



Origin of Right Coronary Artery from the Ascending Aorta: An Extremely Rare Anomaly

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Introduction

The incidence of coronary anomalies in patients undergoing coronary angiography varies from 0.27% to 1.66% [1]. Many of these anomalies are clinically benign; while, others are associated with serious morbidity. We describe the case of a patient undergoing coronary angiogram to evaluate her cardiomyopathy revealing an anomalous right coronary artery arising from the ascending aorta above the left sinotubular junction plane. We discuss the rarity of this anomaly along with its clinical importance.

Case Report

A 54 year old female with a history of benign resected ovarian tumor and poorly controlled hypertension, who presented to the emergency department with three days of worsening lower extremity edema and exertional dyspnea. She denied having any chest pain, orthopnea, paroxysmal nocturnal dyspnea, palpitations, lightheadedness, or prior syncopal episodes. Patient was afebrile on presentation to the emergency department with a blood pressure of 189/143, pulse rate of 136 and respiratory rate of 18. On examination patient was found to have irregularly irregular heart sounds, tachycardia, jugular venous distention of 10cm H₂O, lungs were clear to auscultation bilaterally, and 1+ pitting low extremity edema. EKG revealed atrial fibrillation with rapid ventricular rate in 130s. The transthoracic echocardiogram showed severely reduced Left ventricular ejection fraction of 20%, Left posterior wall thickness of 1.5cm, Intraventricular septal wall thickness of 1.5cm, and severely dilated left atrium suggestive of hypertensive cardiomyopathy.

During the hospital stay the patient was treated with AV nodal blocking agents and diuresis. She underwent coronary angiography to evaluate for ischemia as the etiology of her cardiomyopathy.

During the left heart catheterization procedure, the left main coronary artery was easily engaged using a Judkins left catheter. The angiogram of the left system revealed minimal non-obstructive disease. However, despite multiple attempts with the Judkins right and Williams right catheters, we were unable to cannulate or even visualize the right coronary artery (RCA) ostium. Hence an aortic root shot in LAO projection, 30 degrees angulation was performed, which revealed the location of an anomalous RCA arising from the ascending aorta above the left sinotubular junction (Figure 1). The RCA was then engaged successfully using an Amplatz left-1 (AL-1)

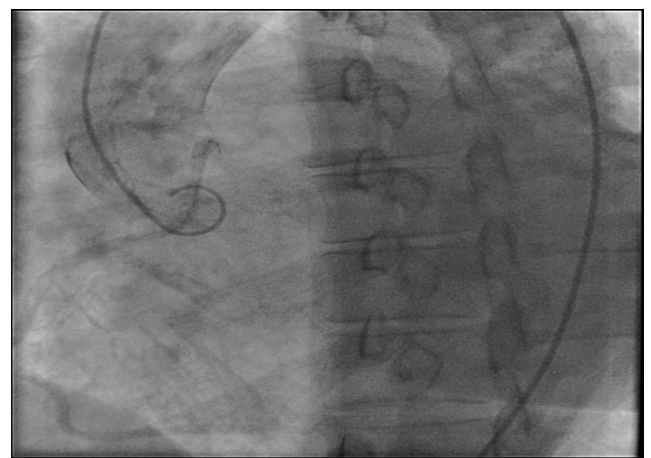


Figure 1: Aortogram in LAO projection

catheter and an angiogram revealed a dominant, large RCA with mild luminal irregularities (Figure 2).

Discussion

Our patient's poor LV function couldn't be explained by the level of atherosclerosis she had, her cardiomyopathy appears to be secondary to her long standing history of poorly controlled hypertension, atrial fibrillation causing tachycardia induced CMP or a combination of both. Anomalous origin of a coronary artery poses a technical challenge during coronary angiograms. It increases the amount of radiation exposure as well as the contrast load utilized to patients. With an increase in the amount of time and the number of catheters, the likelihood of complications also increases, hence it is very important to be aware of the possibilities and have knowledge of them, when it is not possible to cannulate or visualize the coronary by the standard techniques. The most frequent anomalies of coronary origins are associated with the circumflex artery. Anomalies of the origin of RCA are more rare comparatively. Among the anomalous origins of the RCA, the origin from the left sinus of Valsalva was more common in one study 6-27% [1]. Another study found the RCA originating from the LAD or CX to be 12.5% [2]. However, there are only few reported cases of the RCA originating from ascending aorta above the sinotubular junction plane [2-6]. A study by Serkan et al.

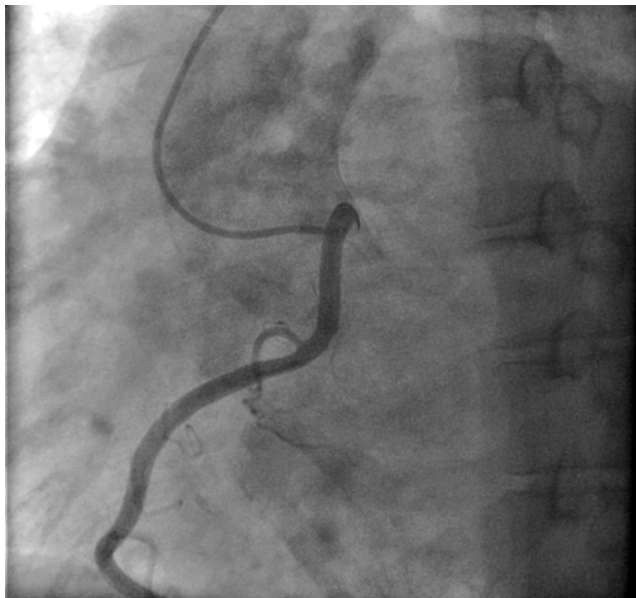


Figure 2: Right coronary artery angiography using an Amplatz Left 1 catheter

reported 0.006% prevalence of this rare anomaly, which constitutes only 2.1% of all the anomalously originating coronary arteries [2,3]. There are reports of typical angina pectoris, myocardial infarction, dyspnea, syncope, ventricular tachycardia, and sudden cardiac death related to anomalously arising RCA [7-11]. The origin of the vessel from the contralateral sinus of Valsalva or artery with subsequent passage between the aorta and right ventricular outflow tract has been clearly shown to be a dangerous anatomy [9,10]. Congenital coronary artery anomalies are frequently found to be responsible for sudden death in young athletes, in approximately 20% of cases, and represent the second most frequent disease responsible for athletic field deaths [11,12]. Since the standard catheters are shaped specifically in order to easily cannulate the normally originating coronaries, the success to engage the anomalously arising ostium using the same catheters is limited. In addition to being extremely rare, the anomalous RCA arising from the aorta above the sinotubular junction plane, poses more challenge in being engaged by the standard catheters, as compared to other anomalously arising coronaries. Literature review revealed that these anomalous RCA were very successfully and easily cannulated using Forward take off Judkins left guide (FL) 3.0 and/or Femoral Curve left (FCL) 3.0 and 3.5 catheters [3]. We had our

success with using an AL-1 catheter. Coronary CT Angiography and Cardiac MRI play an important role in evaluating the exact anatomy without risk of any extra contrast dye or radiation or when it is not feasible to characterize the coronary anatomy by conventional coronary angiogram due to the anomalous origin. In patients undergoing diagnostic coronary angiograms or percutaneous coronary interventions the angiographic recognition of these anomalous coronary origins is vital to the success of the procedure, specifically to the choice of the diagnostic or guiding catheter used to engage the ostium of the anomalous artery.

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