Respir Case Rep 2026;15(2):99-103 DOI: 10.5505/respircase.2026.83798

Anomalous Systemic Arterial Supply to the Left Lower Lobe: Evaluation with Multimodality Imaging and Conservative Management

Sol Akciğer Alt Lobun Anormal Sistemik Arteriyel Beslenmesi: Multimodal Görüntüleme Temelli Değerlendirme ve Konservatif Tedavi Yaklaşımı

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Abstract

Anomalous systemic arterial supply to the left lower lobe is a rare congenital pulmonary vascular anomaly in which normally developed lung parenchyma with intact bronchial communication receives blood directly from a systemic artery rather than from the pulmonary arterial circulation. We report the case of a 35-year-old man who presented with mild hemoptysis and palpitations. CT and MR angiography revealed a 10-mm aberrant systemic artery arising from the descending thoracic aorta and supplying the basal segments of the left lower lobe, with normal pulmonary venous drainage and no dysplastic parenchyma. In the absence of pulmonary hypertension or a significant shunt, conservative management with follow-up was chosen. During one year of follow-up, the patient remained asymptomatic without radiologic progression. This case underscores the critical role of multidetector CT and MR angiography in accurately differentiating this entity from pulmonary sequestration and highlights that noninvasive diagnosis can support safe conservative management in appropriately selected patients.

Keywords: *Anomalous Systemic Arterial Supply, Pulmonary Sequestration, CT Angiography.*

Öz

Sol akciğer alt lobunun anormal sistemik arteriyel beslenmesi, bronşiyal bağlantısı korunmuş ve normal gelişim gösteren akciğer parankiminin pulmoner arter yerine doğrudan sistemik bir arterden kan aldığı, nadir görülen konjenital bir pulmoner vasküler anomalidir. Bu makalede, hafif hemoptizi ve çarpıntı yakınmaları ile başvuran 35 yaşındaki bir erkek olgu sunulmaktadır. Bilgisayarlı tomografi ve manyetik rezonans anjiyografi incelemeleri, inen torasik aortadan köken alan ve sol alt lobun bazal segmentlerini besleyen 10 mm çapında aberan bir sistemik arter ortaya koymuş; pulmoner venöz drenajın normal olduğu ve parankimde herhangi bir displazi bulunmadığı gösterilmiştir. Pulmoner hipertansiyon veya belirgin bir şant akımının saptanmaması nedeni ile konservatif izlem tercih edilmiştir. Bir yıllık takip süresince hasta asemptomatik seyretmiş ve radyolojik olarak progresyon izlenmemiştir. Bu olgu, multidetektör BT ve MR anjiyografinin söz konusu anomalinin pulmoner sekestrasyondan doğru şekilde ayırt edilmesindeki kritik önemini vurgulamakta; noninvazif tanının uygun seçilmiş hastalarda güvenli konservatif yönetimi mümkün kıldığını göstermektedir.

Anahtar Kelimeler: *Anormal Sistemik Arteriyel Beslenme, Pulmoner Sekestrasyon, BT Anjiyografi.*

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Submitted (Başvuru tarihi): 17.11.2025 ***Accepted (Kabul tarihi):*** 11.02.2026

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Anomalous systemic arterial supply to the left lower lobe is a congenital pulmonary vascular anomaly in which normally developed lung parenchyma with preserved bronchial communication receives high-pressure systemic blood flow directly from a systemic artery—most commonly the descending thoracic aorta—rather than from the pulmonary arterial circulation, while venous drainage usually occurs through the normal pulmonary veins (1). It is thought to arise from the persistence of primitive systemic-pulmonary arterial connections that fail to regress during the dual-phase vascular development of the fetal lung (2).

