Giant coronary-pulmonary fistula with pulmonary atresia, ventricular septal defect, and coronary anomaly: A case report and review of literature

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ABSTRACT

Congenital coronary-pulmonary artery fistula is a rare condition and is usually associated with pulmonary atresia. We present a 10-year-old girl with circumflex coronary artery to pulmonary artery (PA) fistula with a giant dilated circumflex coronary artery, ventricular septal defect (VSD), pulmonary atresia, and anomalous originofleft anterior descending (LAD) artery from right coronary sinus. The patient underwent surgical correction. We report the investigation, treatment, and review of literatureof this rare congenital anomaly associated with dilated circumflex coronary artery.

Keywords: Coronary to pulmonary artery fistula, pulmonary atresia, ventricular septal defect

INTRODUCTION

Congenital coronary-pulmonary artery fistula with pulmonary atresia and ventricular septal defect is a rare condition. In this case report we discuss the preoperative evaluation and surgical management of this unusual association.

CASE REPORT

A 10-year-old girl was referred to our institute for evaluation and treatment of cyanotic congenital heart disease. She complained of cyanosis since birth and easy fatigability. She did not complain of chest pain. She was investigated with transthoracic echocardiography, cardiac catheterization, and cardiac computed tomography (CT). Patient was severely cyanotic with room air saturation of 80%. Auscultation revealed a continuous murmur. Transthoracic echocardiography showed findings of tetralogy of Fallot, pulmonary atresia, and dilated circumflex coronary artery. Transthoracic echocardiography in parasternal short axis at the level of great arteries demonstrated the fistula arising from left circumflex coronary artery (LCX) and draining into the pulmonary artery (PA) [Figure 1]. Cardiac catheterization showed a blindright ventricular outlet tract (RVOT), hypertrophied right ventricle (RV), overriding of aorta, and dilated circumflex coronary artery. Retrograde filling of dye to PA from dilated circumflex coronary artery was seen in aortic root angiogram [Figure 2 and Video 1]. CT scan findings were inconclusive. Surgical findings were as follows. Anomalous origin of left anterior descending (LAD) from right coronary cusp (RCC) was seen, which was crossing the RVOT. Circumflex artery originated from left coronary cusp (LCC). Circumflexcoronary artery was hugely dilated with a fistula connecting to main PA [Figure 3]. The fistula was transected and sutured at the coronary end.Rastelli procedure was done using aortic homograft in view of LAD artery crossing the RVOT and associated pulmonary atresia. Associated small collaterals were dissected and ligated. Patient had uneventful recovery and was discharged on ninth postoperative day. Child was on aspirin for first 6 months postoperative period.Patient is doing well at 6 months follow-up.

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DOI: 10.4103/0974-2069.132496

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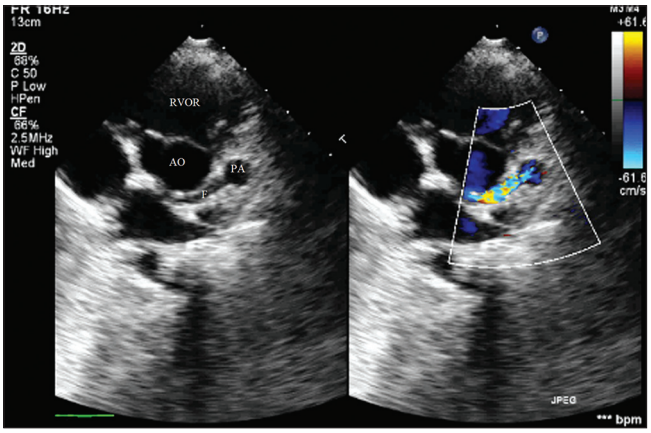

Figure 1: Transthoracic echocardiography in parasternal short axis at the level of great arteries demonstrated the fistula arising from LCX and draining in to PA. LCX = left circumflex coronary artery, PA = pulmonary artery

