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Case Report

Endovascular embolization for massive hemoptysis in intralobar pulmonary sequestration with celiac artery supply: A comprehensive case report☆,☆☆,★,★★,†,††

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ABSTRACT

Pulmonary sequestration (PS) is a rare congenital anomaly characterized by noncommunicative lung tissue supplied by an abnormal systemic vessel. We present a case of a 30-year-old male with intralobar PS, receiving arterial supply from the celiac artery, manifesting as massive hemoptysis. After urgent stabilization, endovascular embolization using polyvinyl alcohol particles was successfully employed. The patient's symptoms resolved, and follow-up confirmed satisfactory recovery. Our case underscores the diverse arterial origins of PS and the efficacy of endovascular embolization as a minimally invasive treatment. The complexity of PS, its diagnostic imaging, and alternative therapeutic options are discussed, emphasizing tailored approaches for optimal outcomes in managing this uncommon congenital anomaly.

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Introduction

Pulmonary sequestration (PS) is an uncommon congenital anomaly affecting the respiratory system, characterized by the presence of a nonfunctional segment of lung tissue lacking normal communication with the tracheobronchial tree. It receives its blood supply from an abnormal vessel originating from the systemic circulation [1]. Its occurrence is rare, ranging from 0.15% to 1.8% in the general population [2]. Classified based on its location relative to the diaphragm, PS is categorized as supra-diaphragmatic, infra-diaphragmatic, or, less commonly, intra-diaphragmatic. Supra-diaphragmatic PS is further divided into intra-lobar and extra-lobar PS [3]. The normal lung's visceral pleura envelops the intra-lobar form, while the extra-lobar type is not and may be situated either within the thoracic cavity (intrathoracic) or externally (ectopic) [3]. The intra-lobar variant is the most prevalent, accounting for approximately 75% of cases [4]. Typically diagnosed in young adults experiencing recurrent pneumonia, the intra-lobar type may occasionally present with hemoptysis, making early recognition challenging [5].

The anomalous artery, arising from the systemic circulation and supplying blood to the PS, may originate from the descending thoracic aorta or abdominal aorta, with less frequent contributions from the intercostal artery, left gastric artery, and renal artery [6]. Surgical resection is considered the standard treatment for PS [7]. However, recent research has demonstrated that endovascular embolization is an effective therapeutic alternative for symptomatic PS. This minimally invasive approach has been associated with lower morbidity and favorable outcomes [7]. There are documented cases in the literature where extra-lobar PS has been successfully treated with endovascular embolization, and although rare, a limited number of cases involving intra-lobar PS treated with this method have also been reported [5].

We present a case of a 30-year-old male with chief complaints of massive hemoptysis, chest pain, and shortness of breath which was later diagnosed to be having intralobar pulmonary sequestration with aberrant arterial supply from celiac artery managed later with endovascular embolization.

Case details

A 30-year-old male presented to the emergency department experiencing multiple episodes of significant hemoptysis over the past 5 hours, accompanied by mild left chest pain and shortness of breath. Notably, there was no history of cough or fever. While the patient had a prior history of occasional hemoptysis, the current episode was severe, impacting his ability to perform daily activities. Additionally, he reported a history of recurrent chest infections treated with antibiotics. At the age of 5, he had been diagnosed with pulmonary tuberculosis and had completed a 6-month course of antitubercular therapy, subsequently testing seronegative. There were no reported incidents of significant trauma, and the patient had no smoking, alcohol, or illicit drug use history. No similar medical condition was noted in any family member.

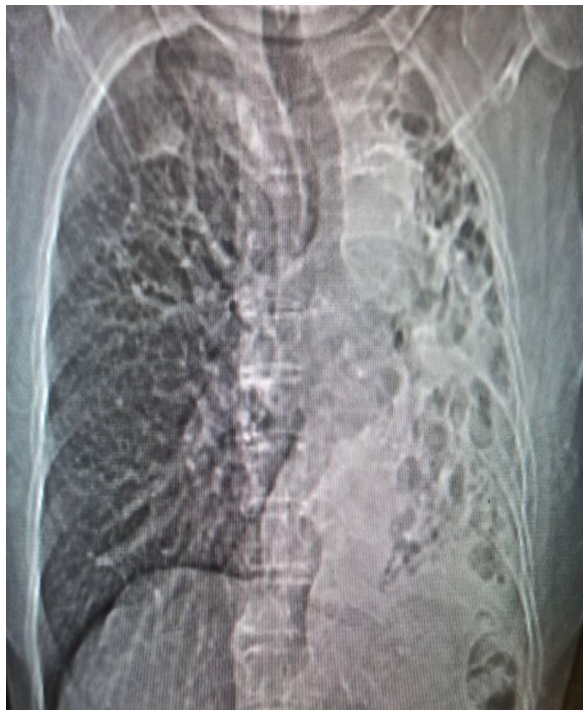

Fig. 1 – Chest X-ray Posteroanterior view showing the collapse of the left lung with ipsilateral mediastinal shift and multiple lucencies towards the periphery.