This anomaly was first described by Pryce in 1946 and has since been referred to in the literature by various terms, including anomalous systemic arterial supply to the lung, isolated systemic arterialization, and the nonsequestrated type (3-5). The clinical presentation of this anomaly varies widely, ranging from asymptomatic cases to recurrent infections, hemoptysis, and, in some instances, the development of pulmonary hypertension (6). The true incidence remains unclear, but the anomaly is considered much rarer than pulmonary sequestration and is most often reported as isolated case reports or small case series (3-7). Because systemic arterial pressure is transmitted to the pulmonary circulation, unrecognized cases may lead to serious complications, and misdiagnosis as pulmonary sequestration can result in inappropriate management (8). Although it most frequently occurs as an isolated anomaly, coexistence with other congenital abnormalities—such as partial anomalous pulmonary venous return, bronchopulmonary foregut malformations, congenital cardiac defects, or diaphragmatic anomalies—has also been described (9).

The aim of this report is to present a rare vascular lung anomaly characterized by anomalous systemic arterial supply to the left lower lobe, emphasize the diagnostic value of noninvasive imaging techniques, highlight the importance of differentiating it from pulmonary sequestration, and demonstrate that conservative management may be a safe and appropriate option in selected patients (10).

CASE

A 35-year-old man presented to our cardiology department with a three-month history of fatigue and palpitations accompanied by intermittent mild hemoptysis. He denied dyspnea, cough, or other respiratory symptoms. The patient had no history of chronic disease and did not smoke, consume alcohol, or use any medications. Physical examination was unremarkable. Except for mild hypertriglyceridemia (194 mg/dL), comprehensive laboratory studies, including hematologic, renal, hepatic, and thyroid function tests, were within normal limits. Electrocardiography (ECG) findings were normal. Transthoracic echocardiography revealed a small atrial septal defect (ASD) measuring approximately 3 mm in the interatrial septum. To further characterize the defect, assess right heart chamber volume and function, quantify shunt flow, and evaluate potential surgical or percutaneous management options, cardiac MRI and MR angiography were performed. Phase-contrast MRI-based flow quantification demonstrated pulmonary (Qp) and systemic (Qs) flow rates of 5.04 L/min and 4.76 L/min, respectively, corresponding to a Qp/Qs ratio of 1.06 and a shunt fraction of approximately 6%. These findings indicated no hemodynamically significant shunt and were consistent with normal right heart volumes. Dynamic contrast-enhanced MR angiography demonstrated a 10-mm aberrant systemic artery arising from the descending thoracic aorta and extending toward the left lower lobe, accompanied by mild ectasia of the inferior pulmonary vein (Figure 1).

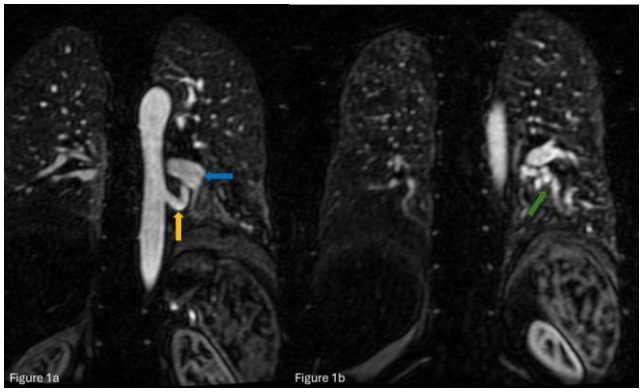

Figure 1: Contrast-enhanced MR angiography images in the coronal plane. The yellow arrow indicates an aberrant arterial vessel originating from the descending thoracic aorta, while the blue arrow demonstrates the ectatic inferior pulmonary vein (a); the green arrow highlights the dilated segmental arterial branches formed by the aberrant artery within the left lower lobe parenchyma (b).

To obtain higher spatial resolution, evaluate the vascular wall and pulmonary parenchyma in greater detail, and more precisely map pulmonary venous drainage using three-dimensional (3D) reformatted images for potential surgical or endovascular planning, thoracic CT angiography was subsequently performed. CT angiography confirmed a 10-mm aberrant systemic artery originating from the descending thoracic aorta at the level of the T8 vertebra and branching into segmental arteries supplying the left lower lobe. The left inferior pulmonary vein was mildly ectatic, and the bronchial tree was normal. Importantly, no dysplastic or cystic parenchymal changes suggestive of pulmonary sequestration were identified. Patchy ground-glass opacities suggestive of increased pulmonary perfusion were observed in the anteromedial basal, lateral basal, and posterior basal segments of the left lower lobe (Figure 2).

