

Endovascular treatment of isolated arterial pulmonary malinosculature

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Abstract

We report the endovascular management of a series of four cases of isolated systemic supply to normal lung or isolated arterial pulmonary malinosculature of the left lung. In these cases, the basal segments of the left lung lacked normal pulmonary arterial supply and instead received systemic arterial blood from the descending thoracic aorta. The relevant anatomy and literature are also reviewed.

Key words: Endovascular management; isolated systemic supply to normal lung, pulmonary malinosculature, pulmonary sequestration

Introduction

Systemic arterial supply to the lungs may be congenital or acquired. One of the reported congenital forms of systemic arterial supply to lungs is isolated systemic arterial supply to normal lung (ISSNL), characterised by abrupt tapering of the left lower lobar pulmonary artery distal to the origin of the superior segmental artery and an aberrant systemic artery originating from the descending thoracic aorta supplying the basal segments of the left lower lobe. This manuscript reports a series of four such cases managed by interventional techniques. All patients gave informed consent for the procedure.

Case Reports

Case 1

A 19-year-old girl presented with a history of recurrent hemoptysis for 3 years. The hemoptysis was insidious in

onset with significant blood loss necessitating multiple blood transfusions on different occasions. There was no history to suggest tuberculosis (TB). Clinical examination of the respiratory and cardiovascular systems did not reveal any abnormality.

A contrast-enhanced computed tomography (Philips Brilliance CT 6- slice scanner, CECT) revealed normally aerated lungs with normal bronchial anatomy and absent basal segmental pulmonary arterial branches to the left lower lobe. There was a large systemic artery arising from the left lateral aspect of the descending thoracic aorta coursing through the pulmonary ligament and supplying the basal segments of the left lower lobe [Figure 1A]. Left inferior pulmonary vein drained normally into the left atrium. These findings were consistent with ISSNL, also known as type B isolated arterial pulmonary malinosculature. Since the lung was otherwise normal, endovascular treatment was planned instead of surgical resection.

A digital subtraction angiogram (Siemens ARTIS ZEE Biplane, Ehrlang, Germany and Siemens Multistar DSA machine) was performed using a percutaneous approach through the right common femoral artery, which confirmed the findings of the CT scan [Figure 1B]. A 100 cm, 8F long vascular sheath was placed with its tip in the descending thoracic aorta. The aberrant artery was selectively cannulated and a 16 mm × 12 mm

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Amplatzer Vascular Plug II (9-AVP2-016; St. Jude Medical, Plymouth, Minnesota, USA) was deployed within the distal portion of the aberrant artery. Since there was persistent blood flow in the artery even after 25 min, two embolization coils (NESTER 0.035" pushable coils; COOK, Bloomington, IN47404, USA) measuring 15 mm × 15 cm and 10 mm × 15 cm were deployed proximal to the Amplatzer plug, followed by injection of a small amount of 80% *n*-butyl cyanoacrylate (NBCA) (Histacryl; B Braun, Tuttlingen, Germany), after which there was complete occlusion of the anomalous artery [Figure 1C]. Post-procedural period was uneventful. The patient was discharged on a course of antibiotics and thrice-daily steroids for 10 days. A follow-up telephonic interview 5 months later revealed the patient to be symptom-free.

Case 2

A 32-year-old lady presented with history of intermittent hemoptysis for the past 20 years. She did not require hospital admissions or blood transfusions. She did not have any clinical or laboratory investigation evidence to suggest diabetes mellitus, hypertension, or TB. Her bronchial lavage and transbronchial lung biopsy as part of the TB workup were normal. CECT thorax revealed features consistent with

