Circular shunt in a pulmonary artery to right atrial tunnel, an anomaly unreported so far

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ABSTRACT

While aortico-right atrial tunnels with left to right shunt from aorta to right atrium are reported widely, pulmonary artery to right atrial tunnels have not been described so far. Such a tunnel will lead to a circular shunt with a recirculation of blood in the right sided cardiac chambers repeatedly bypassing the pulmonary capillary bed. This newly described pulmonary artery to right atrial tunnel was closed nonsurgically with a duct occluder after angiographic delineation.

Keywords: Circular shunt, device closure, pulmonary artery to right atrium tunnel

INTRODUCTION

Circular shunts are defined as intracardiac communications where the shunted blood returns to the same chamber without traversing a capillary bed.[1] An abnormal communication from the pulmonary artery to the right atrium will result in a recirculation of blood through the right sided cardiac chambers bypassing the pulmonary capillary bed, thereby causing a circular shunt without causing any systemic hypoxia. Such a communication has not been reported so far in literature. We report a case of an anomalous fistula between pulmonary artery and right atrium causing enlargement of right atrium and right ventricle, which was closed successfully in the catheterization laboratory.

PATIENT DETAILS AND DISCUSSION

A seven month old baby boy with Trisomy 21 presented with growth failure (weight 5.1 kg) and respiratory symptoms. He had normal oxygen saturations, a long systolic murmur in the right sternal border, normal splitting of the second sound along with mild cardiomegaly on chest X-ray. His electrocardiogram (ECG) showed right axis deviation and prominent right ventricular forces usual for his age. Echocardiogram showed dilated right atrium and right ventricle. On parasternal short axis view, there was an abnormal trifurcation of the main pulmonary artery. [Figure 1a and b, Video 1a and b] The pulmonary artery had the usual right and left pulmonary artery branches and a third abnormal tunnel that arose from the origin of the left pulmonary artery coursed over the roof

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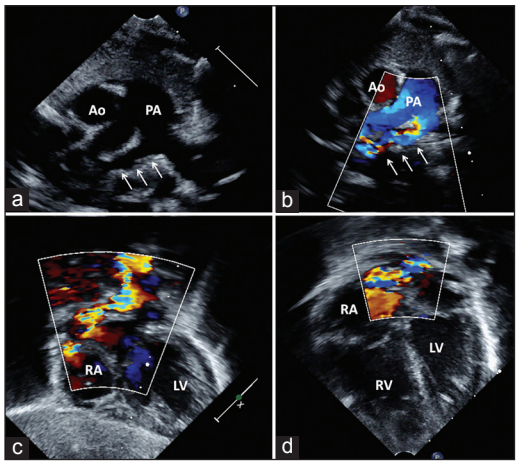

Figure 1: Echocardiogram from parasternal short axis view (a and b, Video 1a and b) showed abnormal trifurcation of the main pulmonary artery into the usual right and left branches and a third abnormal communication (white arrows) into the right atrium. The entry of the communication into the right atrium is delineated well on subcostal coronal (c, Video 1c) and apical views (d, Video 1d). Ao-ascending aorta; PA-pulmonary artery

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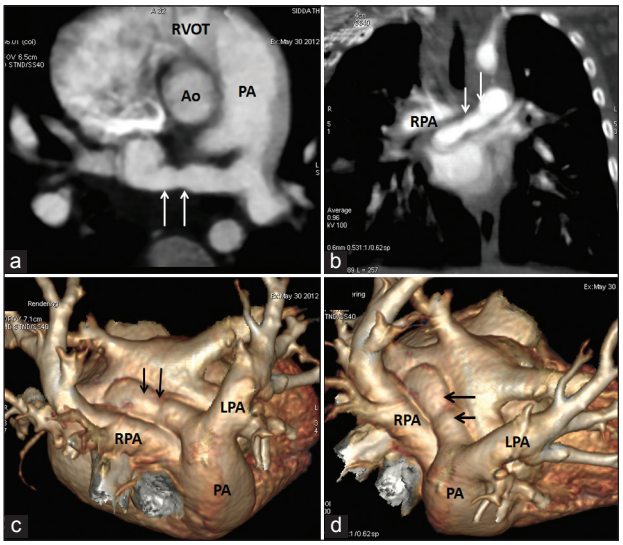

