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Case Report

Intralobar Pulmonary Sequestration in a 53-Year-Old Woman: An Unusual Late Presentation of a Congenital Lung Malformation

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Abstract

Pulmonary sequestration (PS) is a rare congenital bronchopulmonary malformation characterized by nonfunctional lung tissue lacking communication with the tracheobronchial tree and a receiving systemic arterial supply. Intralobar sequestration (ILS), the most common subtype, is usually diagnosed during childhood or early adulthood and is rarely identified after the age of 50. ILS usually manifests with hemoptysis or recurrent pulmonary infection. We report the case of a 53-year-old woman with a history of rheumatic mitral regurgitation and rheumatoid arthritis treated with methotrexate and corticosteroids, who presented with mild exertional dyspnea and constitutional deterioration. Pulmonary function tests showed preserved ventilatory function and normal diffusing capacity, with no exercise-induced desaturation. Chest computed tomography revealed cystic lesions in the left lower lobe supplied by an aberrant artery arising from the descending thoracic aorta, consistent with intralobar pulmonary sequestration. This case highlights an unusual late diagnosis of ILS in a patient with autoimmune disease and emphasizes the diagnostic value of contrast-enhanced computed tomography as well as the importance of individualized therapeutic decision-making.

Keywords: Pulmonary sequestration, Intralobar sequestration, Congenital lung malformation, Computed tomography angiography.

INTRODUCTION

Pulmonary sequestration (PS) is an uncommon congenital pulmonary malformation defined by a nonfunctional portion of lung parenchyma that lacks communication with the tracheobronchial tree and derives its arterial blood supply from the systemic circulation rather than the pulmonary artery [1, 2]. It accounts for approximately 0.15–6.4% of all congenital pulmonary malformations [1, 3].

Two anatomical types are recognized: intralobar sequestration (ILS) and extra lobar sequestration (ELS). ILS represents approximately 75–85% of cases and shares the visceral pleura of the adjacent normal lung, while venous drainage usually occurs through the pulmonary veins [2, 4]. The left lower lobe, particularly its posterior basal segment, is the most common location [3, 5].

Unlike ELS, which is often diagnosed during infancy, ILS tends to present later in life, frequently with recurrent pulmonary infections, hemoptysis, chronic cough, or exertional dyspnea [3, 6]. However, asymptomatic incidental diagnoses in adulthood are increasingly reported due to the widespread use of thoracic computed tomography [4, 7].

We report an unusual case of intralobar pulmonary sequestration diagnosed in a 53-year-old woman with rheumatoid arthritis and valvular heart disease, presenting with mild dyspnea and constitutional symptoms.

CASE PRESENTATION

A 53-year-old female patient was referred for respiratory evaluation because of progressive exertional dyspnea.

Her medical history included rheumatic valvular cardiopathy with mitral regurgitation, treated with digoxin (0.25 mg/day), acenocoumarol (4mg/day), and bisoprolol (5mg/day). She was also followed for

rheumatoid arthritis treated with deflazacort (5 mg/day) and methotrexate (2.5 mg three times a week).

She reported mild exertional dyspnea corresponding to grade 1 on the modified Medical Research Council (mMRC) dyspnea scale, without cough, sputum production, hemoptysis, fever, or chest pain. Symptoms evolved in a context of constitutional syndrome with general deterioration. She also described worsening joint pain related to active rheumatoid arthritis.

On physical examination, oxygen saturation was 96% on room air. She was eupneic at rest, with blood pressure of 118/65 mmHg. Pulmonary auscultation revealed no crackles or wheezing.

Pulmonary function testing showed no restrictive ventilatory defect, with preserved lung volumes and normal diffusing capacity for carbon monoxide (DLCO). A six-minute walk test showed no exercise limitation or oxygen desaturation.

Chest computed tomography demonstrated a heterogeneous oblong parenchymal mass in the basal segment of the left lower lobe, containing a few internal cystic areas. No communication with the tracheobronchial tree is identified. The lesion is embedded within the adjacent lung parenchyma and demonstrates a peripheral incomplete pleural fold without a separate pleural investment. (Fig 1;2)

Contrast-enhanced CT demonstrates an aberrant systemic feeding artery arising directly from the descending thoracic aorta at the D8 vertebral level (Fig 3), measuring approximately 6 mm in diameter and coursing toward and supplying the lesion. (Fig 4;5)

Venous drainage occurs through the left inferior pulmonary vein, with no evidence of anomalous systemic venous drainage.

