13] conducted a retrospective analysis of a large IPS population in China, showing that IPS was primarily located in the posterior basal segment of the left lower lobe (approximately 66%) and the posterior basal segment of the right lower lobe (about 21%), which were consistent with prior studies. Notably, out of 2,625 cases in Wei et al.'s literature, only 2 cases (0.08%) of IPS were found in the lingular segment

of the left upper lobe, the former in the superior lingular lobe and the latter in the inferior lingular lobe [13].

Typically, IPS is supplied by systemic arteries from the descending thoracic aorta or abdominal aorta. However, literature on IPS arterial supply is limited, with only a few studies investigating alternative sources such as the celiac, intercostal, or left gastric artery. An even rarer occurrence is the involvement of the inferior phrenic artery, accounting for only 1.6%, as reported by Wei et al. [13].

The inferior phrenic artery, responsible for supplying the diaphragm, esophagus, stomach, liver, and adrenal gland, also exhibits rare variations. Aslaner [15] reported the rarest variation, which involves the left inferior phrenic artery arising from the left gastric artery. This condition occurs in about 0.1% of all cases, as seen in our case.

Clinical manifestations of IPS vary widely, with some individuals remaining asymptomatic while others experiencing symptoms, including chronic cough, recurrent respiratory infections, breathing difficulty, chest pain, and hemoptysis due to abnormal blood vessels [16].

Diagnosing IPS requires a comprehensive approach, including clinical assessment and imaging. Initial clues can be obtained from a chest radiograph, which may show unusual opacities and, in some instances, visible blood vessels (arteries and veins) near the affected area. However, a conclusive diagnosis typically depends on advanced imaging techniques like CT scans, MRI, or angiography, which provide essential insights into blood supply and assist in surgical planning if needed. CT scans can reveal variable features of IPS, including solid enhancing lesions, cystic lesions, emphysematous changes, cavitary lesions, or focal consolidation, as observed in our case. The key diagnostic feature of IPS on CT is identifying the systemic artery supply in the abnormal lung area [7].

The differential diagnosis of IPS depends on the clinical scenario and radiologic manifestations. Recurrent nonmassive hemoptysis is typically associated with infections, especially tuberculosis, in developing countries, and these were considered as potential differential diagnoses in our case. However, our patient did not present with fever, had no history of contact with tuberculosis, and the CT scan showed persistent consolidation surrounded by ground-glass opacities, without pulmonary nodules, tree-in-bud appearance, lung abscess, calcified granuloma, or pseudoaneurysm formation, which are typically observed in pulmonary infections [17].

Other noninfectious causes of hemoptysis, such as direct invasion of the lung and airway malignancies, were considered unlikely because there was no evidence of a lung or airway mass and no abnormal lymphadenopathy, which are typically seen in malignancies. Furthermore, congenital malformations, including pulmonary arteriovenous malformations (AVM), are also considered as differential diagnoses in patients presenting with hemoptysis. However, in our case, there was no evidence of an abnormal connection between the artery and vein or nidus, as seen in pulmonary AVM [14].

Surgery is the standard treatment for IPS, aiming to remove nonfunctional tissue and correct abnormal blood supply to prevent complications like infections or bleeding [1]. In cases of recurrent hemoptysis, a collaborative team of radiologists, interventionists, pulmonologists, and thoracic surgeons is crucial. This multidisciplinary team assesses the best treatment strategy, which may involve preoperative endovascular approaches to alleviate symptoms and minimize complications before surgical resection.

As of now, only a few reports have detailed the combined techniques of coil embolization before surgical resection in IPS cases. Some studies have explored embolization when surgery is contraindicated [18–20]. While embolization is generally safe and effective, data on its long-term success and complications are limited. In the medical literature, some experts recommend thoracoscopic surgery because it is less invasive, leads to less intraoperative bleeding, allows for faster postoperative recovery, and results in shorter hospital stays compared to traditional thoracotomy. In our case, we showcase a successful approach in which we combined both techniques for the best outcome.

Conclusion

Identifying IPS in unusual locations, such as the lingular region, presents a diagnostic challenge. Due to its rarity and unique characteristics, clinicians and radiologists may not initially consider this diagnosis when evaluating patients with respiratory symptoms or incidental imaging findings. Therefore, gaining a comprehensive understanding of the specific anatomical features and vascular variations associated with these anomalies is crucial for accurately diagnosing and developing effective treatment strategies.

Patient consent

Written informed consent was obtained from the patient's daughter.

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