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

Uei Pua, MBBS, MMed, FRCR, FAMS,1,2 Lawrence Han Hwee Quek, BMBS (Flinders), FRCR, MMED, FRANZCR, FAMS,1,3 and Aneez Dokev Basheer Ahmed, MBBS, FRCS General Surgery (Glas), FRCS Cardiothoracic Surgery, FAMS4

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.1

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...
tioned in the ostium of the segmental artery, followed by an 8F Mach 1 guiding catheter (Boston Scientific, Natick, MA, USA) and a 5F 12.5 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).

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) of 3 to 1 over the same period and remained so at 1 year.

Discussion
BP-AVM is an extremely rare vascular malformation
limited cases described in the English literature.\textsuperscript{3–5} The largest published series is in the Japanese literature and summarized by Narato et al.\textsuperscript{2} 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-AMVs are classified as primary/congenital or secondary related to pulmonary infection/inflammation, trauma or malignancy. Our case was considered a primary BP-AMV 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.\textsuperscript{5} 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-AMV with bronchial artery embolization being described as a potential alternative.\textsuperscript{3,4} From the perspective of embolotherapy, BP-AMV differs from the commonly seen pulmonary AVM. BP-AMV 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-AMV remain mixed due to delayed vascular recanalization and concern for pulmonary embolism in the high-flow condition.\textsuperscript{3,4} 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-AMV 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-AMV.\textsuperscript{4}

While the most common presentation of BP-AMV is that of hemoptysis,\textsuperscript{2–4} 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
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Critical review and revision: all authors
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Accountability for all aspects of the work: all authors

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