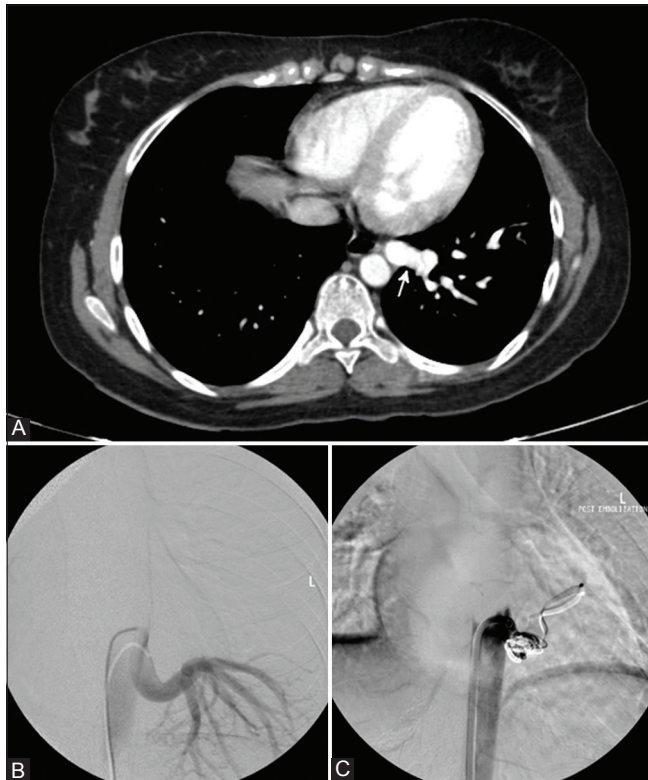


Figure 1: (A) Contrast-enhanced axial CT sections through the lower chest showing anomalous systemic artery (arrow) arising from the aorta (origin not shown) and supplying the left lower lobe. (B) Frontal projection angiogram showing the large systemic artery supplying the basal segments of the left lower lobe. (C) Check angiogram showing complete occlusion of the aberrant artery following deployment of the Amplatzer plug and additional embolization with coils and glue

[Figure 2A] ISSNNL or type B isolated arterial pulmonary malinosculcation.

A digital subtraction angiogram confirmed the findings [Figure 2B]. A 60 cm, 7F vascular sheath was placed with its tip in the descending thoracic aorta. The aberrant artery was selectively cannulated with a glide SIM-1 catheter and tip of catheter was placed in the distal portion of the aberrant vessel. Two 12-mm detachable balloons (Goldbalt-5) were deployed [Figure 2C]. Post-deployment angiogram revealed a small stump with opacification of a segmental branch of the basal lung. There was distal occlusion of the artery. The post-procedural period was uneventful and the patient was discharged on a course of antibiotics. She was symptom-free for 6 months but later presented with recurrent hemoptysis. CECT revealed complete recanalization of the aberrant artery [Figure 2D]. A repeat endovascular management with Amplatzer plug II was planned but the patient did not return for follow-up.

Case 3

A 27-year-old man presented with recurrent episodes of hemoptysis for 2 years. CECT revealed features suggestive of [Figure 3A] ISSNNL or type B isolated arterial pulmonary malinosculcation.

A digital subtraction angiogram was performed. A 7F-long sheath (Arrow, Bernville road, PA, USA) was placed into the aberrant artery [Figure 3B], through which a vertebral glide

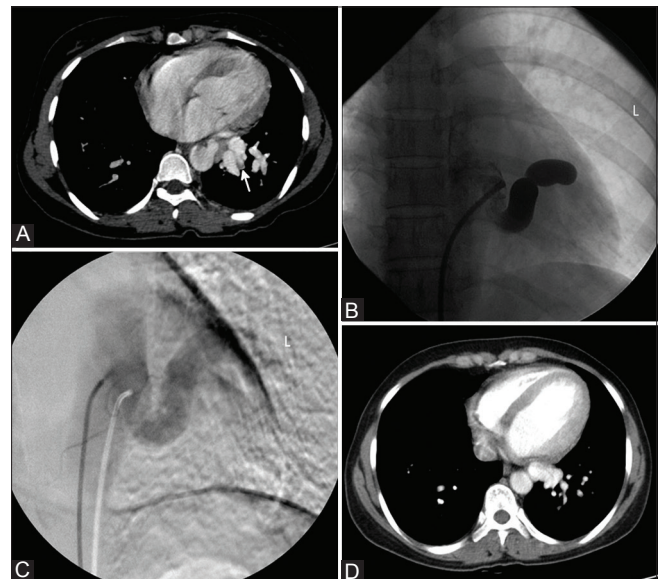


Figure 2: (A) Contrast-enhanced axial CT sections through the lower chest showing anomalous systemic artery (arrow) arising from the aorta and supplying the left lower lobe. (B) Selective angiogram of the aberrant artery. (C) Check angiogram showing complete occlusion of the aberrant artery following deployment of two detachable balloons. (D) Contrast-enhanced axial CT sections through the lower chest, seven months post embolization using detachable balloon, showing recanalization of the anomalous systemic artery

catheter was passed and placed in the distal aspect. Three embolization coils (NESTER 0.035" pushable coils; COOK, Bloomington, IN47404, USA) measuring 10 mm × 14 cm were deployed proximally [Figure 3C] and a check angiogram was performed which showed complete occlusion of the aberrant artery. Post-procedural period was uneventful. The patient was discharged and antibiotics were prescribed for a week. A follow-up telephonic interview conducted 1 year later revealed that the patient was symptom-free for 10 months after which he had one episode of minimal hemoptysis not requiring intervention. Repeat CT has not yet been performed.

Case 4

A 15-year-old boy presented with recurrent hemoptysis for 3 months. He did not require any hospital admission or blood transfusion. Clinical examination of his respiratory and cardiovascular systems was normal. CECT revealed [Figure 4A] ISSNNL or type B isolated arterial pulmonary malinosculcation.

