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Particle embolization for hemoptysis in two patients with cyanotic congenital heart disease☆

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ABSTRACT

Hemoptysis is a known manifestation of complex cyanotic congenital heart disease (CCHD) often related to bleeding from systemic to pulmonary collateral vessels, commonly referred to as aortopulmonary collaterals (APCs). While catheter-directed particle embolization has been historically utilized to treat arterial bleeds causing hemoptysis in other scenarios, coil and vascular plug embolization has been used more frequently to occlude parent arteries in CCHD patients given the concern for shunting of embolic particles. Recently, some centers have shifted to distal particle embolization in treating APCs given its advantages over proximal embolization, including ease of re-treatment if hemoptysis recurs. We describe our experience with distal particle embolization in two CCHD patients experiencing recurrent hemoptysis from APCs following proximal device embolization, highlighting the challenges in endovascular management of these patients.

1. Introduction

Aortopulmonary Collaterals (APCs) are a known sequela of cyanotic congenital heart disease (CCHD) that can cause life-threatening hemoptysis. Although corrective cardiac surgery has reduced the incidence of hemoptysis, APCs are common in CCHD patients, with a prevalence reported at 36% in post-Glenn and post-Fontan patients [1]. We present two cases of hemoptysis from APCs refractory to coil and vascular plug embolization in CCHD patients with prior corrective surgery.

2. Case 1

A 3-year-old girl with cyanotic congenital heart disease presented to our hospital with recurrent hemoptysis following a viral respiratory infection. Her past cardiac history included tetralogy of Fallot with pulmonary atresia and branch pulmonary artery hypoplasia. She previously underwent a unifocalization operation at 2 weeks of age with a right ventricle to pulmonary artery 5 mm polytetrafluoroethylene (PTFE) conduit, later revised at 10 months to an 8 mm PTFE graft. Left pulmonary artery balloon angioplasty was then performed at 26 months of age. Her medical history was also significant for severe tracheal stenosis requiring a tracheostomy. Our service was consulted given the recurrence of her symptoms despite a separate embolization for hemoptysis 9 days prior. Coils and Amplatzer II plugs (Jude Medical, Saint Paul, Minnesota) were used to proximally occlude the bilateral internal mammary arteries (IMAs) at that time. Catheterization data at this time was significant for mildly elevated right heart filling pressures (Right ventricular end diastolic pressure of 10 mm Hg), high-normal left-sided heart filling pressures (Left ventricular end diastolic pressure of 12 mm Hg) and a normal cardiac index (4 L/min/m^2). Competitive flow from APCs was present in the right upper lobe. She did not have additional hemoptysis during the admission and was discharged home in stable condition. However, the following day she was transferred from an outside hospital with recurrent symptoms. Given the severity of her symptoms and short interval of recurrence, she was placed on

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mechanical ventilation. Upon presentation to our hospital, she was hemodynamically stable with a blood pressure of 101/45 and oxygen saturations at her baseline range (84%, range approximately 80%–89%). Lung sounds were decreased in the right upper and middle lobes and her cardiac exam was unchanged with a normal S1, single S2 and a III out of VI medium pitched late peaking systolic ejection murmur along the left sternal border radiating to the axillae followed by a II out of IV diastolic decrescendo murmur at the left lower sternal border. Laboratory studies confirmed a hemoglobin drop from 13.4 to 10.7 g/dL. A chest radiograph correlated with her lung exam, demonstrating consolidation in the right upper and middle lobes. The Otolaryngology service was consulted and subsequently performed rigid bronchoscopy that confirmed clot in the right mainstem and upper lobe bronchi, however they were unable to control the bleeding. The patient was then transported to our angiography suite for embolization. Conventional angiography demonstrated a prominent right bronchointercostal artery (BICA) and persistent flow through the previously embolized right IMA supplying an APC. The distal right BICA was occluded with 350–500 μm Contour particles (Boston Scientific, Marlborough, Massachusetts). The right IMA was also cannulated, however, a wire would not pass across the Amplatzer II (Jude Medical, Saint Paul, Minnesota). The proximal vessel was dissected with a Transcend wire (Stryker, Freemont, California) while attempting to cross the plug (Fig. 1). No distal flow was present following the dissection. The patient recovered and was followed over a 12-month period without further hemoptysis. Computed Tomography (CT) angiography 1 month afterwards demonstrated persistent occlusion of the large right-sided intercostal bronchial trunk and resolution of the prior consolidation. Serial echocardiograms demonstrated preservation of right heart systolic function and evidence of abnormal septal motion suggestive of elevated right ventricular pressures. She underwent another balloon angioplasty of bilateral pulmonary arteries at 38 months of age. At this time a mild increase in right pulmonary artery saturation was noted compared to the left pulmonary artery (72% and 67%, respectively), suggestive of the continued presence of APCs. Since the last embolization, she has not undergone additional corrective surgery and has experienced no episodes of significant cyanosis or dyspnea.