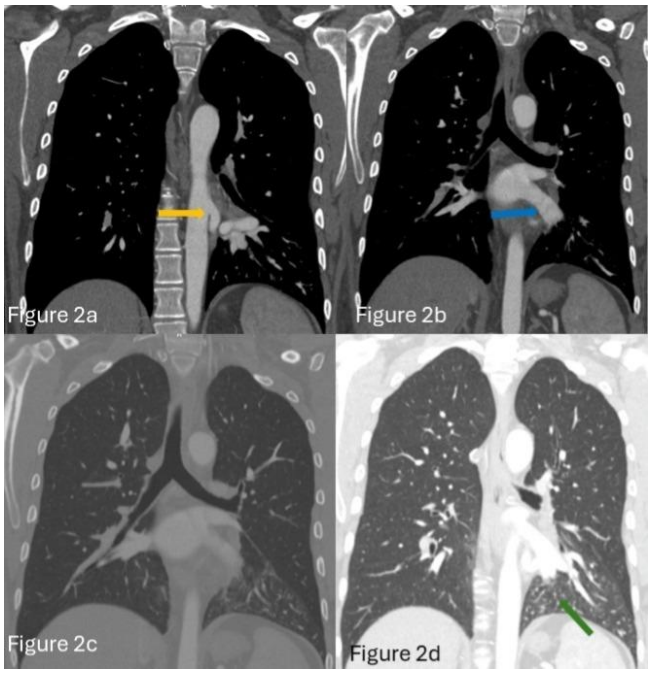

Figure 2: Coronal CT images. Mediastinal window image showing an aberrant arterial vessel originating from the descending thoracic aorta (yellow arrow) (a); mediastinal window image demonstrating the ectatic inferior pulmonary vein (blue arrow) (b); mediastinal window image depicting normal bronchial distribution (c); lung window image showing ground-glass opacities in the left lower lobe secondary to hyperperfusion (green arrow) (d).

Volume-rendered (VR) and maximum intensity projection (MIP) images demonstrated an aberrant systemic artery originating from the descending thoracic aorta and supplying the basal segments of the left lower lobe (Figure 3).

Figure 4 illustrates the imaging features of the left lower lobe, including patchy ground-glass opacities, a mildly ectatic inferior pulmonary vein, the normal course of the left lower lobe bronchus, and the origin and course of the anomalous systemic arterial branch.

Based on these imaging findings, a diagnosis of anomalous systemic arterial supply to the left lower lobe was established. As the patient had no evidence of pulmonary hypertension or a significant shunt, and right heart volumes were within normal limits, conservative management was preferred over surgical or endovascular intervention. During one year of follow-up with evaluations every three months, the patient remained clinically stable without new symptoms or radiologic progression.

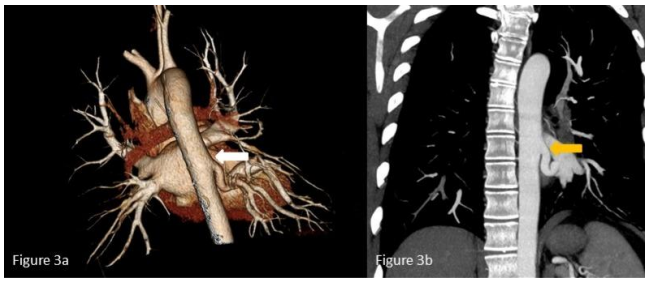

Figure 3: Volume-rendered image showing the aberrant systemic arterial structure originating from the descending thoracic aorta (white arrow) (a); coronal maximum intensity projection (MIP) image demonstrating the same anomalous systemic artery supplying the basal segments of the left lower lobe (yellow arrow) (b).

