

Received: 2020.05.29

Accepted: 2020.10.22

Available online: 2020.10.30

Published: 2020.12.12

Isolated Systemic Arterial Supply to Normal Lung with Aneurysm Formation: A Rare Entity with An Even Rarer Complication

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 61-year-old
Final Diagnosis: Isolated systemic arterial supply to normal lung
Symptoms: Asymptomatic
Medication: —
Clinical Procedure: —
Specialty: Radiology

Objective: Rare disease
Background:

Isolated systemic arterial supply to normal (unsequestered) lung (ISSNL) without associated pulmonary malformation is rare, and lies towards the milder end of the spectrum of congenital lung abnormalities. Aneurysmal dilatation of the anomalous artery is an infrequent complication, with only 5 published cases thus far.

Case Report: We present the case of a 61-year-old man whose screening chest radiograph showed a retrocardiac mass. Further evaluation with axial imaging demonstrated an ISSNL, complicated by aneurysmal dilatation. The genesis of this condition has been postulated to be due to persistence of primitive aortic branches to the developing lung bud. Initially reported in 1777, this entity is now more accurately classified within the spectrum of pulmonary and bronchovascular abnormalities, with refinement of the latter.

The origin of an aberrant artery from the aorta implies that a higher-pressure systemic circulation is being shunted into a lower-pressure pulmonary circulation. While these supplying arteries are known to be large, aneurysmal dilatation is exceptionally rare. Here, we review the cases published in the literature and present a case of our own. We aim to describe its pathogenesis, and touch on the classification systems and management.

Conclusions: ISSNL is usually first suspected on a screening chest radiograph, as many patients are asymptomatic. Based on contrast-enhanced axial imaging, the diagnosis can be established non-invasively. Definitive management includes surgical and endovascular techniques.

MeSH Keywords: Aneurysm • Bronchopulmonary Sequestration • Scimitar Syndrome

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/926409>



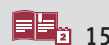
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Background

Isolated systemic arterial supply to normal (unsequestered) lung (ISSNL), without congenital pulmonary abnormalities, is a rarity, with only 51 cases reported in the English medical literature [1]. Although there is high pressure within this vessel and left-to-left shunting, most of the patients are asymptomatic. Aneurysm of this vessel is an exceptionally infrequent complication, with a total of 5 cases reported in the literature to date [2–6].

Our case report presents a rare case of ISSNL associated with aneurysmal dilatation, which is an even rarer complication. We briefly review the classification systems which have evolved since the initial one by Pryce to better incorporate this entity within the spectrum of pulmonary and bronchovascular abnormalities. We also discuss the pathogenesis of aberrant systemic arterial supply to the lungs leading to formation of an aneurysm, as well as management of this complication.

Case Report

We present the case of a 61-year-old man who presented at our public health institution after a screening chest radiograph demonstrated the presence of a retrocardiac mass. Unfortunately, the screening chest radiograph was performed in the private practice sector and could not be obtained.

Nevertheless, the patient subsequently transferred to and was followed up at our institution. A single-phase contrast-enhanced computed tomography (CECT) scan of the thorax was performed as per institutional protocol for evaluation of a lung mass. It revealed an enhancing oval-shaped lesion deriving its arterial supply from the descending thoracic aorta (Figure 1). The findings were discussed during the multidisciplinary team meeting, following which a CT angiogram (CTA) was performed. This scan (Figure 2A–2C) confirmed a conglomerate of tortuous and dilated vessels in the lower lobe of the left lung. The feeding artery arose from the descending thoracic aorta and was noted to be hypertrophied, measuring 11 mm in diameter. This artery had aneurysmal dilatation of its mid-segment, measuring 3.8 cm in maximal diameter. Drainage was via the left inferior pulmonary vein. The absence of communication with the pulmonary artery excluded an arterio-arterial shunt (Figure 2D). As the tracheobronchial tree was normal and no associated sequestration abnormality was detected (Figure 2E), this was diagnosed as anomalous systemic arterial supply to normal lung, complicated by aneurysmal dilation of the supplying systemic artery. Mild scarring and bronchiectasis with areas of atelectasis were also noted at the lung base (Figure 2F). The patient remained asymptomatic and his lung function test results were normal. He opted for conservative management.

