Case report

Selective pulmonary artery occlusion to treat hemoptysis associated with pulmonary venous obstruction

Melissa S.W. Yamauchi, Mary Hunt Martin, Harlan R. Muntz, Ronald W. Day

The patient is a male with trisomy 21 who was born with a fenestrated atrial septum. He developed mild stenosis of the right pulmonary veins, severe stenosis of the left upper pulmonary vein, occlusion of the left lower pulmonary vein and pulmonary hypertension. He underwent surgical repair of the atrial septal defects and stenotic veins at 13 months of age. The right pulmonary veins have remained unobstructed since surgery. However, stenosis of the left upper pulmonary vein recurred within 8 months. There was angiographic evidence of severe stenosis of the central left upper pulmonary vein, occlusion of the central left lower pulmonary vein and diffuse hypoplasia of the peripheral, or upstream, left upper and lower pulmonary veins.

At an age of 15 years, the left upper pulmonary vein was dilated to a diameter of 6 mm with standard and cutting balloon catheters. An 8 mm Amplatzer® Vascular Plug II was placed in the proximal left lower pulmonary artery. An image of the left bronchus, two months after catheter-mediated interventions, is shown in Fig. 1. The prominent vessels of the left bronchus resolved. Pulmonary arterial angiograms immediately before and after placement of the vascular plug are shown in Fig. 2. Echocardiographic estimates of the mean gradient from the left upper pulmonary vein to the left atrium returned to pre-intervention estimates within a period of 1–2 years. Fig. 2 shows a lung-perfusion scan with a limited amount of blood flow to the left lung 42 months following the interventions. His episodes of hemoptysis have not recurred over a period of 4 years.

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ABSTRACT

Hemoptysis may occur in patients with pulmonary venous obstruction and prominent decompressing vessels in the airways adjacent to the affected pulmonary veins. The options for treatment of hemoptysis are limited, particularly when efforts to alleviate pulmonary venous obstruction have failed. Here we describe a patient with hemoptysis associated with stenosis of the central left upper pulmonary vein and occlusion of the central left lower pulmonary vein. The left upper pulmonary vein was dilated with balloon catheters and a vascular plug was placed in the left lower pulmonary artery. Vascular engorgement regressed in the left bronchus and hemoptysis has not recurred for 4 years despite recurrence of left upper pulmonary vein stenosis. Selective occlusion of branch pulmonary arteries may be an effective option for the treatment of hemoptysis from bleeding in lung segments with inoperable pulmonary venous obstruction.

1. Introduction

Pulmonary venous obstruction is a rare though serious condition with one collaborative study citing a 49% survival within three years of diagnosis [1]. Several case reports describe life-threatening hemoptysis in patients with pulmonary venous obstruction and bronchial or tracheal varices [2–4]. Surgical reconstruction of pulmonary venous drainage, anticoagulation for an occluding thrombus, and pulmonaryectomy have been described as options for hemoptysis in this setting [3–5]. Here we report the outcome of catheter-mediated interventions in an individual with refractory hemoptysis associated with severe stenosis of the left upper pulmonary veins and occlusion of the left lower pulmonary veins.

2. Case report

Our patient is a male with trisomy 21 who was born with a fenestrated atrial septum. He developed mild stenosis of the right pulmonary veins, severe stenosis of the left upper pulmonary vein, occlusion of the left lower pulmonary vein and pulmonary hypertension. He underwent surgical repair of the atrial septal defects and stenotic veins at 13 months of age. The right pulmonary veins have remained unobstructed since surgery. However, stenosis of the left upper pulmonary vein recurred within 8 months. There was angiographic evidence of severe stenosis of the central left upper pulmonary vein, occlusion of the central left lower pulmonary vein and diffuse hypoplasia of the peripheral, or upstream, left upper and lower pulmonary veins.

He developed hemoptysis at the age of 13 years. An image of the left bronchus is shown in Fig. 1. He had no large systemic to pulmonary arterial collaterals. Nonetheless, a small left bronchial artery was occluded with a small coil. The severity of hemoptysis gradually progressed with moderate to large volume hemoptysis occurring once or twice a month.