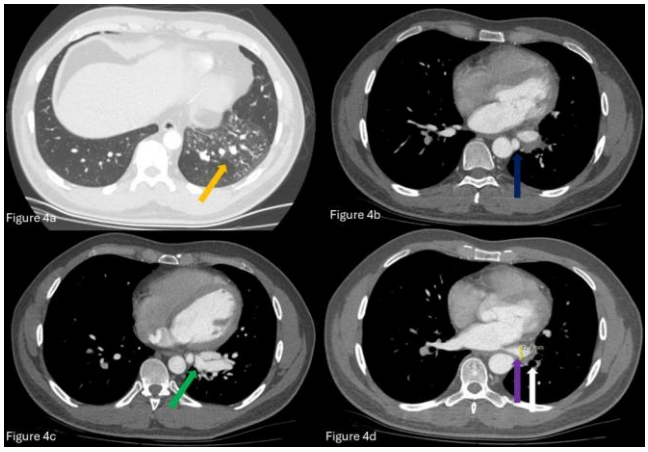

Figure 4: Axial thoracic CT images demonstrating the imaging features of anomalous systemic arterial supply to the left lower lobe. Axial lung window image showing patchy ground-glass opacities in the left lower lobe, indicating abnormal parenchymal hyperperfusion (yellow arrow) (a); axial mediastinal window image demonstrating the origin of an aberrant systemic arterial branch arising from the descending thoracic aorta (dark blue arrow) (b); axial mediastinal window image depicting the course of the dilated anomalous systemic arterial branch supplying the left lower lobe (c); axial mediastinal window image showing a mildly ectatic inferior pulmonary vein (purple arrow) and the normally coursing lower lobe bronchial branch (white arrow) (d).

DISCUSSION

A total of 57 cases of anomalous systemic arterial supply to the left lower lobe have been reported to date, most of which involved the basal segments and received arterial supply from the descending thoracic aorta (3,11). This anatomic pattern is consistent with the findings of Yamanaka et al. (11), whose 15-patient series demonstrated uniform involvement of the basal segments, with aberrant arteries arising exclusively from the descending thoracic aorta. Yan et al. (12) evaluated 23 patients and reported that hemoptysis was the most frequent presenting symptom, occurring in approximately one-third of cases, while all patients exhibited a large-caliber systemic artery arising from the descending thoracic aorta. Wu et al. (10) emphasized the diagnostic superiority of three-dimensional CT angiography, demonstrating its ability to clearly depict the origin, course, and branching pattern of the aberrant systemic artery and to guide preoperative

planning. Most reports in the Western literature consist of isolated case reports, which consistently demonstrate preserved bronchial communication, normal pulmonary parenchyma, and venous drainage via the pulmonary veins (1-5,13,14). These findings suggest that systemic arterialization can occur in the absence of pulmonary sequestration and underscore the diagnostic importance of CT angiography (14).

Imaging plays a central role in the diagnosis of anomalous systemic arterial supply to the left lower lobe (10). Multidetector CT angiography (MDCTA) is considered the gold standard, as it provides high spatial resolution for delineating the origin, course, and caliber of the aberrant artery, as well as pulmonary venous drainage patterns (10-12). Three-dimensional reformatted and multiplanar reconstruction images clearly demonstrate the relationship between the aberrant artery and the adjacent parenchyma, offering valuable anatomic information for surgical or endovascular planning (14,15). Cardiac MRI and contrast-enhanced MR angiography are valuable complementary modalities, especially in young or asymptomatic patients, as they allow noninvasive evaluation of right heart volumes, shunt quantification (Qp/Qs), and potential volume overload without radiation exposure (12).

The main differential diagnosis includes pulmonary sequestration, pulmonary arteriovenous malformation, and bronchopulmonary foregut malformations. Compared with pulmonary sequestration, anomalous systemic arterial supply to the left lower lobe demonstrates normal bronchial distribution, absence of cystic or dysplastic parenchyma, a dilated left inferior pulmonary vein, and an enlarged anomalous systemic artery arising from the descending thoracic aorta. Ground-glass opacities may reflect increased perfusion rather than infection or fibrosis, which are typical findings in pulmonary sequestration (1,5). Pulmonary arteriovenous malformations differ by demonstrating direct arteriovenous communication with early venous filling on contrast-enhanced computed tomography, whereas anomalous systemic arterial supply maintains an intervening capillary bed between the arterial and venous systems (9). Bronchopulmonary foregut malformations may mimic vascular anomalies but usually demonstrate fistulous communication with the esophagus or stomach and are often associated with gastrointestinal symptoms (8). A comprehensive evaluation of the systemic arterial origin, bronchial communication, and pulmonary venous drainage pattern is therefore essential for accurate differential diagnosis.