Upon physical examination, the patient presented with hypotension (100/70 mmHg), a thready and rapid pulse (120/min), normal temperature (38°C), and a respiratory rate of 19/min. Urgent emergency management was initiated. Systemic examination revealed absent air entry on the left side, with all other examination findings within normal limits. The biochemical analysis indicated normal results, except for a decreased hemoglobin level, while the coagulation profile remained within the normal range (Table 1). A Chest X-ray PA view revealed a collapsed left lung with ipsilateral tracheal deviation, along with multiple variable-sized lucencies at the periphery of the left lung field (Fig. 1). Subsequent contrast-enhanced CT chest imaging disclosed a collapsed left lung with multiple cystic and varicoid bronchiectasis, accompanied by ipsilateral mediastinal shift and hyperinflation of the contralateral lung (Figs. 2A-C). No any imaging sign of active tuberculosis was seen. Reconstruction and volume-rendered images delineated the vascular supply to the collapsed left lung originating from a branch of the celiac artery (Figs. 3A and B).

Following hemodynamic stabilization, the patient was referred for radio intervention for further assessment and management. Access was gained through the right femoral artery using a 5 Fr. vascular sheath. Fluoroscopic angiography was conducted through the bronchial artery, revealing no aneurysm or active extravasation. Additionally, no other arterial supply from thoracic aorta branches was observed. Contrast angiography via the celiac artery displayed tortuously dilated blood vessels supplying the left lung field, ac-

Table 1 – Laboratory results of the patient.

Investigation	Result	Reference range
Hemoglobin	9.5	11.5-15 mg/dL
Leukocyte count	11000	4000-11,000
Platelet count	3.2 lakh/cumm	1.5-4.5 lakh/cumm
Eosinophil	1	1%-6%
Prothrombin Time	12	10-13 sec
Erythrocyte Sedimentation Rate (ESR)	18mm/Hr	</=15 mm/Hr
Bleeding Time	6minutes	2-7 minutes

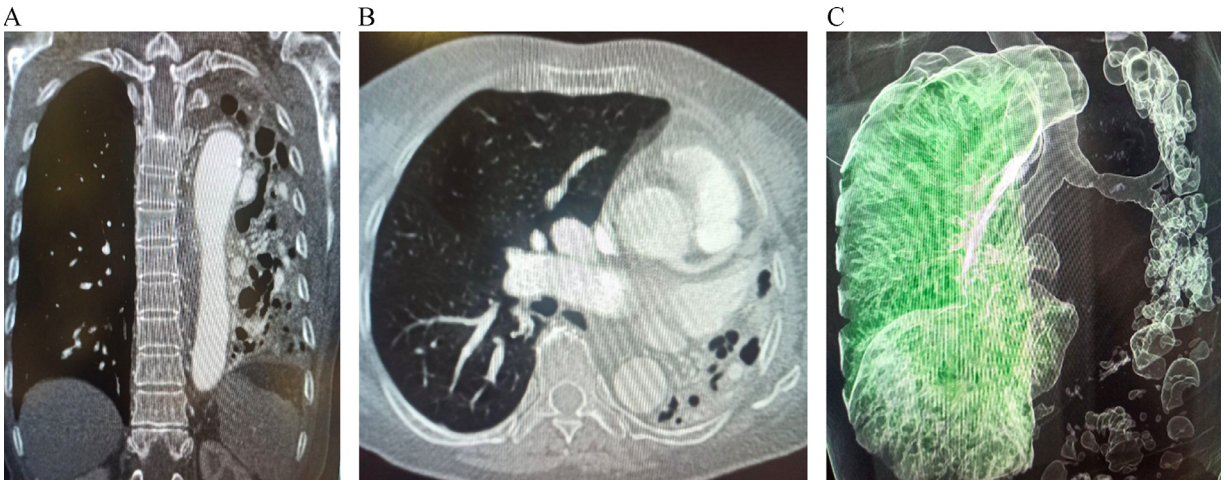

Fig. 2 – A. Coronal Contrast-enhanced CT chest showing cicatrization collapse of left lung with tubular and varicoid bronchiectatic changes in the periphery. B. Axial Contrast-enhanced CT chest showing the cicatrization collapse of the left lung with ipsilateral mediastinal shift and bronchiectatic changes in the periphery. C. 3D Volume rendered reconstruction image of the lung showing a collapsed left lung with multiple cystic lesions in the surroundings.

