A case of systemic arterial supply to the right lower lobe of the lung: imaging findings and review of the literature

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ABSTRACT

Systemic arterialization of the lung without pulmonary sequestration is the rarest form of anomalous systemic arterial supply to the lung. This condition is characterised by an aberrant arterial branch arising from the aorta which supplies an area of lung parenchyma with normal bronchopulmonary anatomy. It is often diagnosed following investigation of an incidental cardiac murmur or based on abnormal imaging, as most patients are asymptomatic or minimally symptomatic. Thoracic computed tomography and computed tomography angiography are generally the most useful diagnostic tests. We present a case of a 22-year old female who was diagnosed with systemic arterial supply to a portion of otherwise normal right lower lobe following investigation of low volume haemoptysis.

CASE REPORT

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A 22-year-old female presented to our institution with a two-day history of low volume haemoptysis. On questioning, she admitted to having several similar, but less pronounced episodes of haemoptysis as well as an otherwise non-productive chronic cough over the preceding three years, for which she had not sought medical attention. She was otherwise well. On presentation, Chest X-Ray (Figure 1) and laboratory findings were unremarkable.

Subsequent thoracic computed tomography (CT) angiography demonstrated a small well circumscribed area of ground glass density in the medial basal segment of the right lower lobe, exclusively supplied by a single systemic artery arising from the descending thoracic aorta (Figure 2). Multiplanar reformatted images clearly demonstrated the exclusive systemic arterial supply to this segment and atresia of the pulmonary artery branch to the right lower lobe, but with a more typical pulmonary venous drainage pattern (Figure 3) - compatible with a diagnosis of systemic arterialization of the lung without sequestration. The patient was also incidentally noted to have an anomalous tracheal bronchus supplying the right upper lobe (Figure 4). The patient was counselled as to potential treatment options prior to discharge and then again at review two weeks later. As she remained well with no further episodes of bleeding, she declined definitive treatment as this time.

DISCUSSION

Anomalous systemic arterial supply to the lungs has been extensively described in association with other congenital abnormalities such as bronchopulmonary sequestration and hypogenetic lung syndrome (scimitar syndrome). In 1946, Pryce introduced the term 'sequestration' to describe congenital abnormalities characterised by an anomalous systemic arterial supply to the lung and atresia or hypoplasia of the pulmonary artery [1,2]. Sequestration was defined by Pryce as a "disconnected (dislocated, ectopic) bronchopulmonary mass or cyst with an anomalous systemic blood supply" [1].

Since Pryce's early description of sequestration, a wide spectrum of bronchopulmonary-vascular malformations have been recognised, many of which do not fulfil the original definition. Our case differs from traditional descriptions of sequestration, in that a portion of normal lung receives an anomalous systemic blood supply. Sade et al. proposed the concept of a 'sequestration spectrum', in order to encompass and describe the various combinations of abnormal bronchial connection, arterial supply and venous drainage [3]. As a similar spectrum concept, Clements and Warner introduced a simple descriptive anatomical approach to such complex bronchopulmonary-vascular malformations named 'pulmonary malinosculation spectrum' [4] (Table 1). This classification includes all congenital lung abnormalities in which there is malinosculation (anomalous communication between blood vessels or other tubular structures) and is constructed in three steps [4,5]. First, the abnormalities of bronchopulmonary airway, arterial supply, or both are classified as bronchopulmonary malinosculation, arterial pulmonary malinosculation and bronchoarterial pulmonary malinosculation, respectively. The second step in the classification defines the associated abnormalities of venous drainage, and the third step describes associated abnormalities of lung parenchyma [4]. According to the concept introduced by Clements and Warner, our case can be classified as arterial pulmonary malinosculation.