A digital subtraction angiogram was performed [Figure 4B]. A 6F Ansel sheath was placed into the aberrant artery from the descending thoracic aorta and a 12 mm × 9 mm Amplatzer Vascular Plug II (9-AVP2-016) was deployed into the distal portion of the aberrant artery. A check angiogram, 25 minutes later, showed complete occlusion of the aberrant artery [Figure 4C]. Post-procedural period was uneventful. The patient was discharged and antibiotics were prescribed for a week. A follow-up CECT showed areas of collapse in the anterior and lateral basal segments of the left lower lobe

with non-opacification of the aberrant artery. Patient was symptom-free at 6 months follow-up. Table 1 summarizes the management of our patients and the various types of pulmonary malinosculcations with proposed standard strategies.

Discussion

The concept of "sequestration" was first described by Pryce in 1946. However, this terminology could not define all variants of sequestration and led to the concept of a "sequestration spectrum" described in 1974.^[1,2] This concept was reviewed in 1984 and the importance of the venous drainage was discussed, leading to the "wheel theory" by Clements, who coined the term "pulmonary malinosculcation."^[3,4] In 2007, Lee proposed the "haphazard theory" to substantiate this concept and described seven types of pulmonary malinosculcation,^[1,3] based on whether the abnormal communication involved the bronchi, arteries of the lungs or veins of the lungs in isolation or in different combinations.

In all cases described in our series, there was systemic arterialization of an otherwise normal lung with normal bronchial tree, normal venous drainage, and absent pulmonary artery branches, suggestive of a type B or isolated arterial malinosculcation. They could also be classified as a Pryce type 1 anomaly. These cases can be managed either surgically or endovascularly. Different surgeries have been reported, including ligation of the anomalous artery and lobectomy, surgical ligation of

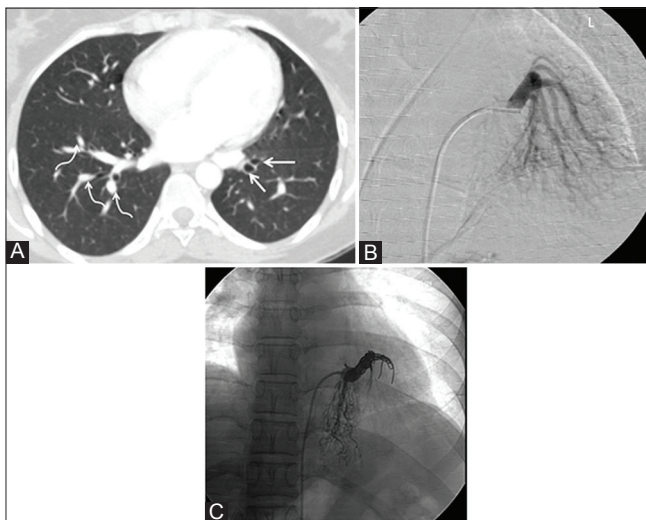


Figure 3: (A) Axial CT sections through the lower chest, in lung window. Note the absence of normal basal segmental pulmonary artery branches accompanying the segmental bronchi in the left lower lobe (arrows). Compare with the normal segmental pulmonary arteries coursing along the bronchi in the right lung lower lobe (curved arrows). (B) Selective angiogram of the aberrant artery showing its branches supplying the basal segments of the left lower lobe. (C) Check angiogram showing complete occlusion of the aberrant artery following deployment of the pushable coils

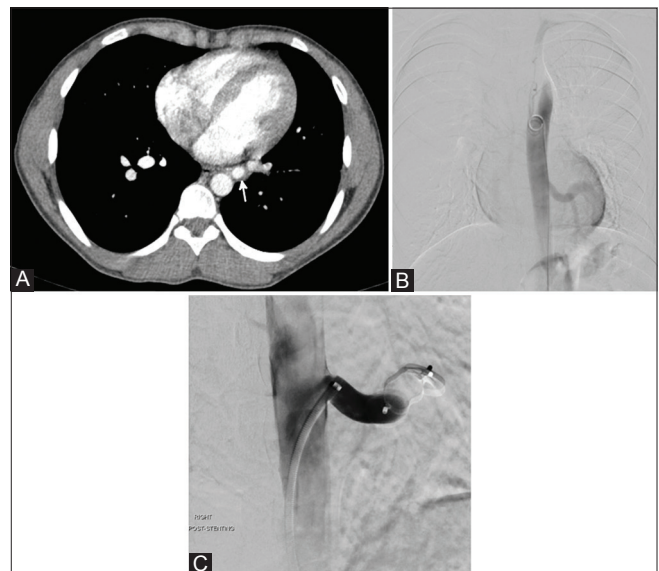


Figure 4: (A) Contrast-enhanced axial CT sections through the lower chest showing anomalous systemic artery (arrow) arising from the aorta and supplying the left lower lobe. (B) Flush aortogram showing the anomalous systemic artery arising from the aorta and supplying the left lower lobe. (C) Check angiogram showing occlusion of the aberrant artery following deployment of the Amplatzer plug, leaving behind an arterial stump