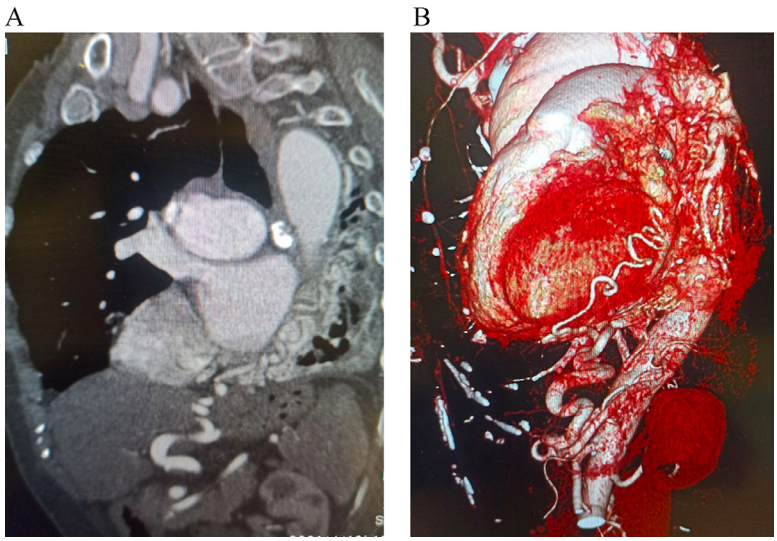

Fig. 3 – A. Sagittal reconstruction image of the chest showing the aberrant arterial supply to the lower segment of the left lung via the branch from the celiac artery. B. 3D volume Rendered reconstruction image showing the origin of the aberrant arterial supply of the lower segment of the left lung via the celiac arterial branch.

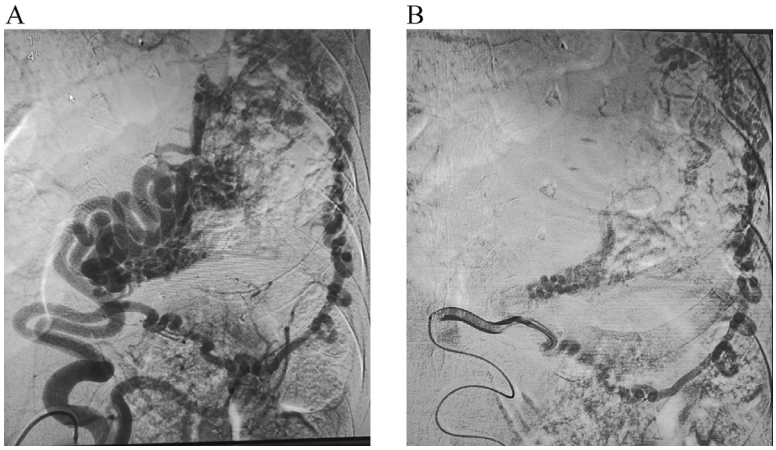

Fig. 4 – A. Fluoroscopy-guided angiography image with the tip of the catheter in the celiac artery showing multiple dilated tortuous vessels supplying the lower segment of the left lung. B. The angiographic image, guided by fluoroscopy and taken post-embolization, revealed the absence of opacification in the feeder vessels originating from the celiac artery.

companied by noted blushing. Embolization of the aberrant vessels was carried out using polyvinyl alcohol particles (250-350 μm), intermittently passing the particles through the aberrant vessels while checking vessel patency in between. Post-embolization angiography demonstrated no opacification of the dilated aberrant vessels, resulting in the resolution of hemoptysis (Fig. 4B). The patient was admitted for observation, and upon improvement in his physical condition, he was discharged after a brief hospital stay of four days. Following the procedure, we commenced regular vital sign monitoring, inspected the access site, and implemented pain management with analgesics and antitussive therapy as a preventative measure against rebleeding. Subsequent follow-up assessments indicated his satisfactory recovery. There were no observed instances of tuberculosis reactivation following the procedure. Despite being advised to undergo a repeat CT scan, he declined due to financial limitations.

