Clinical Image

An Uncommon Chest Pain in a 26-Year-Old Woman: Anomalous Origin of Left Coronary Artery from the Pulmonary Artery

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Received: 13 May 2019
Accepted: 26 May 2019
Published: 02 June 2019

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1. Clinical Image

A 26-year-old woman reported a history longer than 2 months of precordial pain, exertional chest tightness and shortness of breath. Her past medical history was remarkable for remote endocardial fibroelastosis and permanent atrial fibrillation. A grade 2/6 systolic murmur was heard at the cardiac apex. Her serial cardiac enzyme results were negative. Electrocardiographic examination showed left ventricular hypertrophy. Echocardiography revealed a dilated left ventricle and mild pulmonary hypertension. A significantly dilated right coronary artery(RCA) arised from the right aortic sinus of Valsalva (**Figure 1a**). A mildly dilated vessels was observed in the territory of left coronary artery(LCA), while the origin of LCA was not identified. Subsequent coronary angiography demonstrated an inability to visualize LCA. The right coronary artery was a large vessel with the suggestion of right to left coronary filling LCA and main pulmonary artery(PA) (**Figure1a-b**). Because of the poor angiographic visualization and high suspicion of anomalous origin of the left coronary artery from the pulmonary artery(ALCAPA), cardiac MSCT angiography was further required. This confirmed an enlarged tortuous RCA originating from the right coronary sinus of the aortic root with extensive collateralization to LAD and left circumflex artery, which could be traced to the common trunk of LCA connecting with the posterior aspect of the main PA(Figure2a-b). Final diagnosis of ALCAPA was established. Conservative treatment rather than surgical intervention was implemented, and at clinical follow-up 6 months after initial diagnosis at imaging, the patient remained free of previous symptoms.

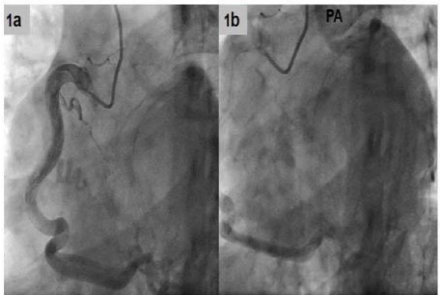

Figure1a-b: Poor opacification of a dilated and tortuous RCA suggests a patent vessel with extensive posterior and apical collateralization. Retrograde filling of the left anterior descending artery and main pulmonary artery are obscurely demonstrated on delayed imaging after contrast injection of the RCA.

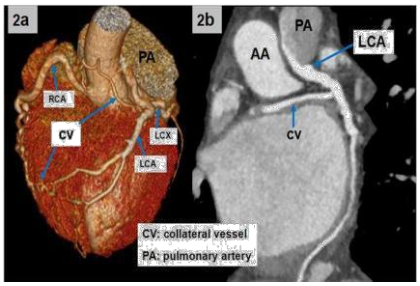

Figure2a-b: Anterior volumetric rendering demonstrates a dilated right coronary artery originating from the right coronary cusp with extensive collateralization to left coronary artery. Curve planar reconstruction reveals anomalous origin of the left coronary artery from the pulmonary artery.

ALCAPA is a rare congenital cardiac anomaly affecting approximately 1 in 300,000 in live births, and the mortality rate in the pediatric population exceeds 90% in untreated population[1]. Survival to adulthood is dependent on the early development of extensive collateral circulation from the RCA to LCA and reversal of flow from the LCA into the PA. Patients with ALCAPA usually present with severe different clinical symptoms like angia, myocardial infarction, heart failure, or sudden death[2]. Those patients lack of symptoms may be associated with well-developed collateral circulation between the left and right coronary arteries. Modern advances in noninvasive cardiac imaging such as CT angiography and MRI, have substantially increased the number of diagnosed cases of adults with benign clinical outcomes, uncovering a large adult population that has not been reviewed. As the gold standard diagnostic modality, coronary angiography is not always necessary in the current era. Coronary CT angiography can accurately depict the origin and proximal course of anomalous coronary artery, such as LCA arising from the main pulmonary artery. Meanwhile, it can also reveal a dilated and tortuous RCA and dilated intracoronary collateral arteries along the epicardial surface of the heart. The reported case emphasizes the importance of multimodality imaging in the diagnosis of ALCAPA. In a series of 11 cases of adult ALCAPA, Jurishica demonstrated a high incidence of sudden death at a young age[3]. When ALCAPA is diagnosed, surgery is often recommended in order to prevent future myocardial ischemia, malignant ventricular arrhythmias and sudden cardiac death.

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