Systemic arterialization of the lung without pulmonary sequestration is the rarest form of anomalous systemic arterial supply to the lung [2,6]. This condition is characterised by an aberrant systemic arterial branch which can arise from the thoracic portion of the descending aorta, or less commonly from the abdominal aorta or coeliac axis, supplying an area of lung parenchyma with normal bronchopulmonary system [2,7,8]. The left lower lobe is most often involved, and there is commonly an atresia of the corresponding pulmonary artery with a normal venous drainage [6,9]. Interestingly, our case presents an anomalous systemic arterial supply to a non-sequestered portion of the right lower lobe in association with an abnormal tracheal bronchus supplying the right upper lobe. Accompanying anomalies are uncommonly seen in systemic arterialization of the normal lung, whereas 14-59% of cases of sequestration are associated with other malformations [5,6], such as diaphragmatic hernia, congenital cystic adenomatoid malformation, and congenital heart disease [10,11].

The aetiology of this anomaly is not completely understood; a persistent remnant of the embryonic connection between the aorta and the pulmonary parenchyma is thought to be responsible [8,12]. It has been proposed that systemic arterial supply to an area of otherwise normal lung may form part of the sequestration spectrum in that it is a mild form of the process that, when more severe, may cause persistence of remnants of the embryonic arterial connection and sequestration of a portion of lung parenchyma [12].

The prevalence of anomalous arterial supply to the lung without sequestration is very low - only 10 cases had been reported in all English and Chinese literature up to 1994 [13]. Yamanaka et al. reviewed 12 cases in the English literature up to 1999 and reported an age distribution ranging from 0 months to 68 years, with an average age of 21.2 years [8]. Male gender with a ratio of 2:1, left side and descending thoracic aorta as the aberrant arterial origin were predominant [8]. Systemic arterial supply to the right lower lobe comprised 16% of cases. An estimated incidence of systemic arterial supply to the normal lung has never been published - this is thought to be due to the rarity of this condition [14].

The majority of patients are asymptomatic, however recurrent pulmonary infection, cardiac failure due to left-to-left shunt and haemoptysis are possible clinical presentations [8]. The proposed explanation for the intermittent episodes of haemoptysis reported by our patient is the higher pressure of systemic circulation generated in the abnormally perfused segments of the right lower lobe resulting in intra-alveolar haemorrage.

Systemic arterial supply to the lung without sequestration may be diagnosed following investigation of an incidental cardiac murmur [6,12], especially in children, or based on abnormal imaging [8]. A retrocardiac tubular structure or a focal area of consolidation may be evident on chest X-Ray [15]. Computed tomography may reveal a focal area of ground glass density, indicating an area of relative hypervascularity and/or intra-alveolar haemorrhage. The CT findings of ground glass density in our patient likely represent air space haemorrhage. CT can also demonstrate an aberrant systemic arterial supply to part of the lung [2,8]. Thoracic CT is the most useful test in the evaluation of patients with suspected abnormal systemic arterial supply to the lung, as it demonstrates both the bronchial and vascular anatomy of the lung [9]. CT angiography can clearly depict the origin of the aberrant systemic artery. MRI has also been reported to be useful in identifying an anomalous vessel originating from the aorta [13].

The radiological differential diagnoses include true sequestration and, less likely, hypogenetic lung syndrome, as both conditions are associated with systemic arterial supply to the lung. On plain radiograph, intralobar sequestration usually manifests as an intrapulmonary lesion in the left lower lobe [16]. A patch of consolidation or a homogenous opacity may be seen in uncomplicated cases. Cystic spaces may become evident as a result of acute infection or chronic inflammation, and they may contain air-fluid level indicating bronchial communication [17]. Extralobar sequestration commonly appears as a single, well-defined homogenous opacity located in the pleural space between the left lower lobe and the

diaphragm [17,18]. Computed tomography has the advantage of revealing the abnormal lung parenchyma and the arterial and venous anatomy [17]. Evidence of a homogenous or heterogenous lung mass supplied by a systemic arterial branch on CT should raise suspicion of lung sequestration. MRI can be useful to differentiate the cystic, solid, haemorrhagic and mucous components of the lung mass, as well as providing details of the venous drainage and the abnormal systemic artery [12,17].

