Cardiology in the Young

cambridge.org/cty

Isolated systemic arterial supply to normal lung – an unusual cause of extracardiac left-to-right shunt

Balaji Arvind, Saurabh Kumar Gupta and Shyam Sunder Kothari

Department of Cardiology, All India Institute of Medical Sciences, New Delhi, India

Brief Report

Cite this article: Arvind B, Gupta SK, and Kothari SS (2020) Isolated systemic arterial supply to normal lung – an unusual cause of extracardiac left-to-right shunt. *Cardiology in the Young* **30**: 1946–1950. doi: 10.1017/S1047951120002851

Received: 13 July 2020
Revised: 29 July 2020
Accepted: 19 August 2020
First published online: 18 September 2020

Keywords:
Isolated systemic arterial supply to normal lung; bronchopulmonary vascular malformation; left-to-right shunt; transcatheter closure

Author for correspondence:
Dr Saurabh Kumar Gupta, Additional Professor of Cardiology, Room No. 9, 8th floor, Department of Cardiology, Cardio-Thoracic Centre, All India Institute of Medical Sciences, New Delhi 110029, India.
Tel: +91-011-26594944; Fax: + 91-11-26588663.
E-mail: drsaurabhmd@gmail.com

Abstract

Isolated systemic arterial supply to a normal lung, a type of bronchopulmonary vascular malformation, is a rare cause of extracardiac left-to-right shunt. We describe such a case that was successfully managed by transcatheter closure of the anomalous arterial supply to otherwise normal lung.

Introduction

While left-to-right shunts commonly occur within the heart or between proximal segments of great arteries, the shunts can also occur within the lungs secondary to bronchopulmonary vascular malformation. In most such cases, the lungs are abnormal, with a sequestrated lung segment receiving blood supply from the aorta. Rarely, the only abnormality is an anomalous systemic arterial supply to an otherwise normal lung. This is termed as an isolated systemic arterial supply to normal lung.[1] In this report, we describe one such case with a significant left-to-right shunt in a young female, which was successfully managed by transcatheter closure of the anomalous arterial supply.

Case presentation

A 21-year-old female presented with exertional palpitations of 3 years duration. She had cardiomegaly and a grade 3/6 continuous murmur in the left infrascapular area. The chest radiograph showed cardiomegaly and a retrocardiac shadow in the left lower lobe of the lung. Echocardiography revealed a dilated left atrium and left ventricle with normal left ventricular contractile function. There was no intracardiac defect, aortopulmonary window or patent arterial duct. Doppler interrogation of descending thoracic aorta revealed an abnormal continuous flow towards the left lung. Computed tomographic angiography confirmed the presence of an anomalous tortuous artery originating from descending thoracic aorta at the level of eighth thoracic vertebra. The anomalous artery supplied all the basal segments of the lower lobe of the left lung (Fig 1) and measured 13.5 mm at its widest diameter. The parenchyma in the affected lung segments was normal with usual blood supply from the left pulmonary artery and drainage through a dilated left inferior pulmonary vein into the left atrium. In view of the large left-to-right shunt, normal lung parenchyma, and an unequivocal supply from the pulmonary artery to the affected segments, transcatheter closure of the anomalous systemic artery was planned.

Cardiac catheterisation confirmed increased pulmonary blood flow with a pulmonary to systemic blood flow ratio of 2:1. The pulmonary artery systolic and mean pressure measured 26 mmHg and 19 mmHg, respectively. Considering the downward orientation of the orifice of the vascular channel, left brachial access was chosen. Following an aortogram, the vascular channel was engaged with a 5 Fr Judkins right coronary catheter with the help of a 0.035-inch angled tip hydrophilic guidewire (Terumo Co, Tokyo, Japan). The catheter was then exchanged with an 8-French Mullins sheath (Cook Medical, Bloomington, Indiana, United States of America) over an 0.035-inch exchange length extra-stiff guidewire. A single-lobed 18 mm Cera vascular plug (Lifetech Scientific Co. Ltd., Shenzhen, China) was deployed approximately

