Closure of right pulmonary artery to left atrium fistula by duct occluder device

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ABSTRACT

We report the successful transcatheter closure of the right pulmonary artery fistula to the left atrium in a 4-year-old boy, who had presented with cyanosis and easy fatigability, identified after two-dimensional echocardiogram with bubble contrast study, computed tomography (CT) angiography, and cardiac catheterization. The fistula was successfully closed by a transcatheter approach using an 18/16 duct occluder device. There was marked improvement clinically and no complication on 2-months follow-up.

Keywords: Duct occluder device closure, left atrium, right pulmonary artery fistula, transcatheter

INTRODUCTION

Right pulmonary artery (RPA) to left atrium (LA) fistula is a rare cause of cyanotic congenital heart disease, first described by Friedlich *et al.* in 1950.[1] There are only about fifty cases reported in the literature.[2] Untreated cases may suffer from hyperviscosity syndrome due to chronic hypoxemia, brain abscess, systemic thromboembolism, or cerebrovascular accident.[2,3] Management options include surgical ligation or transcatheter closure of the fistulous tract by using coils or device. Although one older surgical series has reported a 22% mortality rate,[3] newer surgical techniques and transcatheter procedures using coils and devices can provide a cure at acceptable risks.[4] We report the successful closure of an RPA-to-LA fistula by a transcatheter approach using a duct occluder device in a child with severe cyanosis.

CASE REPORT

A 4-year-old boy presented with the history of cyanosis, easy fatigability, and an episode of loss of consciousness. On examination, he had clubbing and cyanosis. His oxygen saturations were 75% in room air. He had no murmur on auscultation. Electrocardiography (ECG) and chest X-ray were normal [Figure 1]. Bubble contrast study showed delayed bubbles in LA after 4 beats, suggestive of pulmonary arteriovenous malformation. Transthoracic echocardiography showed RPA to LA communication [Figures 2 and 3]. Computed tomography angiogram showed RPA to LA fistula [Figures 4-6]. Cardiac catheterization was done under general anesthesia after informed consent. Access from both side femoral veins and right femoral artery were taken. RPA angiogram with MPA2 4F catheter (Cordis Corp, USA) confirmed RPA to LA fistula. It measured 11.4 mm in diameter [Figure 7]. The septal puncture was performed with Brockenbrough needle and 8F Mullins sheath. A 0.035" Terumo exchange length wire (Japan) was snared in LA with AMPLATZ Goose Neck Snare 25 mm (Medtronic, USA) through an MPA2 6F catheter (Cordis Corp, USA) from the RPA introduced from the left femoral vein. Thus, a venovenous rail was established [Figure 8]. AMPLATZER Sizing balloon 24 mm (St Jude Medical, Inc., USA) was used to size the fistula over the Terumo wire in AP/RAO/LAO views [Figure 9a and b]. The narrowest diameter measured was 10 mm. AMPLATZER Septal Occluder device 16 mm (St Jude Medical, Inc., USA) was deployed through 10 F

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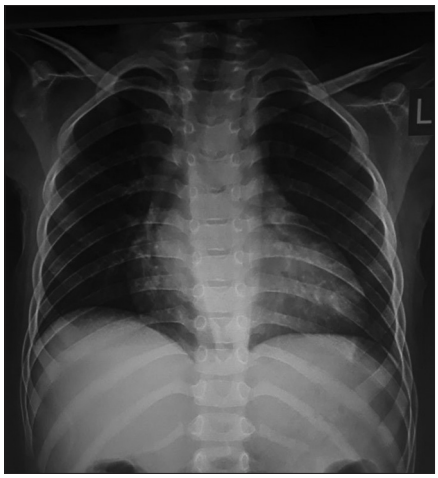

Figure 1: Normal chest X-ray of the patient

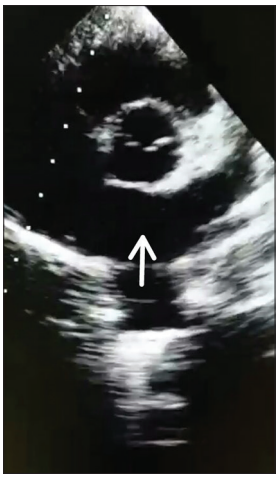

Figure 2: Two-dimensional echocardiogram, parasternal short-axis view, arrow showing direct communication of the right pulmonary artery to the left atrium