At an age of 15 years, the left upper pulmonary vein was dilated to a diameter of 6 mm with standard and cutting balloon catheters. An 8 mm Amplatzer® Vascular Plug II was placed in the proximal left lower pulmonary artery. An image of the left bronchus, two months after catheter-mediated interventions, is shown in Fig. 1. The prominent vessels of the left bronchus resolved. Pulmonary arterial angiograms immediately before and after placement of the vascular plug are shown in Fig. 2. Echocardiographic estimates of the mean gradient from the left upper pulmonary vein to the left atrium returned to pre-intervention estimates within a period of 1–2 years. Fig. 2 shows a lung-perfusion scan with a limited amount of blood flow to the left lung 42 months following the interventions. His episodes of hemoptysis have not recurred over a period of 4 years.
3. Discussion

Our patient developed episodes of hemoptysis, which were sufficiently severe that care providers and family members considered the possibility of treatment with partial lung resection. We were concerned that surgical and catheter-mediated options for the left upper pulmonary vein alone would not provide lasting relief of central vein stenosis, mitigate diffuse disease in the peripheral veins, or prevent recurrent hemoptysis. Alternative options included surgical placement of a left pulmonary artery band, partial intraluminal occlusion of the proximal left pulmonary artery, complete intraluminal occlusion of selective peripheral left pulmonary arteries, and resection of the left lung. We decided to initially attempt non-surgical options. The episodes of hemoptysis resolved following balloon dilation of the left upper pulmonary vein and placement of a vascular plug in the left lower pulmonary artery.

Balloon dilation of the stenotic left pulmonary vein initially decreased the pressure gradient from the vessel to the left atrium. It is possible that this intervention alone was the reason for resolution of bronchial varices. Balasubramanian and associates have reported initial relief of pulmonary vein stenosis by balloon dilation and stent placement [6]. Unfortunately, the reported actuarial survival at 5 years was only 50% and a substantial number of patients underwent subsequent interventions. Further, an increased risk of embolic complications with catheter interventions for pulmonary vein stenosis has been reported [7]. Hemoptysis has not recurred in our patient despite evidence of recurrent stenosis by Doppler echocardiography and evidence of lim-

Fig. 1. Images of the left bronchus during bronchoscopy. Dilated blood vessels with an erythematous and corrugated mucosa are present in the left main bronchus at 13 years of age (upper). Small blood vessels without an erythematous mucosa are present in the left main bronchus two months after balloon dilation of the left upper pulmonary vein and placement of a vascular plug in the left lower pulmonary artery (lower).

Fig. 2. Pulmonary vascular anatomy and perfusion. A manual injection of contrast was performed with transient balloon occlusion of the left lower pulmonary artery. There is pruning and decreased arborization of the peripheral vessels (left upper). There is indistinct filling of small peripheral pulmonary veins with no detectable central left lower pulmonary vein (right upper). A vascular plug is positioned in the proximal left lower pulmonary artery (left lower). Contrast fills the left upper and right pulmonary arteries well. There is a limited amount of blood flow to the left lung 42 months following balloon dilation of the left upper pulmonary vein and occlusion of the left lower pulmonary artery (right lower). The following geometric means for the distribution of flow to each lung zone using anterior and posterior images were reported: right upper 16.6%, right middle 40.0%, right lower 30.3%, left upper 6.3%, left middle 5.8%, left lower 1.0%.
ited left upper lung perfusion following balloon dilation of the left upper pulmonary vein.

A vascular plug was used to selectively occlude the left lower pulmonary artery and isolate the region of lung with the greatest amount of pulmonary venous obstruction. We believe the vascular plug decreased the distal left lower pulmonary arterial pressure and left lower pulmonary venous pressure. Thereby, collateral vessels in the airway were able to regress. In future patients, bronchoscopy could be performed during heart catheterization with transient balloon occlusion of selected vessels. If immediate, a noticeable change in the appearance of bronchial varices might help to identify appropriate vessels to target before placing a vascular plug.

The effects of partial occlusion of selective pulmonary arteries are unknown. We are concerned that partial occlusion may initially lower the distal systolic pulmonary arterial pressure, but will not lower the pulmonary venous pressure or prevent hemoptysis long-term.

In summary, selective occlusion of the left lower pulmonary artery was associated with resolution of hemoptysis in a patient with central left lower pulmonary venous obstruction. Additional experience with selective pulmonary artery occlusion is needed to determine whether it is a safe and effective treatment option for patients with hemoptysis associated with obstructed pulmonary veins that are not readily amenable to surgical repair.

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