On the other hand, hypogenetic lung syndrome is associated with volume loss of the right lung, which is usually evident on chest X-Ray [16]. CT angiography and MRI reveal the typical vascular abnormalities of this rare condition. In its complete form, scimitar syndrome consists of ipsilateral anomalous pulmonary drainage into the inferior vena cava along with hypoplasia or other malformations of the right pulmonary artery, and anomalous systemic arterial supply to the lower lobe of the right lung [16].

Treatment is recommended in both symptomatic and asymptomatic patients in order to prevent haemoptysis [5]. Various treatment modalities have been described, including lobectomy or segmentectomy, systemic artery to pulmonary artery anastomosis and endovascular treatment. Differentiation between this condition from classic sequestration has important treatment implications as it may allow non-operative management (angiographic embolization) of the aberrant systemic artery [5].

TEACHING POINT

Systemic arterialization of the lung without sequestration is a rare condition often diagnosed following investigation of an incidental cardiac murmur or based on abnormal chest X-Ray or CT of the thorax, as most patients are asymptomatic. CT is the most useful diagnostic test as it demonstrates both the bronchial and vascular anatomy of the lung while CT angiography can clearly depict the origin of the aberrant systemic artery, avoiding invasive techniques for the diagnosis.

REFERENCES

1. Pryce, DM. Lower accessory pulmonary artery with intralobar sequestration of lung: a report of seven cases. J Pathol Bacteriol. 1946 Jul;58(3): 457-67. PMID: 20283082

2. Flisak ME, Chandrasekar AJ, Marsan RE, Ali MM. Systemic arterialization of lung without sequestration. AJR Am J Roentgenol. 1982 Apr;138(4):751-3. PMID: 6978045

3. Sade RM, Clouse M, Ellis FH Jr. The spectrum of pulmonary sequestration. Ann Thorac Surg. 1974 Dec;18(6):644-58. PMID: 4611367

4. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. Thorax. 1987 Jun;42(6):401-8. PMID: 3660297

5. Agarwal PP, Matzinger FR, Seely JM, Rasuli P, Shamji FM. An unusual case of systemic arterial supply to the lung with bronchial atresia. AJR Am J Roentgenol. 2005 Jul;185(1):150-3. PMID: 15972415

6. Brühlmann W, Weishaupt D, Goebel N, Imhof E. Therapeutic embolization of a systemic arterialization of lung without sequestration. Eur Radiol. 1998;8(3):355-8. PMID: 9510564

7. Hiramatsu M, Iwashita M, Inagaki T et al. Anomalous systemic arterial supply to separate lingular and basal segments of the lung: an anatomic consideration. Ann Thorac Surg. 2009 Oct;88(4):1358-60. PMID: 19766847

8. Yamanaka A, Hirai T, Fujimoto T, Hase M, Noguchi M, Konishi F. Anomalous systemic arterial supply to normal basal segments of the left lower lobe. Ann Thorac Surg. 1999 Aug;68(2):332-8. PMID: 10475391

9. Irodi A, Cherian R, Keshava SN, James P. Dual arterial supply to normal lung: within the sequestration spectrum. Br J Radiol. 2010 May;83(989):e86-9. PMID: 20418461

10. Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. Thorax. 1979 Feb;34(1):96-101. PMID: 442005

11. Chen W, Wagner L, Boyd T, Nagarajan R, Dasgupta R. Extralobar pulmonary sequestration presenting with torsion: a case report and review of literature. J Pediatr Surg. 2011 Oct;46(10):2025-8. PMID: 22008345

12. Ellis K. Fleischner lecture. Developmental abnormalities in the systemic blood supply to the lungs. AJR Am J Roentgenol. 1991 Apr;156(4):669-679. PMID: 2003427