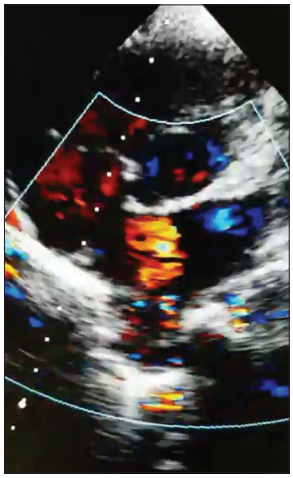

Figure 3: Transthoracic Doppler echocardiogram, parasternal short-axis view showing direct communication of the right pulmonary artery to the left atrium

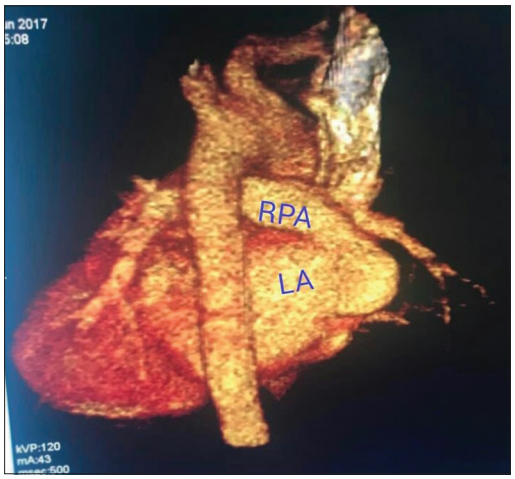

Figure 4: Computed tomography angiogram showing posterior view of the heart

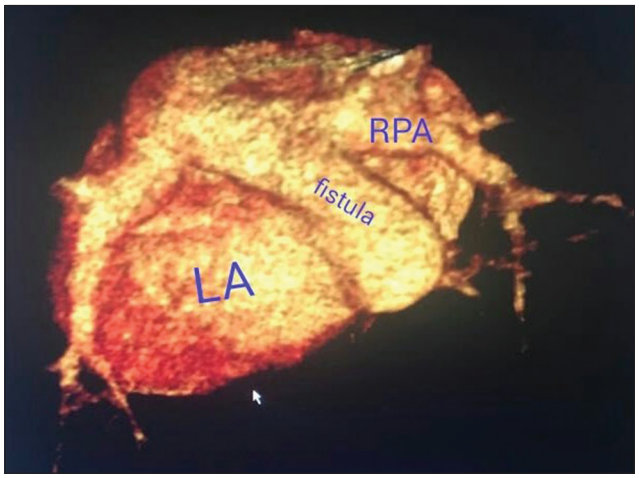

Figure 5: Computed tomography angiogram showing superior view of the heart showing dilated right pulmonary artery and its fistulous connection to the left atrium

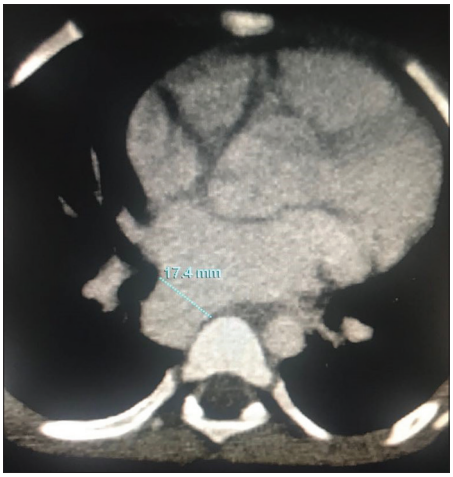

Figure 6: Computed tomography angiogram axial view

delivery system; but, as there was severe hypotension and desaturation, it was retrieved.