Inferior pulmonary vein ectasia constitutes an important ancillary imaging feature in anomalous systemic arterial supply to the left lower lobe and reflects chronic exposure of the pulmonary venous system to high-pressure systemic arterial inflow. In their CT-based comparative study, Qin et al. (16) demonstrated that dilatation of the inferior pulmonary vein is significantly more frequent in anomalous systemic arterial supply than in pulmonary sequestration, in which venous drainage is often abnormal or systemic and parenchymal dysplasia is typically present. This venous finding therefore supports preserved pulmonary venous return and aids in distinguishing these two entities. Similarly, Miyake et al. (17) described characteristic CT features of systemic arterial supply to normal basal lung segments and emphasized that enlargement of the pulmonary veins represents a secondary hemodynamic adaptation rather than a primary venous anomaly. Furthermore, Do et al. (18) highlighted that careful assessment of pulmonary venous caliber and drainage patterns on multidetector CT is essential for accurate diagnosis of systemic arterial supply to the lung and for avoiding misclassification as pulmonary sequestration. Collectively, these observations indicate that inferior pulmonary vein ectasia, when evaluated together with arterial anatomy and preserved bronchial structure, provides valuable diagnostic support in anomalous systemic arterial supply to the left lower lobe.

Therapeutic management depends on the patient's symptoms, hemodynamic status, and the presence of complications. Surgical resection and endovascular embolization are the most commonly employed approaches in symptomatic cases. Yamanaka et al. (11) treated all symptomatic patients surgically and reported complete clinical recovery. Sun et al. (13) highlighted coil or vascular plug embolization as a safe and effective alternative to surgery, especially in patients with smaller aberrant arteries or those unsuitable for operative management. Recently, conservative management has gained attention for asymptomatic or mildly symptomatic patients (9,14). This strategy is generally preferred when the systemic artery is of small caliber, pulmonary venous drainage is normal, and no significant shunt is present (7,9). Overall, current evidence supports surgical or endovascular intervention for symptomatic patients and careful clinical and radiologic follow-up for asymptomatic individuals as an appropriate management approach (7,9,10).

This case illustrates a mildly symptomatic presentation of anomalous systemic arterial supply to the left lower lobe and demonstrates that detailed anatomic assessment can be reliably achieved using noninvasive imaging methods. The patient's stable course without intervention supports the view that conservative management may be a safe option in appropriately selected individuals. In asymptomatic or mildly symptomatic patients, including those with minimal hemoptysis, conservative follow-up with regular clinical and radiologic monitoring appears reasonable.

Several limitations of this case report should be acknowledged. First, as the findings are based on a single patient, the results cannot be generalized to all individuals with this rare anomaly, given the potential for significant anatomic and physiologic variations. Second, although the patient remained clinically and radiologically stable during one year of follow-up, this period may not be sufficient to fully assess the long-term risks of conservative management, such as the late development of pulmonary hypertension or high-output heart failure. Third, the conservative management strategy was not compared with surgical or endovascular treatment, precluding a direct assessment of relative efficacy in this specific case. Larger, multicenter prospective studies with extended surveillance are necessary to establish more robust, evidence-based criteria for selecting candidates for conservative versus interventional management.

In conclusion, anomalous systemic arterial supply to the left lower lobe is a rare vascular anomaly that may be misidentified as pulmonary sequestration but can be accurately differentiated through detailed radiologic evaluation. CT angiography remains the primary modality for assessing the origin, course, and venous drainage of the aberrant vessel. Careful interpretation of noninvasive imaging findings helps prevent unnecessary surgical intervention and supports safe conservative management in suitable patients.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - A.A., B.Y.K.; Planning and Design - A.A., B.Y.K.; Supervision - A.A., B.Y.K.; Funding -; Materials - A.A., B.Y.K.; Data Collection and/or Processing - A.A.; Analysis and/or Interpretation - A.A.; Literature Review - A.A.; Writing - A.A., B.Y.K.; Critical Review - A.A., B.Y.K.

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