13. Muñoz JJ, García JA, Bentabol M, Padín MI, Serrano F. Endovascular treatment of hemoptysis by abnormal systemic pulmonary artery supply. Cardiovasc Intervent Radiol. 2008 Mar-Apr;31(2):427-30. PMID: 17593429

14. Chang JM, Cheng JH, Chen W. An unusual cause of hemoptysis in a young woman. Ann Thorac Surg. 2012 May;93(5):1734. PMID: 22541216

15. Lim HH, Huh JK, Kim JH et al. Therapeutic embolization of systemic arterial supply to normal basal segments of the left lower lobe of the lung. Respirology. 2009 May;14(4):606-8. PMID: 19402839

16. Berrocal T, Madrid C, Novo S, Gutiérrez J, Arjonilla A, Gómez-León N. Congenital anomalies of the tracheobronchial tree, lung, and mediastinum: embryology, radiology, and pathology. Radiographics. 2004 Jan-Feb;24(1):e17. PMID: 14610245

17. Abbey P, Das CJ, Pangtey GS, Seith A, Dutta R, Kumar A. Imaging in bronchopulmonary sequestration. J Med Imaging Radiat Oncol. 2009 Feb;53(1):22-31. PMID: 19453525

18. Alizadeh E, Suliman H. Intralobar pulmonary sequestration. JBR-BTR. 2013 Jul-Aug;96(4):208-9. PMID: 24224295

FIGURES

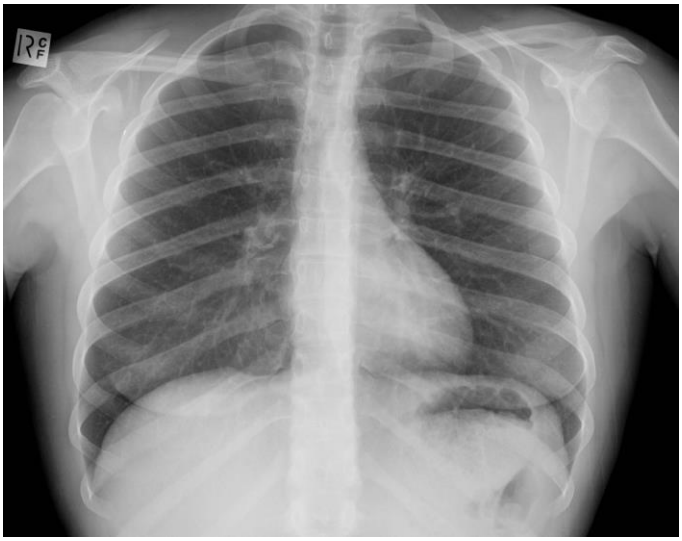
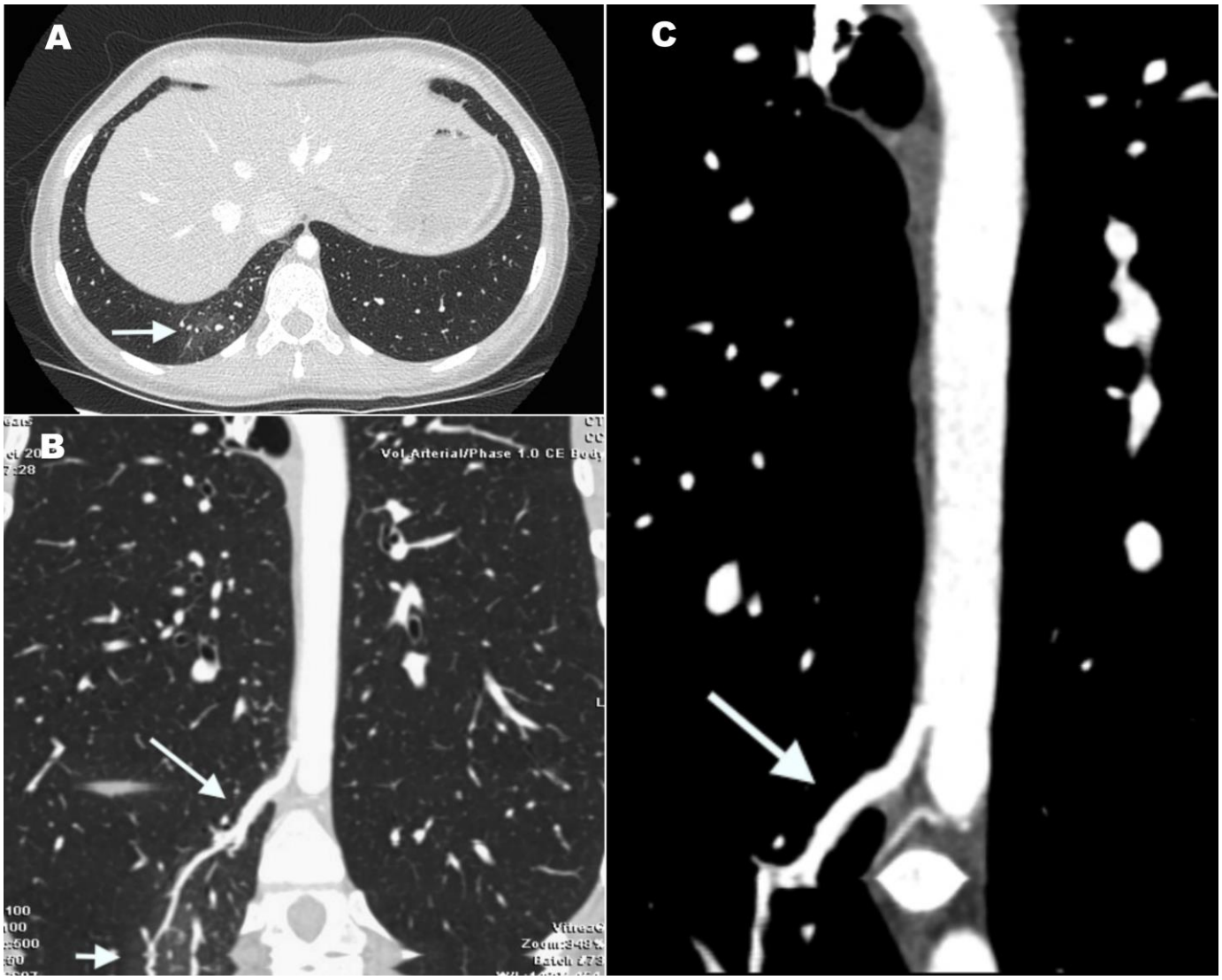


Figure 1 (left): 22 year-old female with systemic arterialization of the lung without sequestration. FINDINGS: Normal postero-anterior chest X-Ray.

Figure 2 (bottom): 22 year-old female with systemic arterialization of the lung without sequestration. FINDINGS: A focal area of ground glass density is evident on axial CT (a) and coronal curved multiplanar reformatted image (MPR) (b, short arrow) on lung windows. Coronal curved MPR also demonstrates an aberrant artery arising from the right, inferior aspect of the descending thoracic aorta, supplying the medial right lower lobe (long arrow in b, and c).
TECHNIQUE: Aquillion 160 CT scanner (Toshiba, Japan), 200mA, 100kV, 3mm slice thickness, 70 cc Vispaque contrast injected IV at 3cc/second.



