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Case Reports

Transcatheter Occlusion of Large Pulmonary Arteriovenous Malformations Using Multiple Devices in a Neonate

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Pulmonary arteriovenous malformations (PAVMs) are usually well-tolerated hemodynamically. Rarely, they cause severe neonatal cyanosis, requiring urgent treatment. We report the use of multiple transcatheter vascular occlusion devices to reduce right-to-left shunting in a neonate with multiple, large PAVMs. © 2014 Wiley Periodicals, Inc.

Key words: PAVM; arteriovenous malformation; percutaneous vascular occlusion; neonatal cyanosis

INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are often diagnosed incidentally on a chest imaging study and are well-tolerated hemodynamically. Rarely, they present with severe neonatal cyanosis due to right-to-left shunting. Historically, PAVM treatment has included surgical segmental resection, lobectomy, or pneumonectomy [1,2]. With advances in transcatheter therapy, PAVM occlusion using embolization coils has been established in infants as an effective alternative to surgical resection [3]. Also, the efficacy of vascular plugs has been demonstrated in children, and more recently in an infant [4,5]. We present a case of multiple, large PAVMs requiring several transcatheter occlusion devices for the treatment of profound neonatal cyanosis.

CASE REPORT

A 3.8 kg full term male infant presented with dyspnea and hypoxemia shortly after birth; room air systemic oxygen saturation was 75%, which did not improve with supplemental oxygen. Physical examination was notable for diminished right-sided breath sounds and a 2/6 systolic ejection murmur at the left mid-sternal border. There were no telangiectasias or hemangiomas. A chest radiograph demonstrated mild cardiomegaly, right middle and upper lobe opacities. An echocardiogram demonstrated left atrial enlargement, small atrial septal defect with left-to-right shunt, moderately diminished left ventricular systolic function, and a moderate sized patent ductus arteriosus with left-to-right shunt; the intracardiac anatomy was otherwise normal. A chest CT scan demonstrated multiple, large, complicated right-sided PAVMs. A head CT scan was normal, without cerebral arteriovenous malformations (AVMs) or intracranial hemorrhage. Genetic studies for hereditary hemorrhagic telangiectasia (HHT) were negative.

On the second day of life, the patient was referred for cardiac catheterization to define the PAVM anatomy and for transcatheter PAVM occlusion. A

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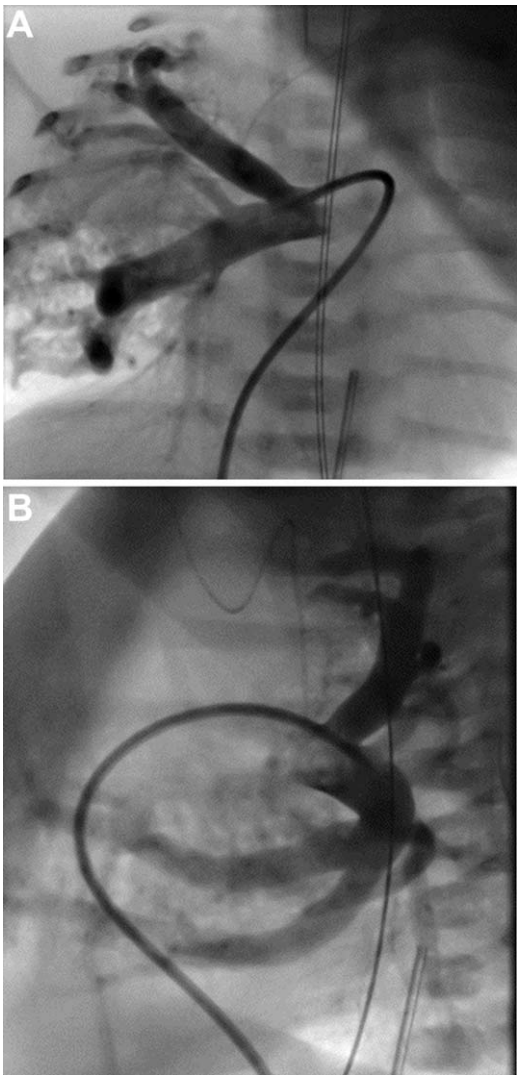

Fig. 1. Right pulmonary artery angiogram in the PA (A) and lateral (B) projections demonstrating two large PAVMs associated with the right upper and middle lobes. The right middle lobe PAVM bifurcates into two large PAVMs. The right lower lobe artery is small.