Then, duct occluder device 18/16 (Lepu Medical Technology, Beijing China) was delivered through 10F delivery system with stable position and no residual

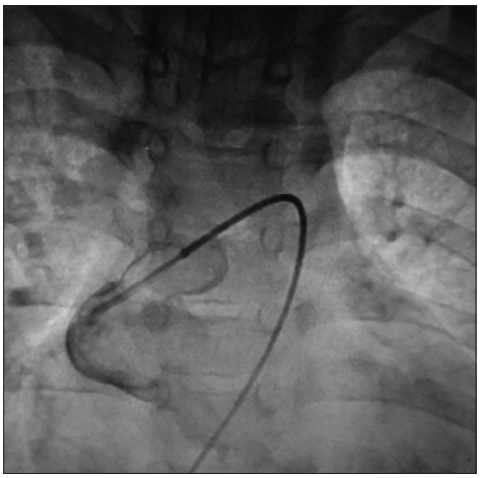

Figure 7: Cine angiogram showing right pulmonary artery-to-left atrium fistula

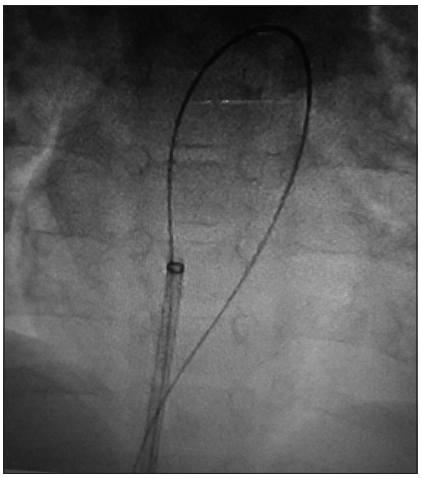

Figure 8: Cine fluoroscopy showing venovenous loop in anteroposterior projection

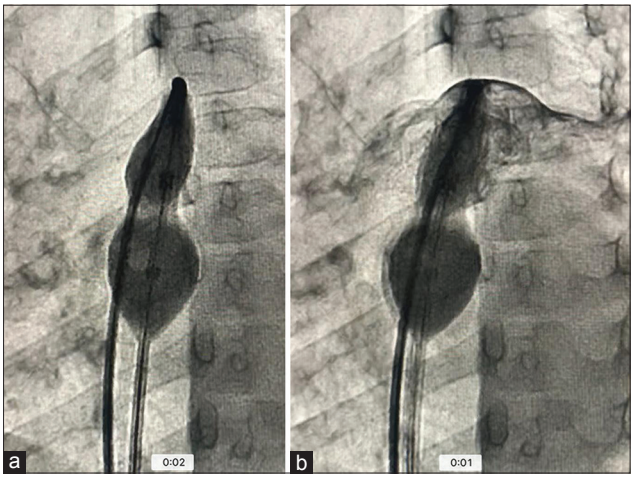

Figure 9: (a and b) Fluoroscopy showing balloon sizing of the narrowest diameter of the communication

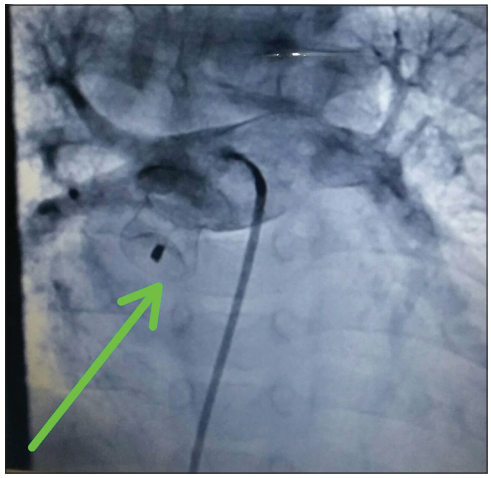

Figure 10: Location of the device was confirmed by angiogram performed in LAO and AP views

shunt noted [Figure 10]. During the procedure, the patient developed SVT twice while attempting interatrial septal puncture, requiring cardioversion, followed by amiodarone infusion. The patient's oxygen saturation increased to 96% at room air. Echo showed no residual shunt with normal flow in the pulmonary artery and pulmonary veins [Figure 11a and b]. At 2-month follow-up, the patient was asymptomatic.