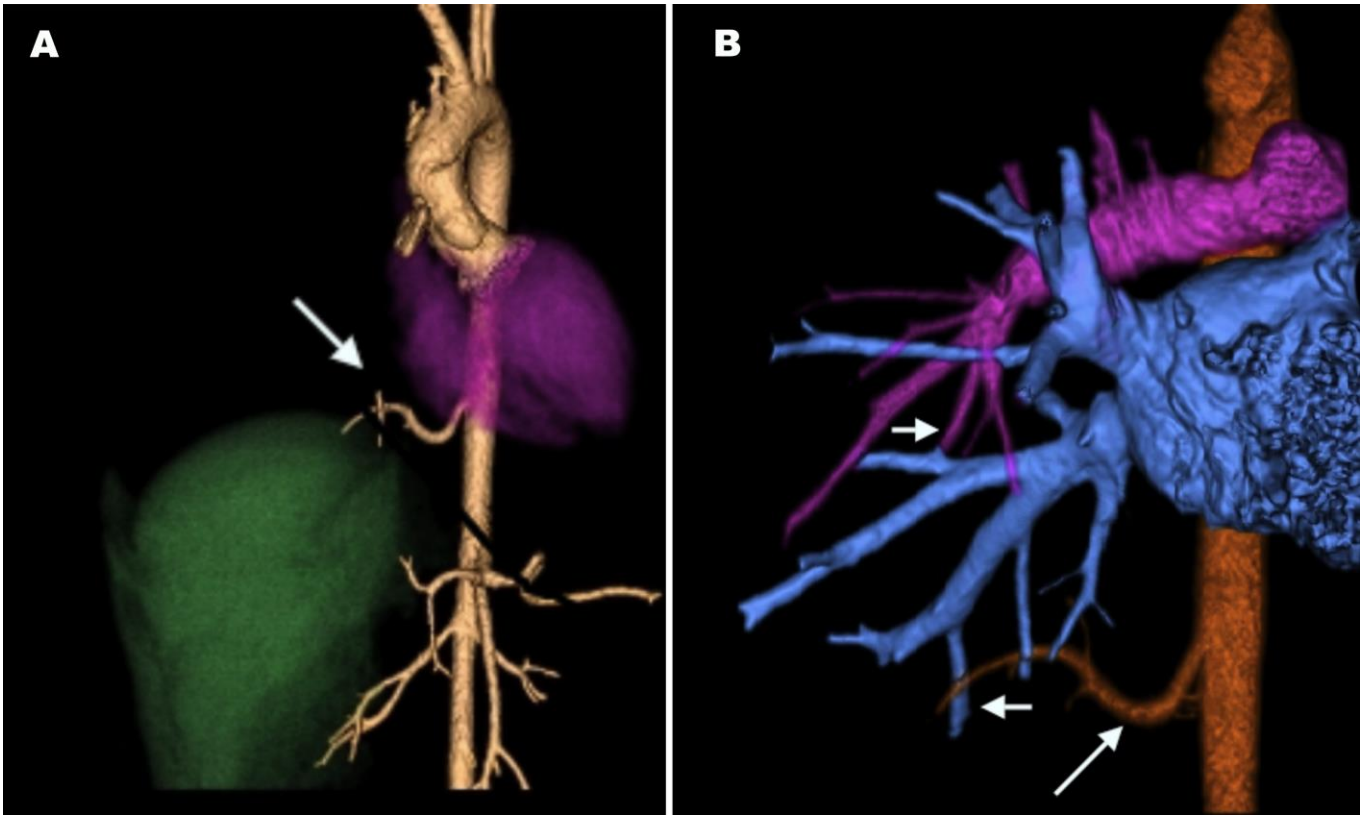


Figure 3: 22 year-old female with systemic arterialization of the lung without sequestration. FINDINGS: Volume rendered, coloured 3D reformatted CT depict the anomalous arterial supply to the right lower lobe (a and b, long arrow) and atretic pulmonary arterial branch to the right lower lobe (b; pink), with preserved pulmonary venous drainage (b; blue).
TECHNIQUE: Aquillion 160 CT scanner (Toshiba, Japan); 200mA, 100kV, 3mm slice thickness, 70 cc Vispaque contrast injected IV at 3cc/second.