CAMBRIDGE UNIVERSITY PRESS

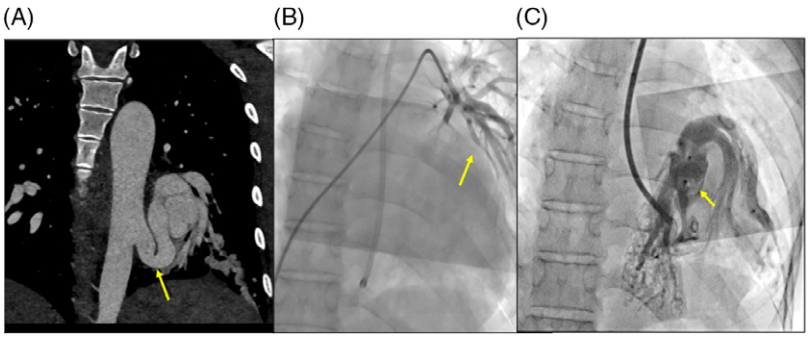


Figure 1. Panel A: CT angiogram in coronal section shows the anomalous artery (arrow) arising from descending thoracic aorta at the level of eighth thoracic vertebra and supplying all the basal segments of the left lower lobe. Panel B: Catheter angiogram in left pulmonary artery confirming intact pulmonary arterial supply to the corresponding lung segments (arrow). Panel C: Catheter angiogram soon after occlusion of the anomalous artery using vascular plug shows a well-positioned device in situ and mild residual flow through the device.

15 mm from the aortic end. The post-deployment angiogram showed minimal residual flow through the device (Fig 1C). She was discharged the next day. At a 3-month follow-up, she was asymptomatic with a normal-sized left ventricle and no residual flow across the anomalous vascular channel.

Discussion

The bronchopulmonary vascular system comprises of bronchial, pulmonary arterial and pulmonary venous systems. Bronchopulmonary vascular malformations include any disruption in the normal communication and/or formation of abnormal communications between one or more of these three components. The spectrum of malformations ranges from simple bronchogenic cyst to the scimitar syndrome. The term "malinosculation" is used to describe the presence of an abnormal communication between the various components of the bronchopulmonary vascular system.[2] Lee and colleagues, based on the extent of involvement of various components, classified these malformations into seven types. (Table 1).[3]

Isolated systemic arterial supply to normal lung is a congenital malformation wherein an anomalous artery from the systemic circulation supplies an otherwise normal lung segment (type B bronchopulmonary vascular malformation) and represents an isolated arterial malinosculation (Table 1).[3] The anomalous artery mostly originates from the descending aorta, usually between the eighth and eleventh thoracic vertebra, and supplies the lower lobe of left lung. Rarely, the anomalous artery may originate from the celiac trunk, subclavian or internal mammary arteries.[4] In the complete variant, there is no alternative blood supply whereas in the incomplete type, the affected lung segment has intact vascular supply from the native pulmonary arteries.[5] Most often, the patients are asymptomatic while others present with recurrent hemoptysis. In some patients as in this case, a relatively lower resistance in the lung vasculature permits intrapulmonary left-to-right shunt resulting in left ventricular volume overload.

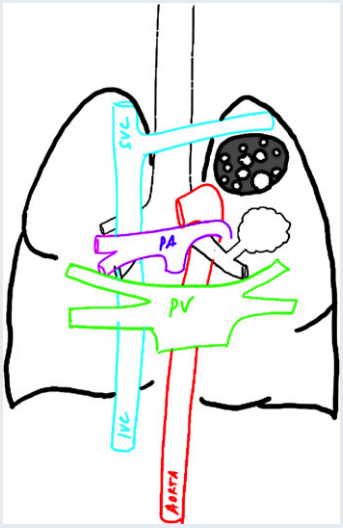
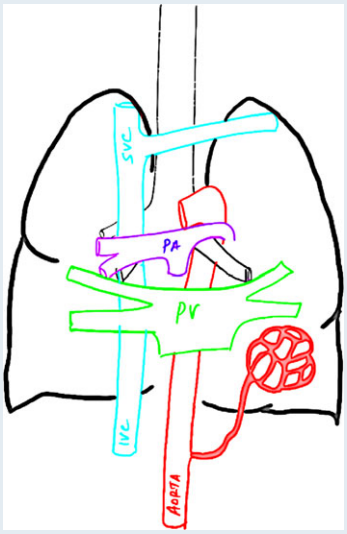
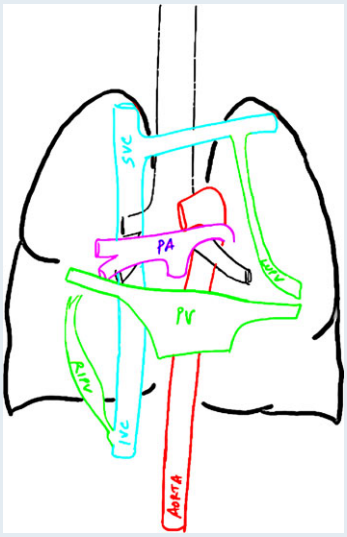
In the absence of any other abnormality, the shunt can be easily abolished by occlusion of the anomalous arterial supply. Intuitively, occlusion of the artery is safer in the incomplete type although, despite concerns, safe occlusion or ligation has been reported in the complete type as well.[6–9] Nevertheless, occlusion of the sole supply to the affected lung segment in the complete type converts the lung segment non-functional with subsequent risk of infection and bronchiectasis. Besides, recanalisation of the arterial channel as well as recruitment of newer bronchial collaterals is known, thus mandating careful follow-up after occlusion of the anomalous artery.[9,10] Incomplete type of systemic supply to normal lung in our patient made it safer for interventional closure.