Table 1: Management of the patients summarized

Gender/age	Clinical manifestation	Side	Origin	No. of arteries	Arterial perfusion to the lower lobe	PA perfusion to the lower lobe	Drainage	Procedure
19/F	Hemoptysis	L	DTA	1	Basal segment		IPV	Amplatzer plug II+ Coils+ 80% glue
32/F	Hemoptysis	L	DTA	1	Basal segment		IPV	Detachable balloons
27/M	Hemoptysis	L	DTA	1	Basal segment		IPV	Coils
15/M	Hemoptysis	L	DTA	1	Basal segment		IPV	Amplatzer plug II

DTA: Descending thoracic aorta, IPV: Inferior pulmonary vein, PA: Pulmonary artery

Type	Abnormal communication	Typical examples	Treatment options
A	Isolated bronchial pulmonary malinosculcation	Tracheal and bronchial abnormalities including bronchial atresia, bronchogenic cysts, cystic pulmonary airway malformations	Usually surgical
B	Isolated arterial pulmonary malinosculcation	Isolated systemic supply to normal lung (pulmonary arterial supply is absent and lung is otherwise normal)	Surgical or endovascular
C	Isolated venous pulmonary malinosculcation	Anomalous pulmonary venous drainages	If shunt is large, may require surgical correction
D	Mixed broncho-arterial pulmonary malinosculcation	Typical intralobar sequestration	Usually surgical as the involved lung would be abnormal
E	Mixed broncho-venous pulmonary malinosculcation	Any type A lesions with associated venous anomalies	Usually surgical
F	Mixed arterio-venous pulmonary malinosculcation	Arterio-venous malformations and fistulas	Usually endovascular
G	Mixed bronchoarteriovenous pulmonary malinosculcation	Typical extralobar sequestration	Usually surgical as the involved lung would be abnormal

the anomalous artery only without lobectomy, and disconnection of the anomalous artery from the aorta and anastomosing it to the pulmonary artery.

Though embolization has been reported to be a safe alternative to surgery in pediatric patients and in severe hemoptysis, surgery still remains the gold standard for severe forms of malinosculcation.^[5] Metallic coils were the preferred embolic materials in the previously reported cases.^[6-10] Most of the times, multiple coils were required. Using NBCA glue along with multiple coils to form a compact mass of coils has also been reported.^[11]

Proposed problems with endovascular treatment include incomplete embolization leading to collateral feeders,^[5] migration of embolization material resulting in embolization of nontarget areas, and development of lung infarction. None of the previously reported 11 cases of endovascular treatment developed any major complications and none of them required surgery.^[6,8-10,12] Mild chest pain after the procedure has been reported frequently.

Lung necrosis has been reported in one patient who was surgically treated with ligation of the anomalous artery alone, without lobectomy, necessitating a re-surgery and lobectomy.

The Amplatzer vascular plug (AVP) has replaced metallic coils, especially in situations where coil migration is possible, like in large, short, and high-flow vessels. The size of the selected device should be 40% larger than the internal diameter of the main trunk of the anomalous

systemic artery. Recanalization has been reported in 7% cases where AVP has been used for pulmonary arteriovenous malformations and, hence, requires long-term follow-up.^[6]

In the first case in our series, we used the Amplatzer Vascular Plug II of size 40% more than the anomalous systemic artery, as recommended. However, since the check angiogram at 25 min showed incomplete occlusion, we decided to deploy a few embolization coils into this artery. Since some blood flow still persisted, a small amount of nBCA glue was injected which resulted in successful complete embolization.

None of our four patients developed any complications such as lung gangrene or infection. One of the patients developed sub-segmental atelectasis, which could be related to ischemia or infarction. However, the patient remained asymptomatic for the same.

Conclusion

Endovascular management is a suitable treatment option in symptomatic patients with ISSNL. Different methods of blocking the anomalous systemic artery, such as detachable balloons, Amplatzer plug, and coils, have been tried. As there are only isolated case reports and case series, there is no consensus on the ideal embolic material.

In our limited experience in treating these types of cases, we suggest that it would be better to avoid detachable balloons. Amplatzer vascular plugs and metallic coils are more preferred embolization materials.

Embolization of the anomalous systemic artery is safe, though it may be the sole arterial supply to an otherwise normal lung, without the risk of any major complications as seen in our patients as well as in a majority of the isolated case reports and case series reported in literature.

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