

## Case Report

# Embolization of a Bronchial Artery–Pulmonary Artery Arterio–Venous Malformation

Uei Pua, MBBS, MMed, FRCR, FAMS,<sup>1,2</sup> Lawrence Han Hwee Quek, BMBS (Flinders), FRCR, MMED, FRANZCR, FAMS,<sup>1,3</sup> and Aneez Dokev Basheer Ahmed, MBBS, FRCS General Surgery (Glas), FRCS Cardiothoracic Surgery, FAMS<sup>4</sup>

Bronchial artery–pulmonary artery arterio–venous malformation, previously known as “bronchial artery primary racemose hemangioma” is a rare vascular malformation. The literature surrounding the imaging findings and treatment is currently limited. We hereby describe the imaging findings and a novel endovascular approach of utilizing bi-directional embolization from both the pulmonary and bronchial artery to occlude the malformation, leading to thrombosis and resolution of symptoms. This strategy represents a potential treatment option of non-surgical patients in this rare malformation.

**Keywords:** bronchial artery, pulmonary artery, arterio–venous malformation

## Introduction

Bronchial artery–pulmonary artery arterio–venous malformation (BP-AVM), previously known as “bronchial artery primary racemose hemangioma” is a rare vascular malformation. The literature surrounding the imaging

findings and treatment is currently limited. We hereby describe the imaging appearance and endovascular treatment of such a case.

## Case Report

A 63-year-old man presented with exertional dyspnea in the background of normal cardiac evaluation, and computer tomography (CT) findings of a vascular malformation in the posterior mediastinum (Fig. 1). The malformation was supplied by 4 arteries (right bronchial artery and three intercostals) arising from the thoracic aorta with a large vascular mass in the posterior mediastinum and single drainage vessel into the right upper lobe pulmonary artery (Fig. 2). The appearance was consistent with a BP-AVM.<sup>1)</sup>

In view of the size and the risk of hemoptysis, the patient was referred for embolization. Through a left common femoral vein access, the right upper lobe pulmonary was accessed. The drainage vessel arising the superior segmental artery of the right upper lobe was first cannulated. However, due to the angulation of the artery with sheath instability, a tri-axial system was needed to allow deep catheterization required for vascular plug embolization (Fig. 3). This consisted of an 8.5F Agilis NxT (St Jude Medical, Minnetonka, MN, USA) steerable sheath posi-


<sup>1</sup>Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore

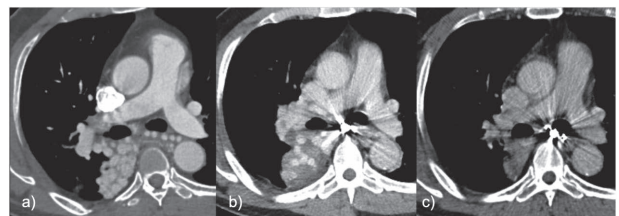
<sup>2</sup>Yong Loo Lin School of Medicine, National University of Singapore, Singapore

<sup>3</sup>Lee Kong Chian School of Medicine, Nanyang Technological University, Singapore

<sup>4</sup>Department of General Surgery, Tan Tock Seng Hospital, Singapore

Received: September 25, 2018; Accepted: March 12, 2019  
Corresponding author: Uei Pua, MBBS, MMed, FRCR, FAMS.  
Department of Diagnostic Radiology, Tan Tock Seng Hospital,  
11 Jalan Tan Tock Seng, Singapore 308433  
Tel: +65-8181-9490, Fax: +65-6759-9940  
E-mail: druei@yahoo.com

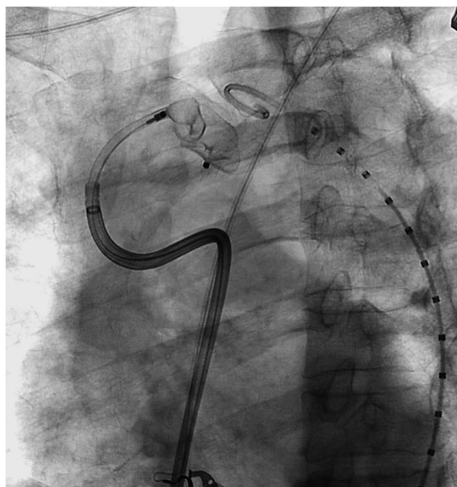
 ©2019 The Editorial Committee of Annals of Vascular Diseases. This article is distributed under the terms of the Creative Commons Attribution License, which permits use, distribution, and reproduction in any medium, provided the credit of the original work, a link to the license, and indication of any change are properly given, and the original work is not used for commercial purposes. Remixed or transformed contributions must be distributed under the same license as the original.