Conclusion

Isolated systemic arterial supply to normal lung, a type of bronchopulmonary vascular malformation, is an extraordinary cause of extracardiac left-to-right shunt. An unusual location of shunt poses challenges during evaluation. However, being extracardiac, this isolated arterial malinosculation is an easy and safe target for transcatheter closure.

https://doi.org/10.1017/S1047951120002851 Published online by Cambridge University Press

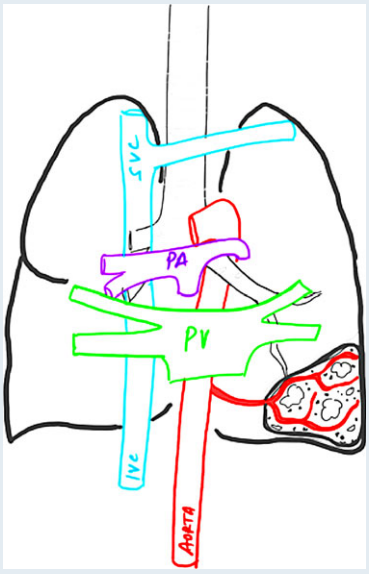
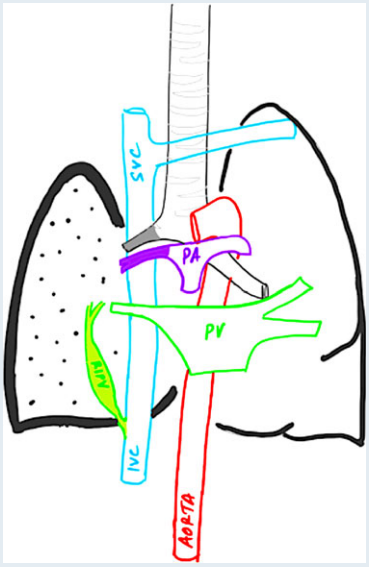
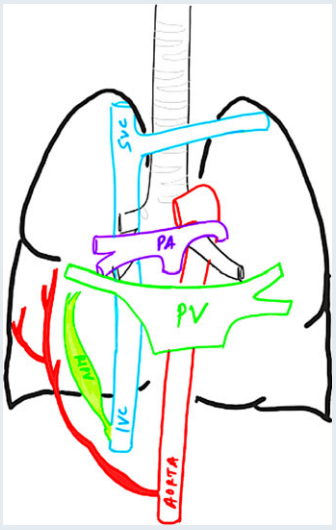
Table 1. Schematic representation of bronchopulmonary vascular malformations with examples

Type	Mechanism	Schematic diagram	Examples
A	Isolated bronchial malinosculation		1. Bronchogenic cyst. 2. Congenital lobar emphysema. 3. Congenital cystic adenomatoid malformation.
B	Isolated arterial malinsoculation		1. Isolated systemic supply to normal lung. 2. Anomalous origin of branch pulmonary artery. 3. Pulmonary artery sling.
C	Isolated venous malinosculation		1. Partial or total anomalous pulmonary venous connections. 2. Isolated scimitar vein.