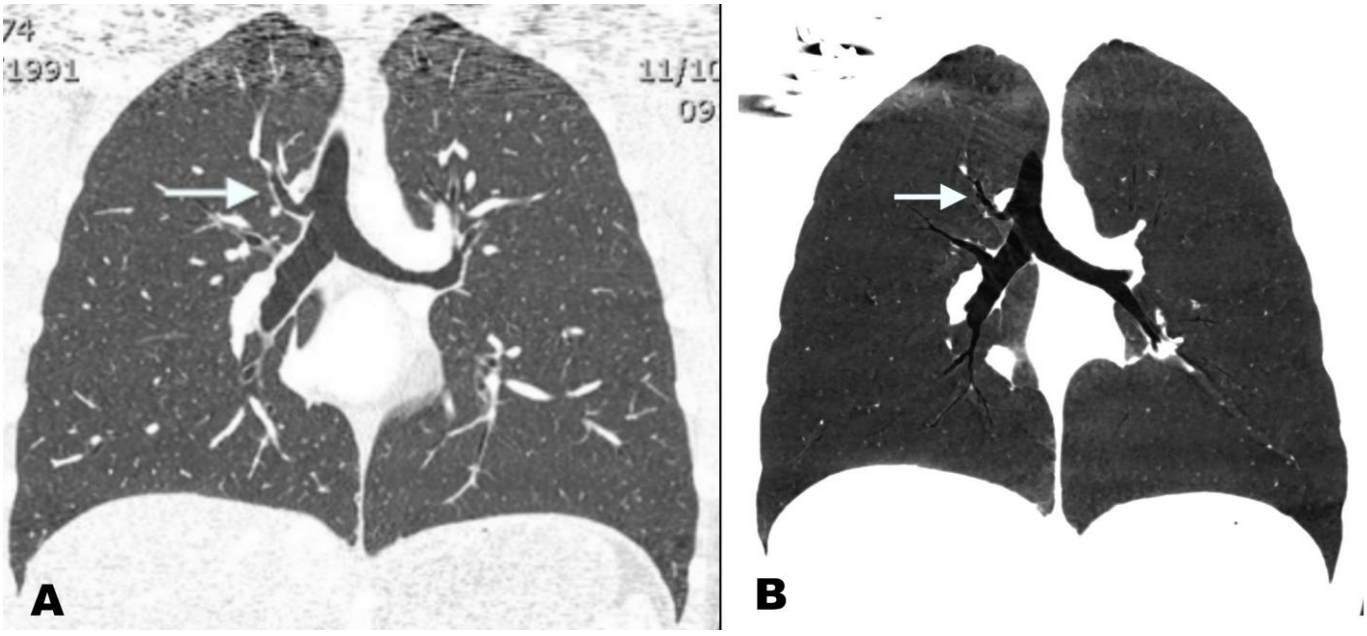


Figure 4: 22 year-old female with systemic arterialization of the lung without sequestration. FINDINGS: Coronal reformatted CT (lung window) (a) and minimum intensity projection (MinIP) (b) reveal an anomalous tracheal bronchus. TECHNIQUE: Aquillion 160 CT scanner (Toshiba, Japan), 200mA, 100kV, 3mm slice thickness, 70 cc Vispaque contrast injected IV at 3cc/second.

'Pulmonary sequestration spectrum' by Sade et al.	**'Pulmonary malinosculation spectrum' by Clements and Warner**		
Independent anatomical variables: 1. A sequestered mass of pulmonary parenchyma may be within or outside the visceral pleura of the ipsilateral normal lung 2. Arterial supply from a systemic or a pulmonary artery, or both 3. Venous drainage to a systemic or a pulmonary vein, or both 4. May or may not have communication with the gastrointestinal tract 5. Normal or abnormal diaphragm	**1. Abnormalities of tracheobronchopulmonary airway and/or arterial blood supply** • Bronchopulmonary malinosculation • Arterial pulmonary malinosculation • Bronchoarterial pulmonary malinosculation	**2. Abnormalities of venous drainage** • Normal • Anomalous • Multiple • Mixed • Mismatched	**3. Lung parenchymal abnormalities** • Intra or extrapulmonary • Cystic, adenomatous, emphysematous, ectopic, foregut inclusions • Abnormal lobation, lobulation, hypoplasia