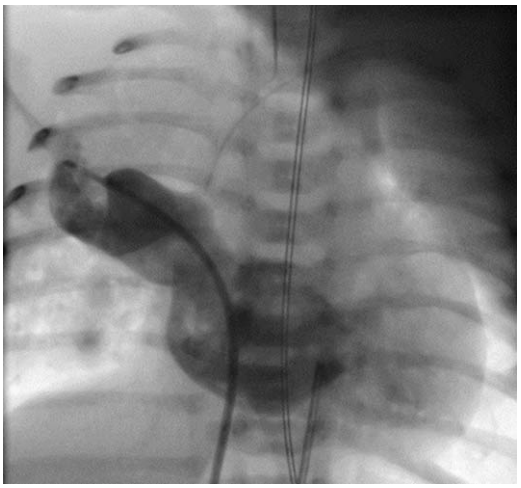

Fig. 2. A contrast injection reveals an enormous right upper pulmonary vein draining to the left atrium.

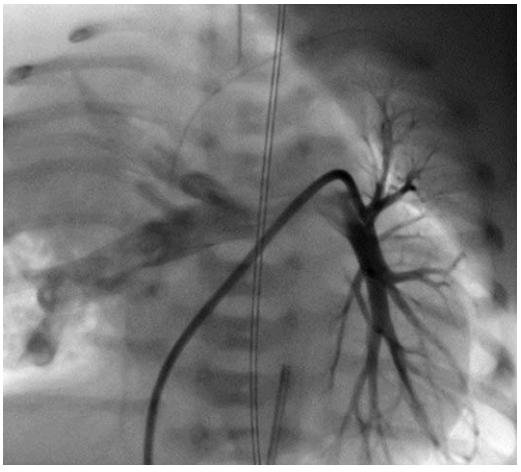

Fig. 3. An angiogram is performed in the proximal left pulmonary artery. The left pulmonary artery bifurcates normally; no PAVMs or other abnormalities are noted.

selective right pulmonary artery angiogram demonstrated two large PAVMs (Fig. 1A and B). The largest PAVM (7.8 mm) was associated with the right middle lobe pulmonary artery and bifurcated into two moderate-large sized superior (4.5 mm) and inferior (4.8 mm) PAVMs. The second large PAVM (6.6 mm) was associated with the right upper lobe pulmonary artery and bifurcated into two smaller PAVMs. There was markedly increased flow through these vessels, with contrast returning through an enormously dilated right upper pulmonary vein to the left atrium (Fig. 2). The right lower pulmonary artery was small, but without PAVMs. A left pulmonary artery angiogram displayed normal vasculature (Fig. 3).

A 5 Fr transseptal sheath was positioned in the right middle lobe PAVM for vascular plug delivery. During device delivery, with the transseptal sheath across the tricuspid valve, there was transient, exacerbated hypoxemia, which resolved promptly when the sheath was withdrawn to the right atrium following deployment of each device. The superior branch of the right middle lobe PAVM was occluded with a 6 mm Amplatzer Vascular Plug II (AVP II) (St. Jude Medical, St. Paul, MN). As there was some residual flow through this device, an 8 mm AVP II was deployed just proximal to the first device. The inferior branch of the right middle lobe PAVM was occluded with two 8 mm AVP II devices. The right upper lobe PAVM was occluded with a 10 mm AVP II. A repeat angiogram in the right pulmonary artery demonstrated no residual flow