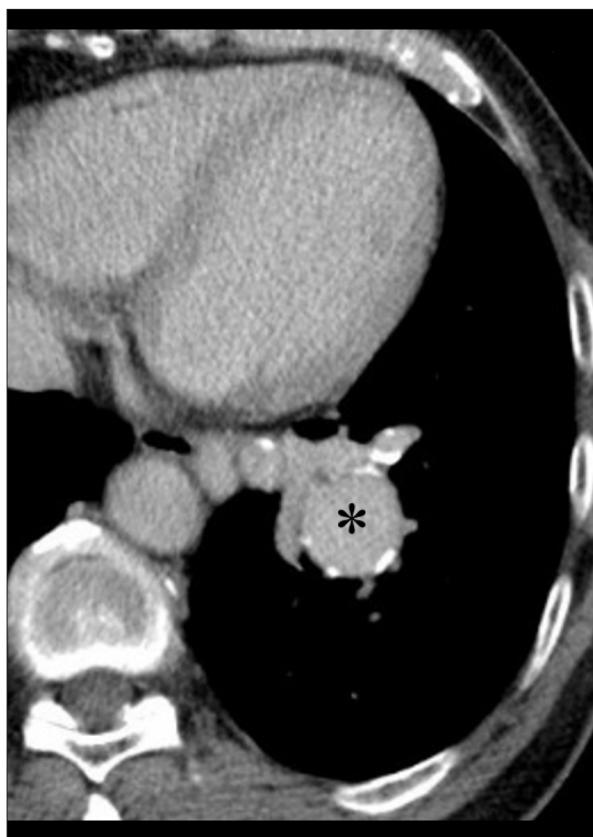


Figure 1. Axial CECT image in the portal venous phase showing an enhancing oval-shaped lesion (asterisk) in the lower lobe of the left lung. Portions of its walls show mural calcification. Adjacent to it was a conglomerate of tortuous and dilated vessels deriving its arterial supply from the descending thoracic aorta.

Discussion

Aberrant systemic arterial supply to the lungs occurring in conjunction with other congenital pulmonary malformations, including pulmonary sequestration and hypogenetic lung or scimitar syndrome, have been extensively described in the literature [7]. ISSNL, which is devoid of any associated pulmonary abnormalities, is rare [4,7–9]. Fifty-one cases have appeared in the English medical literature, 92% of which were of Asian descent [1]. These authors postulate a genetic predisposition and an expected rise in its incidence in this region. This condition also has a predilection for males, and is more commonly seen on the left side, as was the case in our patient [2,3,7,8,10]. This has previously been postulated to be the result of persistence of primitive aortic branches to the developing lung bud, instead of regression in lieu of the pulmonary artery taking over [2–4,7,8,10,11]. More often than not, the anomalous arterial supply arises from the descending thoracic aorta, although the literature has described origins which have also occurred from below the level of the diaphragm, such as from