3. Case 2

A 5-year-old boy with complex congenital heart disease presented to our service with recurrent hemoptysis in the setting of a viral upper respiratory tract infection (URI). His structural cardiac disease included atrial situs inversus, a hypoplastic double outlet right ventricle with a hypoplastic tricuspid valve, D-malposition of the great vessels, a hypoplastic main pulmonary artery with severe valvar and subvalvar stenosis, and a ventricular septal defect. At 3 months of age, he underwent a corrective left-sided bidirectional cavopulmonary anastomosis and atrial septectomy followed by catheter-directed right ventricular outflow tract occlusion at 5 months of age. He also underwent coil embolization of the bilateral IMAs and a right thyrocervical branch 7 months prior for a similar episode of hemoptysis. The mean pressure of his bidirectional cavopulmonary anastomosis was 14 mm Hg with a wedge of 10 to 11 mm Hg during this catheterization. A paucity of pulmonary markings in the right upper lobe was noted, likely related to alternate supply by APCs. His most recent episode of hemoptysis occurred while he was already in the hospital for a URI. A rapid response was called the first night of his PICU stay for 100 mL of hemoptysis. Oxygen saturations remained at baseline (range approximately 75%–85%). He was closely monitored without intubation or endoscopic intervention. His aspirin was held indefinitely after this episode. During his hospital course he had no further hemoptysis and was discharged home. Unfortunately, he returned 1 week later with 60–90 mL of recurrent hemoptysis. In the ED, he was hemodynamically stable with a blood pressure of 97/61 and oxygen saturations at his baseline (81%; range approximately 75%–85%). Physical examination revealed clear and equal breath sounds; a normal S1 and single S2; and a II out of VI systolic murmur along the left sternal border. Laboratory studies were significant for a drop in hemoglobin from 13.5 to 10.8 g/dL. Chest radiography did not reveal a focal lung consolidation. Given his recurrent hemoptysis, the consensus of the Critical Care, Cardiology, Otolaryngology, and Interventional Radiology services was to intubate the child and proceed with catheter-directed embolization. Angiography demonstrated a prominent left BICA, which was also present on images from the previous catheterization, a left subclavian artery collateral, and a recanalized left IMA collateral. The left BICA was embolized with 350–500 μm Contour particles. A microwire was used to cross the proximal IMA coils and particle embolization was performed (Fig. 2). The subclavian APC was cannulated with the parent catheter, however our Renegade microcatheter (Boston Scientific, Marlborough, Massachusetts) would not track distally into the vessel, thus embolization was not performed. He stabilized and was sent home. Echocardiography performed 1 month after discharge demonstrated a patent bidirectional cavopulmonary anastomosis and a mildly dilated left ventricle with normal systolic function. Unfortunately, he presented 2 months later with hemoptysis. On this presentation, he was hemodynamically stable with a blood pressure of 97/51 and oxygen saturations at his baseline (78%, range approximately 75–85%). Initial

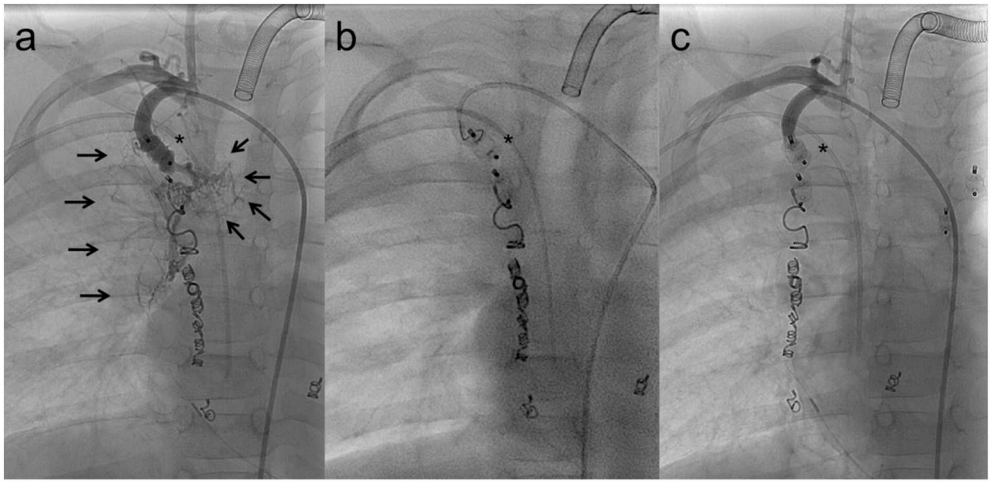

Fig. 1. Dissection of the right internal mammary artery at Amplatzer apposition.
(a) Recanalization of a right internal mammary APC (arrows) following Amplatzer (*) embolization; (b) Focal wire dissection at the location of Amplatzer deployment; (c) Occlusion of the distal internal mammary artery following dissection.