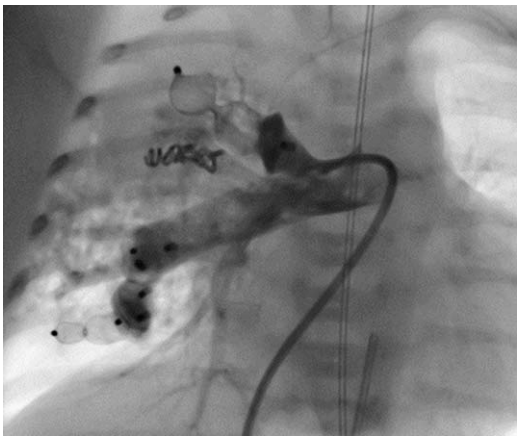

Fig. 4. Following vascular plug deployment in the right upper and right middle lobe AVMs, and placement of two coils in the right upper lobe AVM, there is still contrast flow through residual AVMs, mostly associated with the right middle lobe.

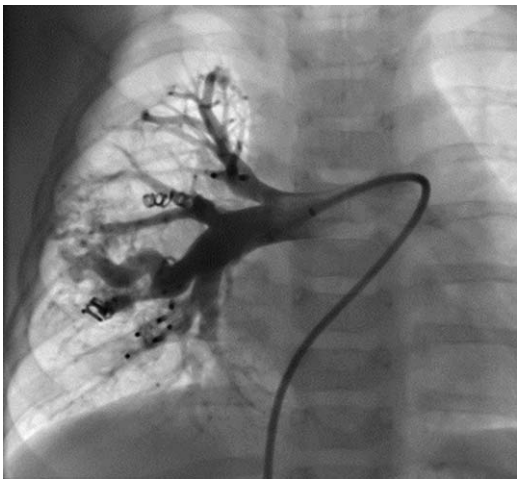

Fig. 5. Right pulmonary artery angiogram at age 7 months demonstrates a new, large right middle lobe PAVM.

through the occluded PAVMs. However, following occlusion of these large PAVMs, smaller PAVMs were evident. A right upper lobe PAVM (2.9 mm) originated just distal to the previously occluded right upper lobe PAVM. A right middle lobe PAVM (3.2 mm) originated between the previously occluded superior and inferior branches. A 4 Fr snare catheter was used to deploy two 0.038″ × 4 cm × 3 mm MReye embolization coils (Cook, Bloomington, IN) in two small right upper lobe PAVMs (Fig. 4). The snare catheter was positioned in the right middle lobe PAVM to deploy a 0.038″ × 5 cm × 5 mm MReye embolization coil. Two more proximal, small PAVMs were identified arising from the right middle lobe pulmonary artery, and each was occluded with a 0.038″ × 4 cm × 3 mm MReye embolization coil. At the conclusion of the procedure, his systemic arterial oxygen saturation was 90% while receiving FiO_2 35%. He weaned promptly from mechanical ventilation, extubated easily, and discharged home one week following the catheterization with a systemic oxygen saturation of 88% in room air.

He was evaluated monthly in the Pediatric Cardiology clinic, and his oxygen saturation rose gradually to 95%. However, over several months this declined to 85%. At age 7 months, he returned to the cardiac catheterization lab for diagnostic catheterization and occlusion of additional PAVMs. A selective left pulmonary artery angiogram was normal. A right pulmonary artery angiogram demonstrated no flow through the previously placed, well positioned occlusion devices. There was a large right middle lobe PAVM (5.5 mm) (Fig. 5). This was occluded with a 6 mm AVP II. As there was residual flow through this device, an 8 mm AVP II was placed just proximal to the 6 mm device. A follow-up angiogram demonstrated several small vessels, arising proximal to these plugs, supplying this large PAVM. Two 0.038″ × 4 cm × 3 mm MReye embolization coils were deployed to occlude the small tributary vessels. A right pulmonary artery angiogram demonstrated complete occlusion of the large right middle lobe PAVM and small tributary vessels (Fig. 6A and B). There was flow through a small right upper lobe PAVM; however stable catheter positioned could not be maintained for coil delivery. His systemic oxygen saturation, which was 85% at the start of the catheterization, improved to 92% at the conclusion of the procedure. He extubated easily, recovered uneventfully, and was discharged home the following day.