Figure 2: Computed tomographic images in axial (a) and coronal planes (b) show the abnormal communication (two arrows) from the pulmonary artery bifurcation to the right atrium. Volume rendering (c and d, Video 2) delineate the communication well. RPA-right pulmonary artery; LPA-left pulmonary artery; RVOT-right ventricular outflow tract

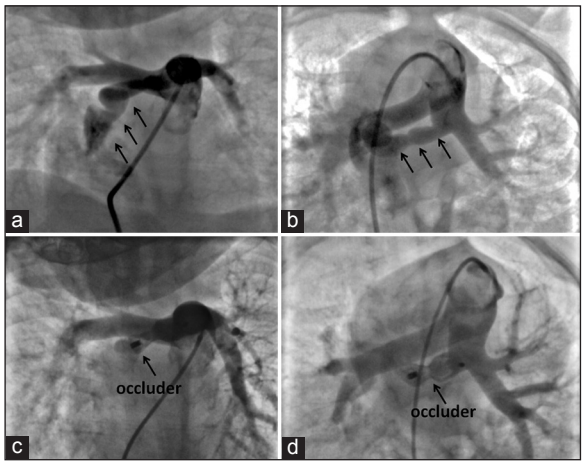

Figure 3: Pulmonary angiogram (Video 3a and b) show the abnormal communication from pulmonary artery to right atrium (three arrows) in cranial (a) and caudal (b) projections. The communication was successfully occluded (c and d, Video 3c and d) with an Amplatzer duct occluder device during the cardiac catheterization

of the left atrium in the transverse sinus of the heart towards the right and drained near the superior vena caval orifice of right atrium. The tortuous course of this channel and entry into the right atrium was well seen from the subxiphoid view in coronal plane [Figure 1c, Video 1c] and apical view [Figure 1d, Video 1d]. There was abnormal continuous flows into the right atrium through this tunnel. Contrast enhanced multislice computed tomography in axial and coronal slices [Figure 2a and b] and volume rendered images [Figure 2c and d, Video 2] confirmed the abnormal tunnel like communication from pulmonary artery to right atrium coursing in the transverse sinus of heart causing a circular shunt and dilatation of right atrium and right ventricle. Diagnostic pulmonary artery angiogram in cranial and caudal projections [Figure 3a and b, Video 3a and b] delineated this 6 mm wide tunnel. The pulmonary artery pressure and right atrial pressures were mildly elevated. Two femoral venous access were taken, and the tunnel was crossed from the pulmonary artery with a guidewire, which was snared in the right atrium to get a veno venous railroad. A long 6F sheath was advanced through the right atrium into the tunnel. This tunnel was closed with a 8-6 Amplatzer duct occluder (St Jude Medical, Minnesota, MN) in the cardiac catheterization laboratory [Figure 3c and d, Video 3c and d] and position was checked with pulmonary angiogram through the other venous access. Follow up echocardiogram showed cessation of flows in the tunnel and reduction in size of the right ventricle. While different cardiac lesions causing circular shunts have been reported in the past, this rare variation of circular shunt from a pulmonary artery to right atrium involving only the right sided cardiac chambers without causing systemic hypoxia is unreported so far.[2]

This anatomy may be embryologically akin to the aortico-right atrial tunnel which often arises from the left side of the aortic root or from the undersurface of the aortic arch, courses in the transverse sinus of the heart and drains in to the right atrium and causes a left to right shunt.[3] The anatomical location of the fistula in this patient was also similar in its course through the transverse sinus of the heart over the roof of the left atrium and drainage near the superior vena caval orifice of the right atrium. While the proximal origin of aortico-right atrial tunnel has been documented from the left side of the aortic root and also from the undersurface of the aortic arch, this patient had the proximal origin from the main pulmonary artery near the confluence. In the recent years, aortico-right atrial tunnels with significant left to right shunts have been managed with transcatheter interventions.[4] We opted to close the tunnel in our patient too in the catheterization laboratory. If left alone, the obligatory shunt from the pulmonary artery to right atrium may result in progressive dilatation of the right sided cardiac chambers. Since this anomaly has not been reported in the past in literature, the natural history of an uncorrected pulmonary artery to right atrial tunnel is unknown.

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