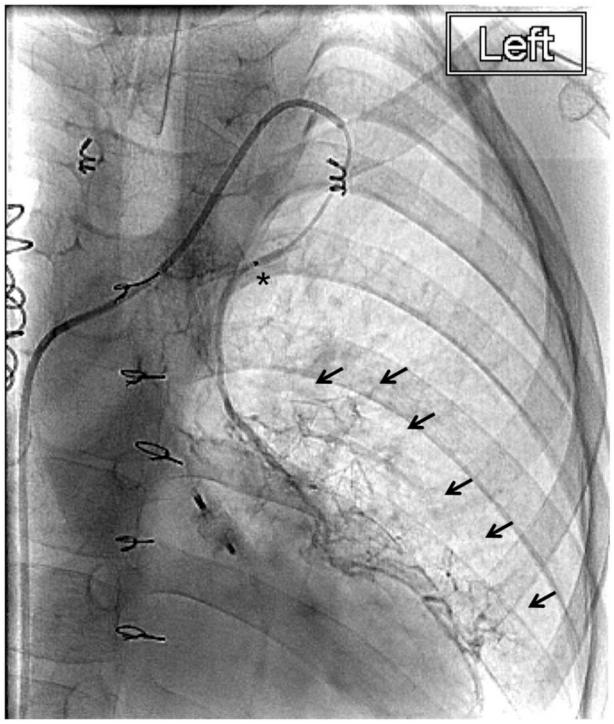

Fig. 2. Left internal mammary embolization past coils.
Left internal mammary APC cannulation (arrows) using a microcatheter (*) to cross a previously deployed coil.

physical exam revealed clear and equal breath sounds. Laboratory work up again demonstrated a drop in hemoglobin from 12.1 to 11.2 g/dL. CT angiography did not show active extravasation of contrast to localize the hemorrhage or consolidation of lung parenchyma. On arrival to the PICU, he suffered another episode of approximately 60 mL hemoptysis. After a multidisciplinary discussion, the consensus was to proceed with catheter directed embolization. Repeat angiography demonstrated left BICA and right IMA recanalization despite prior occlusion. Particle embolization was performed of the left BICA, right IMA, and a left subclavian artery APC through an Excelsior SL10 microcatheter (Stryker Neurovascular, Freemont, California). No additional hemoptysis occurred and the child was discharged home. He underwent a successful fenestrated Fontan operation 1 month later.

4. Discussion

These cases highlight the difficulties in managing CCHD patients with hemoptysis. Delayed bleeding from APCs 14 years after a corrective or staged procedure has been described with a prevalence of late hemoptysis as high as 12% in patients with pulmonary atresia and VSD [2–4]. Identifying CCHD children at risk for bleeding can be difficult, although the above cases demonstrate a temporal association with respiratory tract infections.

We began by conservatively managing both patients, but proceeded to aggressive treatment given their persistent hemoptysis [5]. Unfortunately, there are no reliable predictors of re-bleeding in patients with submassive hemoptysis (< 8 mL/kg in 24 h), making the decision to observe or proceed with catheter-directed therapy difficult [6]. Averin et al. observed a mortality rate of 75% in patients with hemoptysis prior to Fontan completion, supporting aggressive treatment of submassive hemoptysis in this group [7]. In both cases, multidisciplinary collaboration from our Critical Care, Cardiology, Otolaryngology, and Interventional Radiology services was utilized to develop a therapeutic plan.

Proximal embolization of the systemic arteries supplying APCs rather than the collateral vessels themselves can result in recanalization as was observed in both cases [8]. Though thought to cause permanent occlusion, Amplatzer plug (Jude Medical, Saint Paul, Minnesota) recanalization has also been reported in the literature [9]. While coils can be traversed with a microcatheter and wire for distal particle embolization, crossing a vascular plug with a microcatheter can be more difficult. Focal dissection at the location of the plug apposition with the vessel wall was successful in the first case.

Recently, a few medical centers have shifted towards the use of embolization particles to occlude APCs given their efficacy, ability to track distally, and patency of the parent vessel allowing easy access for re-embolization [7]. Although these particles are considered permanent, recanalization has been demonstrated in porcine models 4 weeks after embolization as was seen in the second case [10]. Higher rates were specifically associated with the 500–700 μm Contour SE brand [10]. Hypotheses for short-term recanalization include an incomplete redistribution of particles into distal vessels and non-elastic deformation of the spherical particle shape. However, as our cases demonstrate, even if recanalization occurs particle embolization can be repeated.

5. Conclusions

In summary, we describe two patients with CCHD and hemoptysis from APCs refractory to proximal embolization. Both cases demonstrate the challenges in endovascular management of these patients and highlight our experience with particle embolization. Particle embolization can be an effective alternative to proximal device occlusion, especially for refractory hemoptysis in CCHD patients.

Conflicts of interest

James Ryan Loftus: Declarations of interest: None.
Joseph Reis III: Declarations of interest: None.

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