Anomalous Origin of all Coronary Arteries from Right Sinus of Valsalva (RSOV)

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Received: 21 December 2018; Accepted: 15 January 2019

Citation: Manousopoulos Konstantinos, Bourboulis Nikolaos. Anomalous Origin of all Coronary Arteries from Right Sinus of Valsalva (RSOV). Cardi Vasc Res. 2019; 3(1); 1-6.

Case
A 70-year-old man admitted for chest tightness on exertion for approximately 5 days. The patient had a history of systemic hypertension as risk factor for coronary artery disease. He had no history of tobacco or alcohol use. Physical examination was normal. His resting electrocardiogram was normal. Electrocardiography revealed normal sinus rhythm with no ST segment changes. Echocardiography revealed normal left ventricular function. The patient referred for catheterization due to suspected coronary artery disease with angina. Angiography through the right radial artery and use of a 6F left Judkins catheter was unable to cannulate the ostium of main stem. With the use of the same catheter selective injection of the right coronary artery revealed a single coronary artery (right coronary artery) and left main (LM) arising from the same right coronary ostium. Following the course of the coronary vessel LM was divided to left anterior descending artery (LAD) and left circumflex artery (LCX). The coronary artery was free of atherosclerotic changes and no intervention was planned.

Figure 1: Anomalous origin of the LM from the RSOV.

The incidence of an anomalous origin of the LM from the RSOV among patients who undergo angiography has been estimated between 0.008 and 0.017% [1]. LM either arises independently from the ostium of the RCA, or less frequently as in our case the two arteries share a common ostium [3]. Four different types of this extremely rare coronary abnormality have been described based on the course of the ectopic LM. Type A: Anterior to the right ventricular outflow tract, Type B: Between the aorta and the pulmonary trunk, Type C: Cristal, through the supraventricular crest and interventricular septum, Type D: Dorsal or posterior to the aorta [2,3].

The anomalous origin of the left coronary artery from the RSOV is the most frequent and has been consistently related to myocardial infarction and sudden cardiac death. The incidence is significantly higher in patients with interarterial course of the LM [4]. Although few cases of sudden death and myocardial ischemia associated with a posterior course of the LM have been described, this type of anomaly is considered mostly benign [5]. Anginal symptoms are usually related to exercise and are caused by compression of the proximal part of the LM between the expanded aortic root and the pulmonary trunk. Therapeutic approach must be individualized according to symptoms, age and the anatomy of the aberrant LM. Surgical correction is generally indicated in young symptomatic patients who are at high risk of sudden death. Multidetector computed tomography scan (MDCT), cardiac magnetic resonance (CMR) and transesophageal echocardiography (TEE) are commonly used for the diagnosis and imaging of the origin and course of coronary arteries disease. However, coronary angiography remains the gold standard for the diagnosis. Three dimension multiplane TEE is a minimally invasive imaging modality that can portray directly the proximal and interarterial course of the LM [6]. Cannulation of the ectopic coronary arteries during angiography can be
extremely challenging and success depends mostly on physicians’ experience. Intravascular ultrasound (IVUS) has also been used in order to obtain cross-sectional luminal images.

Figure 2: Anomalous origin of the LM from the RSOV in a RAO view.

Conclusion
In conclusion, ectopic origin of coronary arteries from the same ostium is a very rare entity during angiography. The majority of patients remain with no symptoms during their lifetime, certain types of these malformations lead to myocardial infarction and sudden cardiac death. Ectopic coronaries arteries have challenging and time consuming cannulation; therefore, such other diagnostic tools such as TEE, MDCT or CMR may have a complementary role to make the correct diagnosis.

References