**Fig. 1** Axial computer tomography image obtained at the level of the pulmonary trunk of the large vascular malformation in the posterior aspect of the right hemithorax. (a) pre- (b) 1-day post- (c) 6 months. Significant thrombosis could be seen in the immediate post-embolization period (b) with complete thrombosis and reduction in size by 6 months (c).

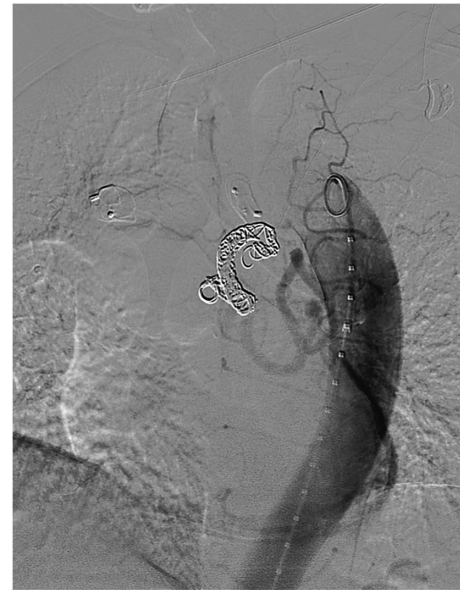


**Fig. 2** Composite image of the aortogram showing multiple feeding artery arising from the aorta into the vascular malformation. The drainage pulmonary artery could not be demonstrated. The direction of flow confirmed a left-to-right shunt physiology.



**Fig. 3** Fluoroscopic images of the drainage pulmonary artery which was accessed using a tri-axial system consisting of a deflectable sheath, guiding catheter and diagnostic catheter to obtain a position for deployment of an Amplatzer vascular plug II. A pigtail catheter could be seen in the aorta.

tioned in the ostium of the segmental artery, followed by an 8F Mach 1 guiding catheter (Boston Scientific, Natick, MA, USA) and a 5F 125 cm long Ultimate 1 diagnostic catheter (Merit Medical Systems, Inc., South Jordan, UT, USA) inserted co-axially. In a telescoping fashion, the guiding catheter was advanced over the diagnostic catheter and guidewire to reach a stable position within the segmental pulmonary artery, and a 16 mm Amplatzer AVP II vascular plug (St Jude Medical) was deployed, occluding the solitary drainage pulmonary artery (Fig. 3).



**Fig. 4** Completion aortogram showed near cessation of flow through the malformation and the two smaller feeding arteries that were not embolized. Nevertheless, together with embolization of the outflow pulmonary artery, complete thrombosis of the vascular malformation was achieved.

Through a right common femoral artery access, aortography demonstrated the 4 feeding arteries as seen on CT (Fig. 2). The plan was for staged coil/plug embolization (without particulate embolics) of the feeding arteries (2 arteries in a single sitting) due to the difficulty in resolving any potential spinal supply. If needed, we would return for an interval embolization of the remaining two vessels. The largest supplying artery (intercostal artery) was first cannulated and a 6F Brite-tip sheath (Cordis, Miami, FL, USA) was used to deliver a 12 mm AVP II plug successfully deployed (Fig. 4). The hypertrophied right bronchial artery was then accessed using a Progreat microcatheter and embolized using a total of 8 Interlock coils ranging from 5 to 12 mm in size (Boston Scientific) (Fig. 4). Completion angiography showed complete embolization of the two vessels with significant slow flow into the malformation through the remaining two untreated arteries (Fig. 4).

The immediate post-embolization CT at day 1 showed significant thrombosis of the BP-AVM (Fig. 1b) and the patient was discharged well. At 6 months follow-up CT, complete thrombosis with a significant reduction in size of the BP-AVM was demonstrated (Fig. 1c). The patient reported improvement in effort tolerance from previous modified Medical Research Council score (mMRC)<sup>1)</sup> of 3 to 1 over the same period and remained so at 1 year.

