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ALCAPA with right ventricular diverticulum – a rare dual anomaly

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Abbreviations: ALCAPA: Anomalous Origin of Left Coronary Artery from Main Pulmonary Artery; RVD: Right Ventricular Diverticulum; BWG: Bland-White-Garland Syndrome.

Description

A 1-month-old neonate with known atrial septal defect (ASD) and congestive heart failure (CCF) was referred to our centre for further evaluation through Computed Tomographic Angiography (CTA). CTA revealed anomalous origin of left coronary artery from main pulmonary artery (ALCAPA) (Figure 1), moderate right ventricular enlargement and hypertrophy with thickened trabeculae. Further, a tubular outpouching (15 x 7 x 13 mm, CC, AP and TR dimensions) arose from the anterior wall of right ventricle (RV). It showed full thickness ventricular wall and contracted along with rest of the RV which suggested muscular type of right ventricular diverticulum (RVD) (Figure 2). Additional findings included large ostium secundum ASD (Figure 3a), dilated main pulmonary artery (PA). Further rare finding included anomalous origin of left lower lobe PA from right descending PA (Figure 3b). The neonate was initially managed medically with a plan for surgery later on. Unfortunately, the neonate passed away a week after CTA. ALCAPA is also known as Bland-White-Garland syndrome [1] is associated with other cardiac anomalies such as ASD, ventricular septal defect, patent ductus arteriosus etc [2]. Only 8 cases of muscular RV diverticula have been reported [3]. ALCAPA associated with RVD is an extremely rare presentation and to our knowledge is the first reported association of ALCAPA, ASD and RVD. It is clinically important to distinguish RVD from aneurysm as diverticulum could be the source of arrhythmia, cardiac rupture, and thrombosis. Diverticulum have a narrow neck and contract synchronously along with rest of RV myocardium whereas aneurysm is akinetic or dyskinetic. ALCAPA is treated with coronary artery implantation to left coronary cusp, ASD with open heart surgery or by device placement through cardiac catheterization. If the diverticulum is small and asymptomatic; it can be left for conservative follow-up. If large and presenting with symptoms, it needs to be excised.

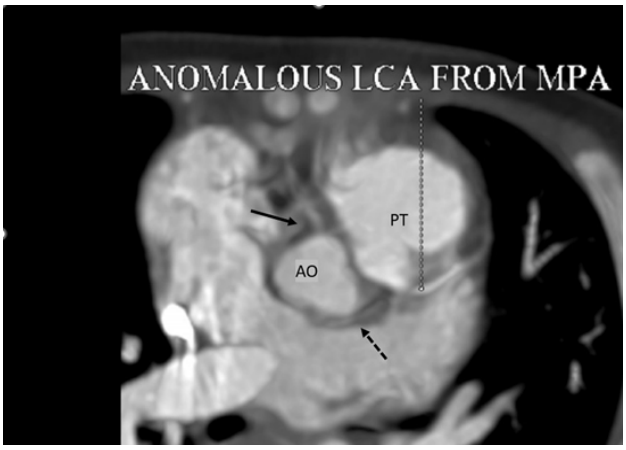

Figure 1: Computed tomographic angiography shows anomalous origin of the left coronary artery from the pulmonary trunk (ALCAPA). Note the origin of right coronary artery (RCA) from right coronary cusp (line arrow) and absent left coronary artery from left coronary cusp (dotted arrow). Ao=aorta; PT=pulmonary trunk.

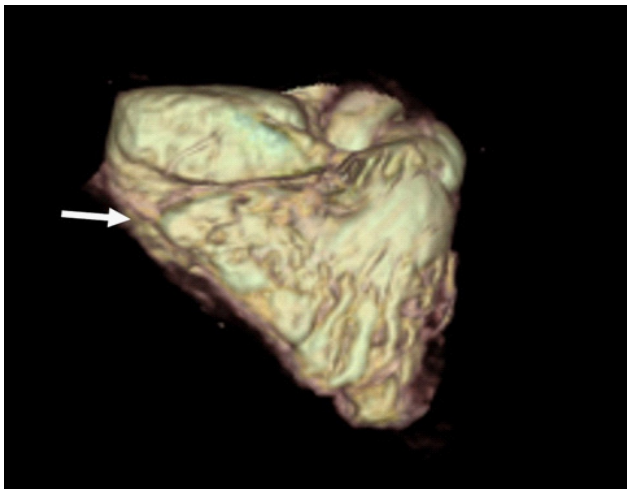

Figure 2: A 3D volume-rendered computed tomographic angiography showing a diverticulum arising from anterior margin of right ventricle (white arrow). RV=Right Ventricle.

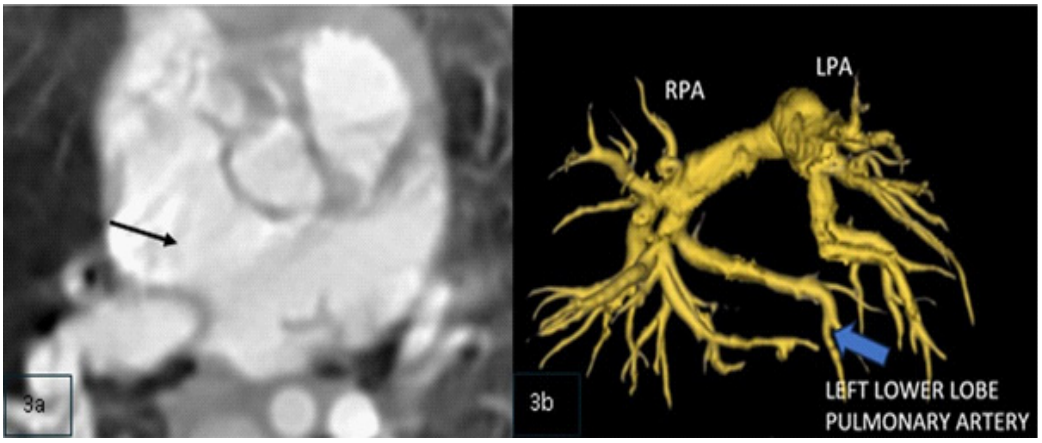

Figure 3: (a) Axial image shows a large atrial secundum defect (line arrow). **(b)** 3D volume-rendered computed tomographic angiography shows anomalous origin of the left lower lobe pulmonary artery from right descending pulmonary artery. RPA =Right pulmonary artery. LPA= Left pulmonary artery.

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