DISCUSSION

Pulmonary artery–LA fistula has been categorized into three types.[5] A fourth type has been added subsequently.[6] In Type I, RPA branches normally. However, an additional fistulous channel connects RPA to LA. Pulmonary venous return is normal. In Type II, the lower lobe branch of RPA drains directly into LA, forming an aneurysmal sac. This communication drains into LA in place of the absent right lower pulmonary vein. Right lung abnormalities are the rule[7]; one or more lobes may be absent. Type III is similar to Type I with pulmonary veins draining into the abnormal channel that connects RPA and LA. Type IV is similar to Type II with pulmonary veins entering the aneurysmal sac. Our patient had abnormal communication between RPA and LA with normal branching of RPA distal to the communication, normal pulmonary venous drainage, and lung development. He had Type I pulmonary artery-LA fistula.

Although they have manifested from day one to middle age,[6] most cases present in the third decade.[8] Among symptoms and signs, dyspnea is seen in 50%, neurological symptoms in 20%,[9] telangiectasia in 33%,[10] and cyanosis in 60% patients. Among the reported cases, only one patient had murmur (continuous murmur).[11] ECG may be normal.[6] An atrial septal defect (ASD) is the most commonly associated intracardiac anomaly; however, pulmonary abnormalities include the absence of the lower or middle lobe, right lung sequestration, and diverticulum of the right main bronchus, as have been reported in the literature.[11]

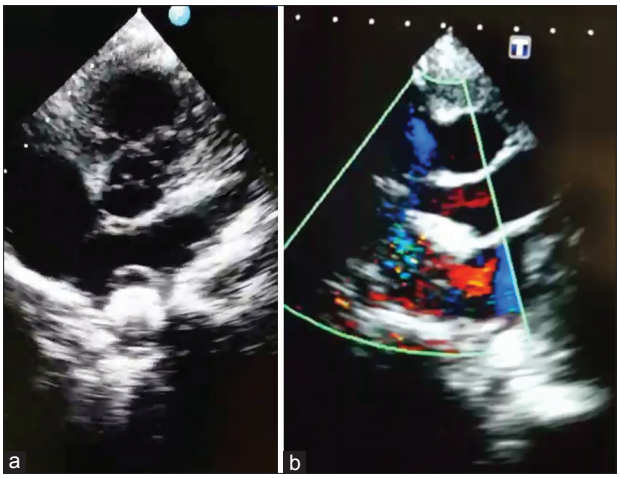

Figure 11: (a) Transthoracic echocardiography parasternal short-axis view after deployment of device across the communication. (b) Transthoracic echocardiography Doppler showing no residual shunt across the device

Intervention is required to prevent paradoxical embolism. Other complications are rupture, hemoptysis, polycythemia and brain abscess.[12] In 1984, a case of 22-month-old girl with Down syndrome was reported who had a direct communication of RPA to LA that led to systemic desaturation; she also had ASD secundum that spontaneously closed. She underwent successful ligation of the abnormal communication.[13] In 1997, a case of a 4-year-old girl was reported who presented with unexplained cyanosis, fistula between RPA and LA was identified after echocardiography and angiography, which was successfully ligated using extracorporeal circulation.[3] Transcatheter closure of RPA to LA fistula in a 6-year-old boy using duct occluder device was reported in 2013 who presented with cyanosis and dyspnea.[14] A case of the RPA-to-left atrial fistula with ASD was reported in 2018. The fistula was detected after the patient developed desaturation following surgical closure of the ASD. It was managed with a transcatheter (trans-RPA route) closure of the fistula using an AMPLATZER ventricular septal defect closure device.[15]

We conclude that although it is a rare entity, suspecting the diagnosis and making accurate radiological and clinical evaluation can bring forth a treatable pathology. Closure of the defect by duct occluder device is a successful treatment option with marked clinical improvement.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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