Discussion

Pulmonary sequestration (PS) was initially reported in 1777, with the term 'Sequestration' introduced by Pryce in 1946, derived from the Latin verb "sequestare," meaning "to separate" [8]. Categorized based on its relation to the pleura, PS comprises 2 types: (a) Intra-lobar PS (ILS) and (b) Extra-lobar PS (ELS). ILS, the more prevalent type, involves dysplastic parenchyma within normal lung tissue, typically affecting the lower lung lobes, particularly on the left side [9]. Conversely, ELS, less common, possesses its pulmonary investment [9]. Venous drainage for most ILS occurs through pulmonary veins, while most ELS drain into the azygos or hemiazygos vein or the inferior vena cava [10].

In 1946, Pryce classified ILS into 3 types based on the aberrant artery supply (Table 2), later recognizing that Type 1 ILS represented an anomalous systemic arterial supply to the lung [11]. Savic et al. reported that 74% of ILS cases have aberrant arteries branching from the descending thoracic aorta, with only 1% branching from the celiac artery, as seen in our case [10]. The embryogenesis of ILS remains unclear, with a widely accepted theory suggesting disturbances in embryological development leading to a nonfunctioning lung segment without communication with the tracheobronchial tree but supplied by anomalous systemic blood vessels [12]. Chronic pulmonary infection-induced aberrant arterial vessel proliferation is also a contributing factor [13].

Table 2 – Pryce classification of ILS (Kanno-7).

Type 1	The aberrant artery flows through part of the normal lung but not through the sequestration.
Type 2	The aberrant artery flows through both the normal lung and sequestration.
Type 3	The aberrant artery flows only to the sequestration.

Patients with extra-lobar PS may exhibit symptoms in the first weeks of life due to a left-to-right shunt, whereas those with intra-lobar PS are frequently diagnosed in young adults [5]. Intra-lobar PS may remain asymptomatic or present with chronic cough, fever, pneumonia, chest pain, and, in severe cases, life-threatening complications such as massive hemoptysis and superimposed fungal infections like aspergillosis [14]. Increased capillary pressure secondary to elevated systemic pressures in feeding arteries is implicated in the occurrence of massive hemoptysis [15]. Extra-lobar PS is more often associated with congenital anomalies, with 50%-60% of ILS patients having associated congenital anomalies, most commonly congenital diaphragmatic hernia [10]. Our case exhibited repeated occurrences of hemoptysis, along with episodes of chest pain and shortness of breath, and a prior history marked by recurrent infections.

The diagnosis of pulmonary sequestration relies on imaging, with chest radiographs offering initial clues and advanced

tools such as CT scans, MRI, or angiography providing definitive information on blood supply and aiding in surgical planning if necessary. For ILS, CT scans may display solid enhancing, cystic, emphysematous, cavitary lesions, or focal consolidation [16]. CT angiography stands as the gold standard for diagnosis, showcasing abnormal arterial vessels and pathologic venous drainage accurately [17]. While an initial X-ray was performed, the definitive diagnostic tool for our case was a contrast-enhanced CT of the chest, revealing the aberrant arterial supply to the collapsed left lung through the celiac artery, which was further supported by Fluoroscopic angiography. Cicatrization collapse of the left lung with bronchiectatic changes were likely sequelae due to post-tuberculosis and recurrent childhood infection.

Although surgical resection traditionally remains the treatment of choice for ILS, even in asymptomatic cases, recent considerations include endovascular embolization and coiling as alternative approaches. Embolization occludes arterial supply, leading to sequestration regression via infarction, necrosis, and fibrosis [18,19]. Various embolic agents, including polyvinyl alcohol particles, micro-coils, n-butyl cyanoacrylate glue, Amplatzer occlusion devices, alcohol, and gelatin, have been employed [20]. We employed polyvinyl alcohol particles sized between 250-350 μm for the permanent embolization of feeder vessels, yielding a satisfactory outcome.

While embolization complications may include fever, access site thrombosis, transient limb ischemia, and non-target embolization, sequestration recurrence post-embolization is possible due to shunt formation, incomplete closure, and embolic agent displacement [19]. None of those complications was seen in our study.

Conclusion

This case report highlights the successful management of intralobar pulmonary sequestration, emphasizing the pivotal role of endovascular embolization in addressing massive hemoptysis. The aberrant arterial supply from the celiac artery further accentuates the complexity of this rare congenital anomaly, highlighting the significance of tailored therapeutic approaches for optimal patient outcomes.

Author contributions

Dinesh Chataut: Conceptualization, as mentor and reviewer for this case report and for data interpretation.

Shailendra Katwal: Contributed in conceptualization and reviewer.

Sundar Suwal: Contributed in performing literature review and editing.

Ajit Thapa: Contributed in performing literature review and editing.

Amrit Bhusal: Contributed in writing the paper and reviewer for this case.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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