Taken together, these findings are consistent with intralobar pulmonary sequestration. No pleural effusion or significant mediastinal or hilar lymphadenopathy is identified.

Given the absence of infectious complications, hemoptysis, or significant functional impairment, a multidisciplinary discussion was conducted, and conservative management with radiological surveillance was proposed.

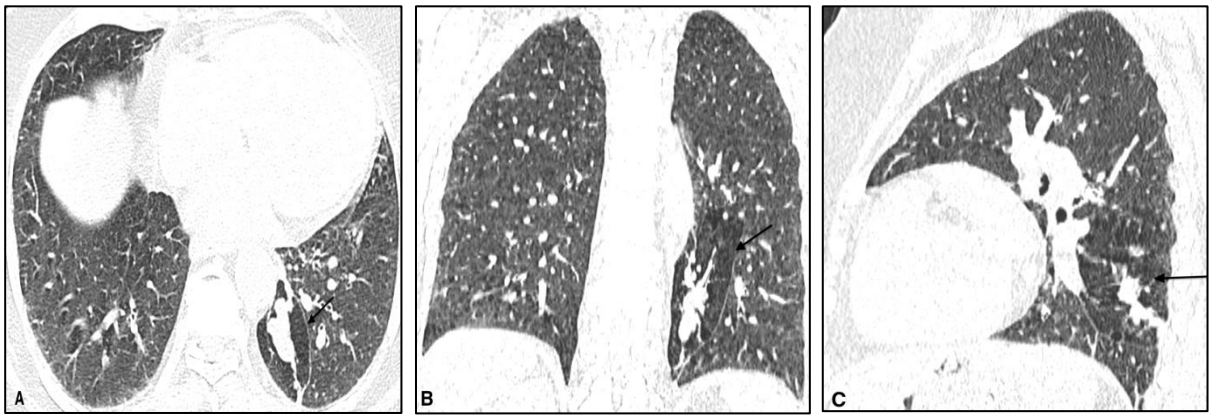

Fig. 1: Chest CT images (lung window) in axial (A), coronal (B), and sagittal (C) planes show a cystic parenchymal lesion in the lateral basal segment of the left lower lobe (Arrow)

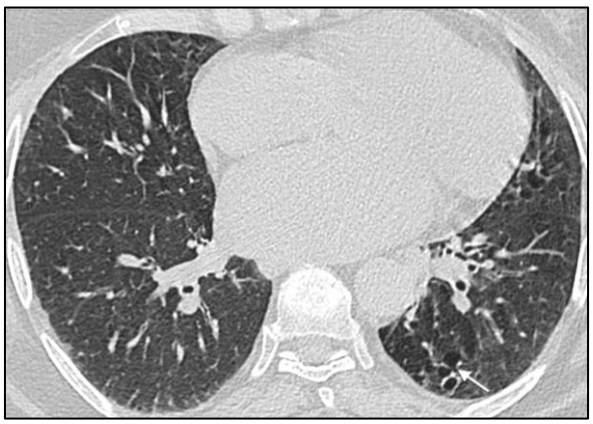

Fig. 2: Axial chest CT images (lung window) demonstrate multiple intraparenchymal cystic lesions within the sequestration (Arrow)

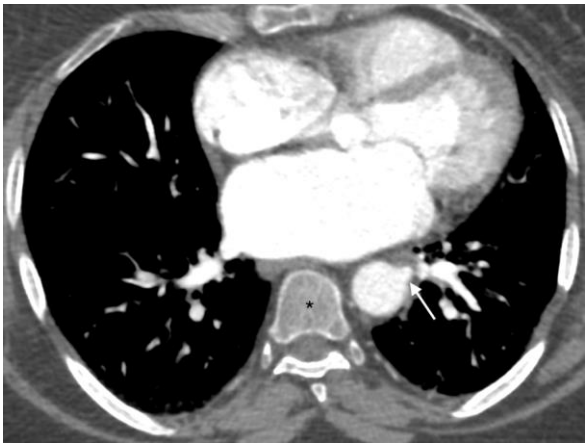

Fig. 3: Axial contrast-enhanced chest CT (mediastinal window, arterial phase) shows an aberrant systemic artery arising directly from the descending thoracic Aorta (arrow), at the T8 vertebral level (asterisk)