## Discussion

BP-AVM is an extremely rare vascular malformation with

limited cases described in the English literature.<sup>3–5)</sup> The largest published series is in the Japanese literature and summarized by Narato et al.<sup>2)</sup> and included 34 cases in the Japanese literature with hemoptysis reported in 30 of the cases, a right-sided predominance (25 cases) and no gender predilection. Broadly, BP-AVMs are classified as primary/congenital or secondary related to pulmonary infection/inflammation, trauma or malignancy. Our case was considered a primary BP-AVM in view of the absence of risk factors.

We classified our case as an AVM, although the malformation was between two arterial systems (bronchial and pulmonary), strictly without a venous component. The rationale being that the underlying pathophysiology is similar. Namely; a high pressure and high velocity system (systemic) draining into a low pressure and low velocity system (pulmonary artery), which would be considered an AVM under the ISSVA classification.<sup>5)</sup> This distinction allowed us to treat the lesion conceptually similar to single venous outflow AVM in other areas (e.g., in dural AVM), in which closure of the venous end (pulmonary artery) is more crucial than the arterial inflow.

Surgery (lobectomy) remains the main treatment BP-AVM with bronchial artery embolization being described as a potential alternative.<sup>3,4)</sup> From the perspective of embolotherapy, BP-AVM differs from the commonly seen pulmonary AVM. BP-AVM represents a systemic artery to pulmonary artery shunt, that is; a left to right shunt versus a right to left shunt, and therefore has a much higher shunt speed. Within the literature, success with bronchial artery embolization in BP-AVM remain mixed due to delayed vascular recanalization and concern for pulmonary embolism in the high-flow condition.<sup>3,4)</sup> More importantly, the authors in the described cases performed solely arterial side embolization without embolization of the outflow. Based on this, we chose to first embolize the outflow pulmonary artery to reduce the shunting rate (left-to-right shunt physiology) which not only reduces the risk of pulmonary embolism but also makes subsequent arterial side embolization easier. After obtaining outflow control, we performed partial inflow embolization (2 out of 4 arterial feeders) in a rather proximal segment of the feeders. Proximal embolization while technically easier poses the potential of the BP-AVM recruiting new collateral inflow. However, we believed that embolization of the solitary outflow would result in significant stasis in the malformation and mitigate the risk of collateral revascularization. And indeed, this strategy resulted in complete thrombosis of the malformation.

Our “bi-directional” embolization approach deviates from the described “bronchial artery embolization only approach” and is further supported by another case in the literature which pulmonary artery embolization (outflow)

was combined with thoracic endografting (inflow) in a case of BP-AVM.<sup>4)</sup>

While the most common presentation of BP-AVM is that of hemoptysis,<sup>2–4)</sup> our case presented with exertional dyspnea which markedly improved following embolization. We are not fully certain of the etiology but postulate that it could be due to resolution of pulmonary hypertension and congestion with embolization of the AVM. Future cases will benefit from pulmonary arterial pressure measure prior and after embolization to confirm this pathophysiology.

## Conclusion

Bidirectional pulmonary arterial and systemic artery embolization is a possible alternative to surgical resection in the treatment of this rare vascular malformation.

## Disclosure Statement

The authors declare no conflict of interest in the preparation of this manuscript.

## Author Contributions

Study conception: UP

Data collection: UP

Analysis: UP

Investigation: all authors

Writing: UP

Funding acquisition: N/A

Critical review and revision: all authors

Final approval of the article: all authors

Accountability for all aspects of the work: all authors

## References

- 1) Mahler DA, Wells CK. Evaluation of clinical methods for rating dyspnea. *Chest* 1988; **93**: 580-6.
- 2) Narato R, Enomoto T, Ono H, et al. A case of successful bronchial artery embolization for primary racemose hemangioma with massive hemoptysis. *Nihon Kokyuki Gakkai Zasshi* 2006; **44**: 641-6. (in Japanese)
- 3) Iwasaki M, Kobayashi H, Nomoto T, et al. Primary racemose hemangioma of the bronchial artery. *Intern Med* 2001; **40**: 650-3.
- 4) Matsubara Y, Morisaki K, Matsuda D, et al. Thoracic endovascular aortic repair and coil embolization of the pulmonary artery for primary racemose hemangioma of the bronchial artery with a bronchial-pulmonary artery fistula. *Vascular* 2015; **23**: 436-9.
- 5) ISSVA Classification of Vascular Anomalies ©2014 International Society for the Study of Vascular Anomalies. Available at [issva.org/classification](http://issva.org/classification) (Accessed: 29/10/2017)