(*Continued*)

https://doi.org/10.1017/S1047951120002851 Published online by Cambridge University Press

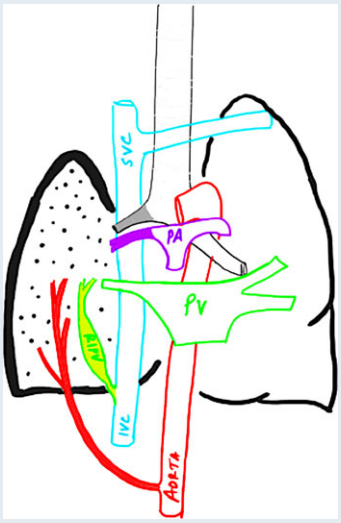
Table 1. (*Continued*)

Type	Mechanism	Schematic diagram	Examples
D	Mixed broncho-arterial malinosculation		1. Intralobar sequestration.
E	Mixed broncho-venous malinosculation		1. Scimitar vein associated with lung hypoplasia.
F	Mixed arterio-venous malinosculation		1. Scimitar vein associated with systemic to pulmonary collateral.

(*Continued*)

https://doi.org/10.1017/S1047951120002851 Published online by Cambridge University Press

Table 1. (*Continued*)

Type	Mechanism	Schematic diagram	Examples
G	Mixed broncho-arterio-venous malinosculation		1. Classic scimitar syndrome. 2. Extralobar sequestration.

IVC=inferior caval vein; LUPV=left upper pulmonary vein; PA=pulmonary artery; PV=pulmonary veins; RIPV=right inferior pulmonary vein; SVC=superior caval vein.

Acknowledgements. The authors acknowledge help received from Dr Priya Jagia, Professor, Department of Cardiovascular Imaging and Endovascular Interventions, AIIMS, New Delhi in the interpretation of CT angiogram.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflict of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards as mentioned in the Helsinki Declaration of 1975, as revised in 2008.

References

1. Flisak M, Chandrasekar A, Marsan R, et al. Systemic arterialization of lung without sequestration. Am J Roentgenol 1982; 138, 751–753.
2. Clements BS, Warner JO. Pulmonary sequestration and related congenital bronchopulmonary-vascular malformations: nomenclature and classification based on anatomical and embryological considerations. Thorax 1987; 42, 401–408.
3. Lee M-L, Lue H-C, Chiu I-S, et al. A systematic classification of the congenital bronchopulmonary vascular malformations: dysmorphogeneses of the primitive foregut system and the primitive aortic arch system. Yonsei Med J 2008; 49, 90–102.
4. Miyake H, Hori Y, Takeoka H, et al. Systemic arterial supply to normal basal segments of the left lung: characteristic features on chest radiography and CT. Am J Roentgenol 1998; 171, 387–392.
5. Yamanaka A, Hirai T, Fujimoto T, et al. Anomalous systemic arterial supply to normal basal segments of the left lower lobe. Ann Thorac Surg 1999; 68, 332–338.
6. Izzillo R, El Hajjam M, Qanadli S, et al. Intralobar sequestration of the lung in an adult (type 1 of pryce). Treatment by coil embolization. J Radiol 2000; 81, 996–999.
7. Canyigit M, Gumus M, Kilic E, et al. Aneurysm of an Anomalous Systemic Artery Supplying the Normal Basal Segments of the Left Lower Lobe: Endovascular Treatment with the Amplatzer Vascular Plug II and Coils. Cardiovasc Intervent Radiol. 2011; 34, 126–130.
8. Mammen S, Keshava SN, Moses V, Chiramel GK, Irodi A, Gnanamuthu BR. Endovascular treatment of isolated arterial pulmonary malinosculation. Indian J Radiol Imaging. 2015; 25, 239–243.
9. Jiang S, Shi J-Y, Zhu X-H, et al. Endovascular embolization of the complete type of anomalous systemic arterial supply to normal basal lung segments: a report of four cases and literature review. Chest 2011; 139, 1506–1513.
10. Abe T, Mori K, Shiigai M, et al. Systemic arterial supply to the normal basal segments of the left lower lobe of the lung—treatment by coil embolization—and a literature review. Cardiovasc Interv Radiol 2011; 34, 117–121.

https://doi.org/10.1017/S1047951120002851 Published online by Cambridge University Press