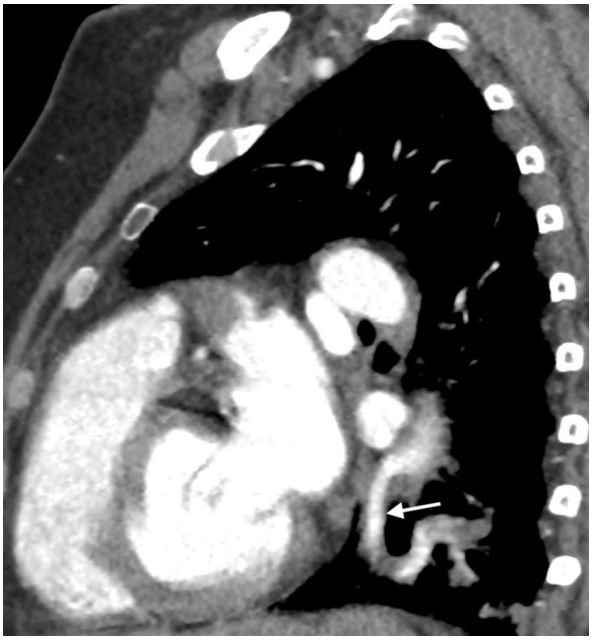

Fig. 4: Sagittal contrast-enhanced chest CT (mediastinal window, arterial phase) shows an aberrant systemic artery arising from the descending thoracic Aorta, consistent with intralobar pulmonary sequestration

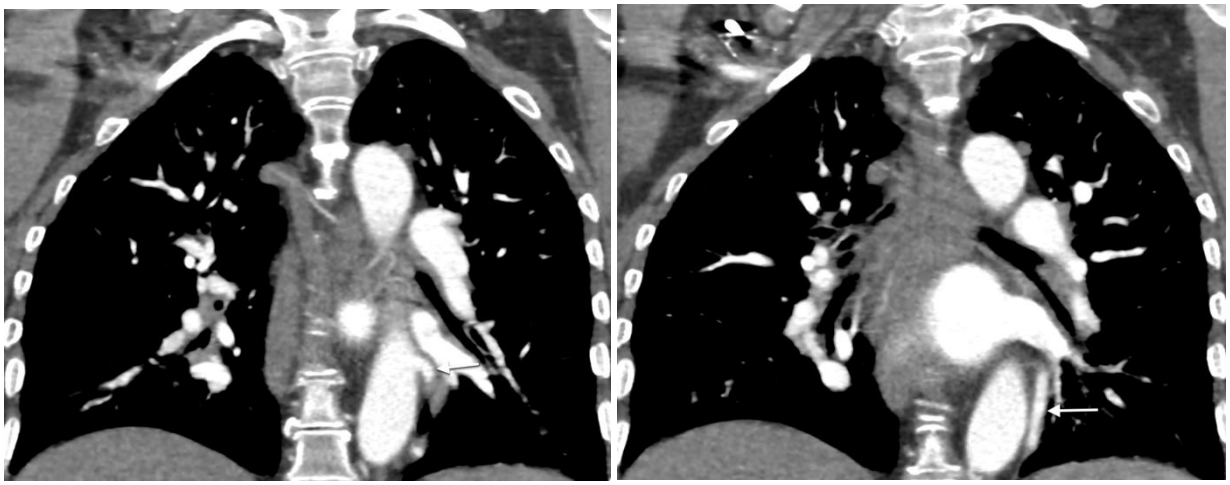

Fig. 5: Coronal contrast-enhanced chest CT (mediastinal window, arterial phase) shows an aberrant systemic artery arising from the descending thoracic Aorta, consistent with intralobar pulmonary sequestration

DISCUSSION

Pulmonary sequestration remains a rare congenital anomaly, although its real prevalence is probably underestimated because many cases remain asymptomatic until adulthood [1, 4]. Adult diagnosis after the age of 50 remains exceptional, making our case noteworthy [3, 6].

The embryological mechanism remains debated. The most widely accepted theory suggests the formation of an accessory supernumerary lung bud during early embryogenesis, which subsequently acquires systemic arterial supply [2, 5]. In ILS, this occurs before pleural development, resulting in incorporation within the normal lung pleura [2, 4].

ILS classically involves the lower lobes, predominantly the left lower lobe, as observed in our patient [3, 4]. The arterial supply usually arises from the descending thoracic aorta, which was also demonstrated in this case [4, 7].

Clinical presentation in adults is highly variable. Most symptomatic patients present with recurrent localized infections, productive cough, hemoptysis, or chest pain [4, 6]. Interestingly, our patient had none of these classical manifestations. Dyspnea was mild and likely multifactorial, potentially related to both underlying cardiac disease and constitutional symptoms.