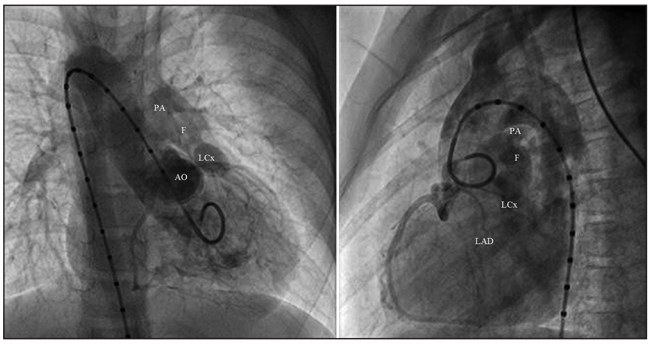

Figure 2: Cardiac catheterization showing retrograde filling of the dye to PA from dilated circumflex coronary artery through the coronary to pulmonary fistula in aortic angiogram. PA = pulmonary artery

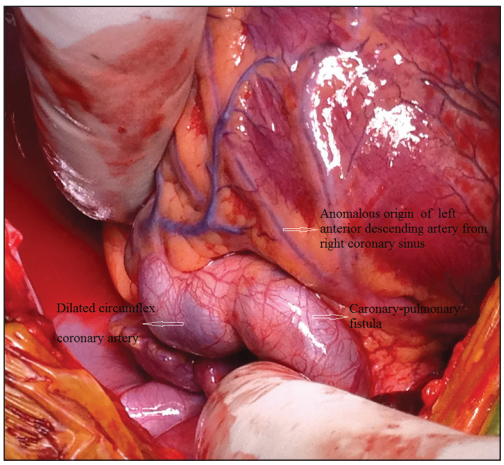

Figure 3: Operative picture showing hugely dilated circumflex artery and the fistula

DISCUSSION

Pulmonary atresia with ventricular septal defect (VSD) and major systemic to pulmonary arterial collaterals is a complex congenital heart lesion with an incidence of 4.2/100,000 live births.[1] The embryological basis of coronary to pulmonary fistula is explained by Amin *et al.*[2] In cases of pulmonary atresia with VSD, congenital coronary artery to PA fistula is reported with an incidence of 10%.[2] In a study done by Changela *et al.*,[3] from a single center series of more than 2200 cases they reported only one case of coronary artery to PA fistula. There have been many retrospective studies and case reports[4-6] of coronary artery to PA fistula, but none of them reported the combination of coronary anomaly and aneurysm of circumflex artery that we have reported in this case. Coronary to pulmonary fistula may provide the sole supply to the pulmonary arterial system but most often it is associated with major aortopulmonary collaterals.[7] Coronary-pulmonary artery fistula with pulmonary atresia and VSD should be managed by any technique that allows preservation of the coronary circulation and connection of the RV to the PA. Rastelli operation was done because this is the procedure of choice in pulmonary artesia and more so with LAD crossing the RVOT. In our case, we transected the fistula and sutured the coronary end and used pulmonary side of fistula to complete anastomosis for Rastelli procedure. In previously reported studies and case reports, the diagnosis was made by transthoracic echocardiography, cardiac cauterization, cardiac CT, magnetic resonance imaging, and during surgery.[7,8] In our reported case, the cardiac CT images were inconclusive in interpretation because child had tachycardia during the procedurethat resulted in imaging artefacts. The important clinical implication of coronary to PA fistula is the need for detailed investigations, recognition, before taking up for surgery. Hence, all cases of VSD with pulmonary atresia should be evaluated for coronary-pulmonary artery fistula.

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How to cite this article: Seetharama Bhat PS, Girish Gowda SL, Mahimarangaiah J, Manjunath CN. Giant coronary-pulmonary fistula with pulmonary atresia, ventricular septal defect, and coronary anomaly: A case report and review of literature. Ann Pediatr Card 2014;7:142-4.

Source of Support: Nil, **Conflict of Interest:** None declared