Table 1: Summary table of classifications by Sade and colleagues, and Clements and Warner

Aetiology	Unknown. The most recognised theory is the persistence of an embryonic connection between the aorta and the pulmonary parenchyma.
Incidence	An estimated incidence of systemic arterial supply to the normal lung has never been published – this is thought to be due to the rarity of this condition.
Gender ratio	Unknown – a review of 12 cases found higher prevalence in males (2:1).
Age predilection	Commonly congenital – a review of 12 cases reported an average age of diagnosis of 21.2 years (range 0-68).
Risk factors	Unknown
Treatment	Lobectomy or segmentectomy; angiographic embolization; systemic artery to pulmonary artery anastomosis
Prognosis	Unknown
Findings on imaging	*Chest X-Ray findings:* a retrocardiac tubular structure; focal area of consolidation. *Thoracic CT findings:* focal area of ground glass density, indicating an area of relative hypervascularity and/or intra-alveolar haemorrhage; may reveal an aberrant systemic artery supplying an area of normal lung parenchyma. *CT angiography findings:* depicts an aberrant systemic artery originating from the aorta supplying a segment of the lower lobe.

Table 2: Summary table of arterialization of the lung without sequestration

Condition	Chest X-Ray	CT	CT angiography	MRI
Systemic arterialization of the lung without sequestration	• focal area of consolidation • a retrocardiac tubular structure • plain radiographs may be normal	• normal lung parenchyma or a focal area of ground glass density indicating an area of relative hypervascularity and/or intra-alveolar haemorrhage • may reveal an aberrant systemic artery supplying an area of normal lung parenchyma	• reveals a normal lung parenchyma or a focal area of ground glass density, indicating relative hypervascularity and/or intra-alveolar haemorrhage • delineates the aberrant systemic artery originating from the aorta • defines a normal venous system	• reveals an anomalous systemic vessel supplying an area of normal lung parenchyma
Pulmonary sequestration	Intralobar sequestration: • opacity or a patch of consolidation typically in the lung base • may show air-fluid levels caused by bronchial communication Extralobar sequestration: • homogeneous mass typically in the pleural space between the left lower lobe and the diaphragm	• homogenous or heterogeneous soft-tissue opacity; may reveal cysts with air-fluid level, and focal emphysema • may demonstrate the aberrant systemic vessel as a linear enhancing structure adjacent to the aorta	• homogenous or heterogeneous soft-tissue opacity; may reveal cysts with air-fluid level, and focal emphysema • delineates the feeding vessel to the sequestration along with its venous system	• differentiates the cystic, solid, haemorrhagic, and mucous components of the mass • defines the course of both the aberrant systemic artery and the venous drainage
Hypogenetic lung syndrome (scimitar syndrome)	• decreased aeration of the right hemithorax with ipsilateral mediastinal shift • may reveal the pulmonary vein descending along the right cardiac border (scimitar sign)	• shows the size of the right hemithorax • may reveal the bronchial anomalies and the anomalous vein	• depicts the anomalous vein and systemic arterial supply to the lower lobe of the right lung from the subdiaphragmatic aorta or its main branches	• shows the anomalous vein draining into the inferior vena cava

Table 3: Differential diagnosis of systemic arterialization of the lung without sequestration

ABBREVIATIONS

CT = Computed Tomography
MRI = Magnetic Resonance Imaging

KEYWORDS

Systemic arterialization of the lung without sequestration; systemic arterial supply; pulmonary artery abnormality; lung; haemoptysis; pulmonary sequestration