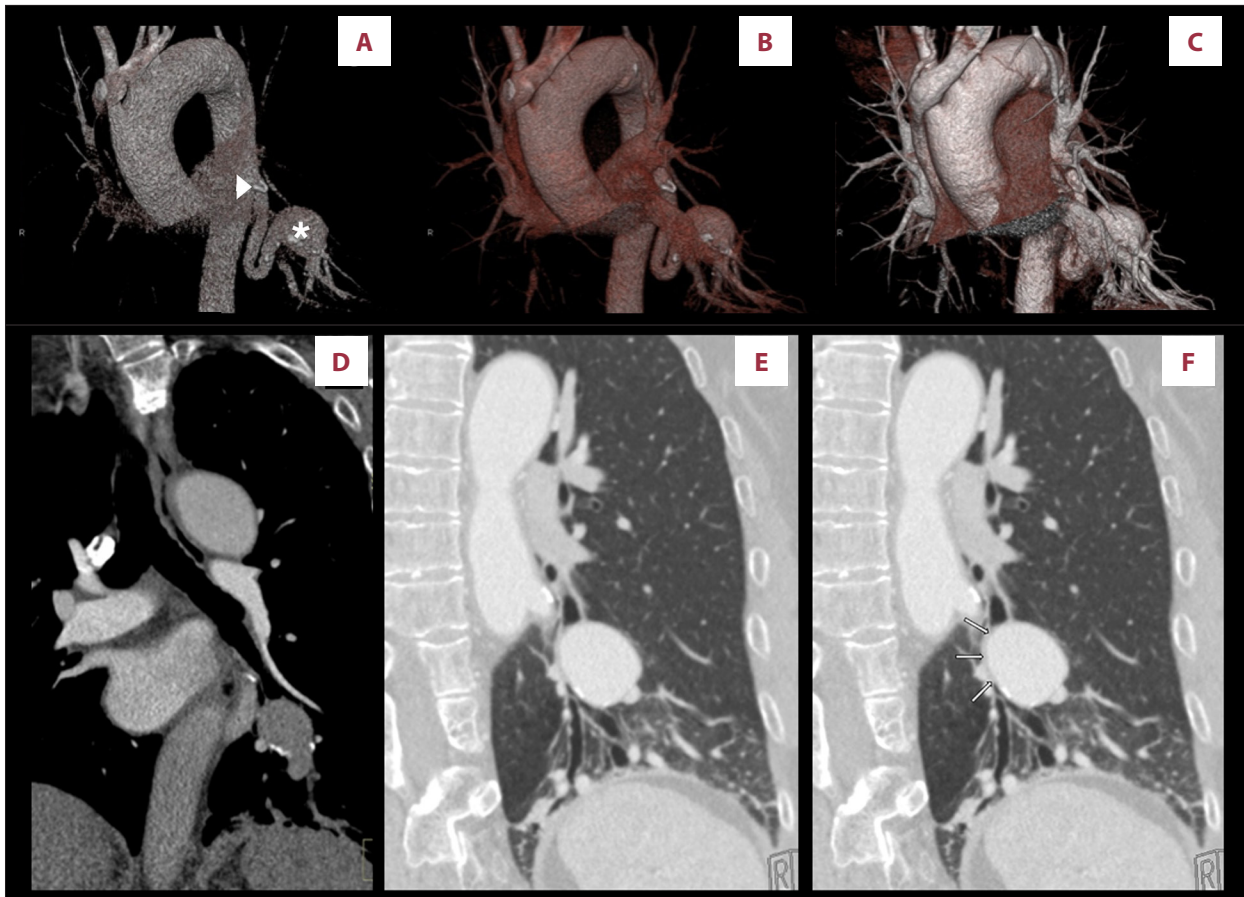


Figure 2. Volume-rendered, coloured 3D reformat images of the CTA using opacity threshold for segmentation. (A) Image showing arterial supply originating from the descending thoracic aorta (arrowhead). The proximal segment is tortuous with aneurysm formation at its mid-segment (asterix). The distal branches have a normal caliber, with gradual tapering. (B) The left inferior pulmonary vein (V) commences below the aneurysm sac, then courses anteromedially before joining with the left superior pulmonary vein to drain into the left atrium. There is no direct communication between these veins and the systemic artery. (C) The pulmonary arteries have been included, showing paucity of branches to the left lower lobe, which is supplied by the aberrant systemic artery. CTA with curved coronal reformats. (D) Image in the pulmonary angiographic phase shows a gracile left lower-lobe pulmonary artery coursing alongside the lower-lobe bronchus before terminating early. There is no communication with the aberrant arterial supply and no opacification of the aneurysm sac. (E) Image showing continuation of the tracheobronchial tree into the left lower lobe. (F) Mild scarring and bronchiectasis with areas of atelectasis at the left lung base were attributed to compression of the lower-lobe segmental bronchi by the aneurysm (arrows).

the suprarenal abdominal aorta or celiac axis, and, more rarely, from the subclavian or internal mammary vessels [8,12,13]. In addition, a rare case of anomalous systemic arterial supply to normal lung coexisting with the pulmonary artery has also been described [13]. Venous drainage is normal, via the inferior pulmonary vein into the left atrium.