The most recent evaluation was at age 36 months. He has not required any interventions. He is growing and developing normally (weight > 95th percentile for age), with a normal respiratory and cardiovascular examination, and systemic oxygen saturation 93% in room air. Chest radiograph demonstrated normal heart size, normal pulmonary vascular markings in the left lung and right lower lobe, occlusion devices in stable position in the right middle and right upper lobes, and interval decrease in the size of the enlarged right upper pulmonary vein. The oxygen saturation of 93%, although markedly improved, suggests persistence of small PAVMs.

DISCUSSION

The differential diagnosis for neonatal cyanosis encompasses many illnesses. In the presence of hypoxemia refractory to oxygen and the absence of congenital heart defects with right-to-left shunting or severe neonatal respiratory disease, congenital PAVM should be considered. Embryologically, PAVMs result from

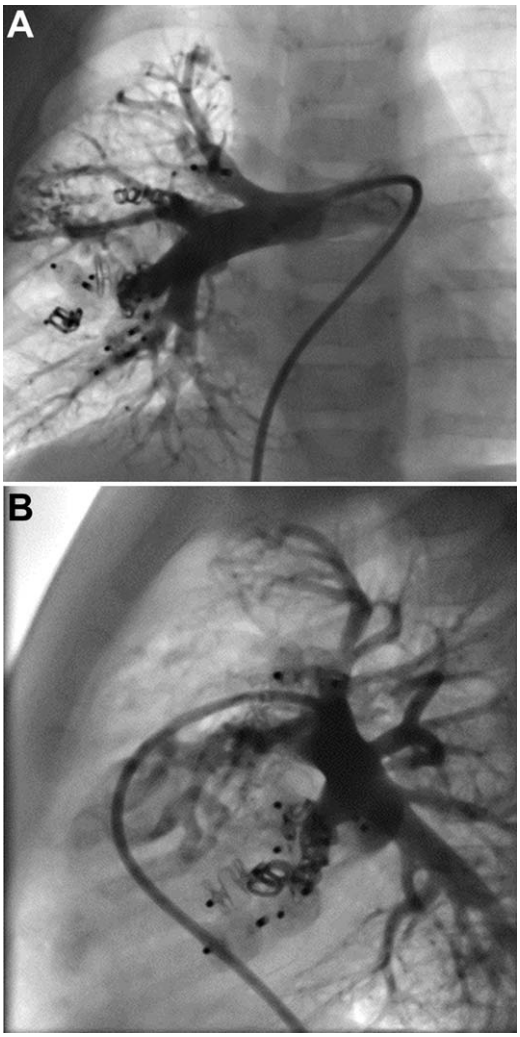

Fig. 6. Right pulmonary artery angiogram in PA (A) and lateral (B) projections demonstrates occlusion of right middle lobe PAVM and tributary vessels, with small residual right upper lobe PAVM.

persistent pulmonary arteriovenous connections without an intervening capillary bed, due to incomplete vascular septae development between primitive pulmonary arteries and venous plexuses [6]. Physiologically, PAVMs are low resistance channels through which blood is preferentially shunted, bypassing capillary gas exchange, resulting in an intrapulmonary right-to-left shunt. This leads to hypoxemia, polycythemia, and risk for paradoxical embolism. Though rare, neonates that are symptomatic due to PAVMs can be critically ill, as in our case, presenting with profound cyanosis and dyspnea, as well as congestive heart failure or lung hemorrhage. Mortality is high, up to 39% in the first week of life; 33% of these infants were suspected to have HHT [7].