The coexistence of rheumatoid arthritis added diagnostic complexity. Pulmonary manifestations of rheumatoid arthritis, including interstitial lung disease, bronchiolitis, rheumatoid nodules, and methotrexate-induced pneumonitis, may mimic or obscure congenital pulmonary abnormalities. Differential diagnoses therefore included rheumatoid arthritis-associated interstitial lung disease, methotrexate-induced pneumonitis, chronic infection, cystic bronchiectasis, and malignancy. In this setting, chest CT played a decisive diagnostic role.

Computed tomography angiography (CTA) is currently considered the diagnostic gold standard and has largely replaced conventional angiography because of its non-invasive nature and its ability to simultaneously assess both the pulmonary parenchyma and the anomalous vascular supply [1–4]. The main diagnostic objectives of CT are to identify the sequestered lung tissue and to demonstrate its aberrant systemic arterial supply, which remains the hallmark of diagnosis [1, 3]. Radiologically, pulmonary sequestration is classified into intralobar sequestration (ILS) and extralobar sequestration (ELS). Both share the hallmark feature of anomalous systemic arterial supply but differ significantly in their anatomical relationships, venous drainage, and imaging appearance [1–4].

Intralobar sequestration is the most common form, accounting for approximately 75–85% of pulmonary sequestrations, and is typically located within the normal visceral pleura of the adjacent lung, most commonly in the posterior basal segment of the left lower lobe [1–3]. On chest CT, ILS usually appears as an intraparenchymal lesion without its own pleural covering, often embedded within normal lung tissue [2, 3].

The key diagnostic feature is the identification of a feeding systemic artery, usually arising from the descending thoracic aorta, but occasionally from the abdominal aorta, celiac trunk, intercostal, gastric, renal, or subclavian arteries [1, 3, 8]. Venous drainage typically occurs through the pulmonary veins into the left atrium, which helps distinguish ILS from ELS [2, 3].

Extralobar sequestration is less common (15–25% of cases) and differs by having its own pleural covering and systemic venous drainage, usually into the azygos system, inferior vena cava, or portal vein [1–3]. On CT, it typically appears as a well-circumscribed soft-tissue mass adjacent to, but separate from, the normal lung parenchyma, most often located near the diaphragm [2, 3]. Rare infradiaphragmatic forms may mimic retroperitoneal or adrenal masses [1, 3].

Management remains controversial in asymptomatic adults. Surgical resection is generally recommended in symptomatic patients to prevent recurrent infection and potentially life-threatening hemoptysis [6, 8]. Video-assisted thoracoscopic surgery (VATS) has become the preferred surgical approach due to reduced morbidity [8].

Endovascular embolization is an alternative or adjunctive treatment for pulmonary sequestration, consisting of occlusion of the aberrant systemic feeding artery to reduce blood flow to the sequestered lung tissue [1–3]. It may be used as a definitive treatment in selected non-surgical candidates or as a preoperative procedure to reduce intraoperative bleeding risk [2, 3, 8]. The procedure is performed under angiographic guidance using coils, vascular plugs, glue, or other embolic agents [3, 8]. Its main advantages are its minimally invasive nature and its ability to decrease hemorrhagic risk. However, incomplete occlusion, collateral vessel recruitment, and symptom recurrence remain potential limitations, making patient selection and multidisciplinary evaluation essential [6, 8].

In our patient, conservative management was chosen because of minimal respiratory symptoms, normal functional evaluation, and significant comorbidities, particularly anticoagulation and valvular heart disease. Regular clinical and radiological follow-up was planned to monitor disease progression and detect potential complications that might require surgical or endovascular intervention.

CONCLUSION

Intralobar pulmonary sequestration is a rare congenital pulmonary malformation that may remain undiagnosed until late adulthood. Its diagnosis should be considered in adults with atypical cystic lower lobe lesions, even in the absence of recurrent infection. Contrast-enhanced CT angiography remains essential for diagnosis by identifying the aberrant systemic arterial supply. Management should be individualized according to symptoms, complications, and comorbidities. This case illustrates the importance of considering congenital pulmonary anomalies in the differential diagnosis of respiratory symptoms in patients with complex multisystem diseases.

Consent: Written informed consent was obtained from the patient for publication of this case report.

Conflicts of Interest: The authors declare no conflicts of interest.

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