ISSNL was first reported by Huber in 1777, but remained unclassified until discovered later in association with lung parenchymal abnormalities and sequestration. These included extralobar sequestration in 1861 and intralobar sequestration by Pryce in 1946 [14]. The initial attempt at classification by Pryce was unsatisfactory as it assumed ISSNL to be a type of sequestration, which was a misnomer. The concept of

a ‘sequestration spectrum’ was proposed by Sade in 1974 and advanced by Clements and Warner as the ‘pulmonary malinoculation spectrum’ in 1987 [14,15]. The latter classification was more inclusive, as it considered all permutations of anomalies involving (1) the tracheobronchial tree and/or arterial supply, (2) venous drainage, and (3) lung parenchyma. In the present era, our case is best classified as ISSNL or “pseudosequestration of the lung” [2,10], which is in a category of its own, but at the milder end of this spectrum [4,7,11].

Whether an aberrant artery supplies a normal or an abnormal lung, its origin from the aorta implies that a higher-pressure systemic circulation is being shunted into a lower-pressure pulmonary circulation. While these supplying arteries are known to be

large, aneurysmal dilation of this vessel is exceptionally rare. There have been only 5 reported cases of aneurysm of the aberrant vessel supplying an ISSNL in the English literature [2–6]. The patients ranged from 25 to 78 years old, and all had a mass on imaging, similar to our patient. Four of them were asymptomatic, while 1 presented with hemoptysis. Imaging of all these patients revealed an aberrant arterial supply to either the right (n=1) or left (n=4) lower lobes. The diameters of the aneurysms ranged from 1.8 to 5 cm in diameter. One patient developed a further complication of thrombosis of the aneurysmal artery [6]. These aberrant vessels, being major aortopulmonary collateral arteries (MAPCAs), are predisposed to aneurysm formation, as they have a more elastic rather than muscular wall [4,11,12]. Wall thickening is often seen secondary to systemic arterial hypertension, with a tendency to develop atheromatous changes [3,4,12]. The mild scarring and bronchiectasis in our patient might have been related to compression of the lower-lobe segmental bronchi by this aneurysm. Although this has yet to be reported, the aneurysm can potentially rupture, resulting in life-threatening outcomes and increased mortality [3,4].

Chest radiographs often demonstrate a lobulated retrocardiac or paraspinal mass, representing the conglomerate of tortuous vasculature [7]. Further evaluation with CECT and CTA of the thorax plays an important role by confirming vascular nature of the mass, identifying the origin and course of the aberrant supplying artery, and excluding any possible anomalous venous drainage [7,8,13]. These scans also aid in ruling out associated lung parenchymal anomalies, which would be present in sequestration, or, more rarely, hypogenetic lung syndrome [7]. Finally, imaging is useful in demonstrating complications such as aneurysmal dilation of the anomalous systemic arterial supply or scarring and bronchiectasis secondary to its mass-effect, as in our example.

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Early radiological diagnosis is imperative to effect definitive treatment, especially in clinically symptomatic patients who experience hemoptysis and heart failure. Differentiating ISSNL from sequestration is important, as minimally invasive techniques such as angioembolization may be employed in lieu of surgical options such as segmentectomy or lobectomy [7,8,13]. There is no consensus regarding the choice of embolic material [12]. In the case of complication by aneurysmal dilation, the potential risk of rupture will also be abolished [4]. Three of the reported aneurysmal cases subsequently underwent surgical resection [4–6], another opted for endovascular embolization [2], while the last declined intervention [3].

Conclusions

ISSNL is usually first suspected on a screening chest radiograph, as the majority of these patients are asymptomatic. Based on CECT and CTA of the thorax, the diagnosis can be established in a non-invasive fashion. Demonstrating a normal bronchial anatomy on imaging is crucial in distinguishing this entity from sequestration or hypogenetic lung syndrome. Definitive management includes surgical and, more recently, minimally invasive endovascular techniques.

Department and Institution where work was done

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