Between 47% and 80%, all congenital PAVMs are associated with HHT; conversely, the incidence of PAVM in all patients with HHT is 5–15% [8]. The diagnosis of HHT is made clinically based on the presence of 3 of 4 Curaçao Criteria: epistaxis, telangiectasia, visceral AVM, or positive family history. While our patient does not meet diagnostic criteria for HHT, a high degree of suspicion is required to diagnose HHT in a newborn, as clinical findings are often absent until later in life. Genetic testing can be helpful in infants with a family history of HHT with a known mutation. Genetic testing in the absence of a family history of HHT is not as sensitive, as these patients often have novel mutations in the *ENG* or *ACVRL1* genes [9]. Determining HHT status in neonates with PAVM is important, as patients with HHT are more likely to have multiple PAVMs, more rapid disease progression, and more complications [8]. While uncommon, cerebral AVM in infants with HHT is especially concerning, as intracranial hemorrhage in this population is almost universally fatal [10].

Prior to 1994, surgical resection, lobectomy, or total pneumonectomy was recommended for symptomatic neonatal PAVM [2,11]. Transcatheter occlusion of neonatal PAVM using Gianturco stainless steel coils was first described by Grady et al [3]. Transcatheter occlusion is preferable to surgical resection to avoid surgical morbidity and mortality. Also, by selectively embolizing the abnormal vasculature, the normal pulmonary vasculature is preserved.

Our case emphasizes the importance of a persistent and thorough catheterization evaluation, and using various types of occlusion devices, depending on vessel size and morphology, to occlude as many PAVMs as possible. We used MReye embolization coils and the AVP II. While use of the Amplatzer Vascular Plug has been described in the occlusion of PAVMs in older children, and recently in one neonate, to our knowledge the AVP II has not been described for the treatment of neonatal PAVM [4,5]. The AVP II is advantageous in large PAVM occlusion, due to its stability in high flow vessels and rapid embolization properties [12]. The recently released lower profile AVP 4 can be delivered through a catheter with 0.038″ inner diameter, as opposed to a larger sheath. Using the AVP 4 may have prevented the transient oxygen desaturation during device delivery in our patient. However, given the recommendation for 30% device oversizing, the largest AVP 4 (8 mm), might not have occluded the largest PAVM in our patient [13]. Additionally, this case reinforces the importance of assiduous clinical follow-up. As has been described previously, and noted in our case, these patients may have residual PAVMs or develop enlargement of previously minor PAVMs, necessitating re-intervention [11,14]. In one large series, 19% of patients developed significant enlargement of feeding arteries to previously small PAVMs, and 7% developed

recanalization of previously occluded PAVMs, usually through embolization coils [15]. Serial occlusion of PAVMs facilitates the detection of smaller malformations which have the potential to become hemodynamically significant over time. Also, this technique promotes growth of the normal pulmonary vasculature. In our patient, the normal, but small, right lower lobe pulmonary artery has grown following occlusion of the PAVMs. Normal pulmonary artery branches are now visible in the right middle and upper lobes. Clinical follow-up is ongoing, as the systemic oxygen saturation of 93% suggests residual small PAVMs, along with evaluation for signs and symptoms of HHT.

CONCLUSION

PAVM is a rare, though treatable and reversible cause of marked cyanosis in neonates. In all cases, an evaluation should be performed for extra-PAVMs, especially cerebral AVM and HHT. Over the last decade, several transcatheter vascular occlusion devices have become available, and are especially useful for treating neonatal PAVMs. Careful angiographic evaluation of vessel size and morphology ensures use of the best device for each lesion. Serial clinical follow-up evaluations are required after initial treatment, as additional interventions may be required to treat persistent, enlarging, or new PAVMs, as noted by recurrent hypoxemia or other sequelae.

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