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CASE REPORT

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Variant origin of three main coronary ostia from the right sinus of Valsalva: report of a rare case

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Observing anomalies in the origin of the coronary arteries is a rare but recognised scenario during coronarography. All the major coronary arteries originating from the right sinus of Valsalva is an extremely rare anomaly, its reported incidence being 0.008% in angiographic studies. Most coronary artery variations are benign and are therefore found accidentally or postmortem. However, some anomalies in the origin of the coronary arteries are associated with myocardial ischaemia and a higher risk of sudden cardiac death.

Herein, we report a sporadic case of anomalous origin of the coronary arteries, in which the right coronary artery, anterior interventricular artery and left circumflex artery arise separately from the right sinus of Valsalva, each originating from a separate ostium.

Regardless of their low incidence rate, coronary artery anomalies can cause serious technical challenges during coronary angiography and percutaneous interventions because of the unusual location and course of the artery. Echocardiography, computed tomography, and magnetic resonance imaging can be useful in such cases. (Folia Morphol 2023; 82, 4: 932–935)

Key words: coronary arteries, ostia, variation, coronarography, angiogram

INTRODUCTION

The heart's blood supply is usually carried via the coronary arteries (CAs). Normally, there are two of these, the right (RCA) and left (LCA) coronary arteries. The LCA further separates into the left anterior descending (LAD) (anterior interventricular artery) and left circumflex coronary (LCx) artery. The RCA normally originates from the right sinus of Valsalva (RSV), and the LCA from the left sinus of Valsalva (LSV) [10]. Coronary artery anomalies (CAAs) arise from

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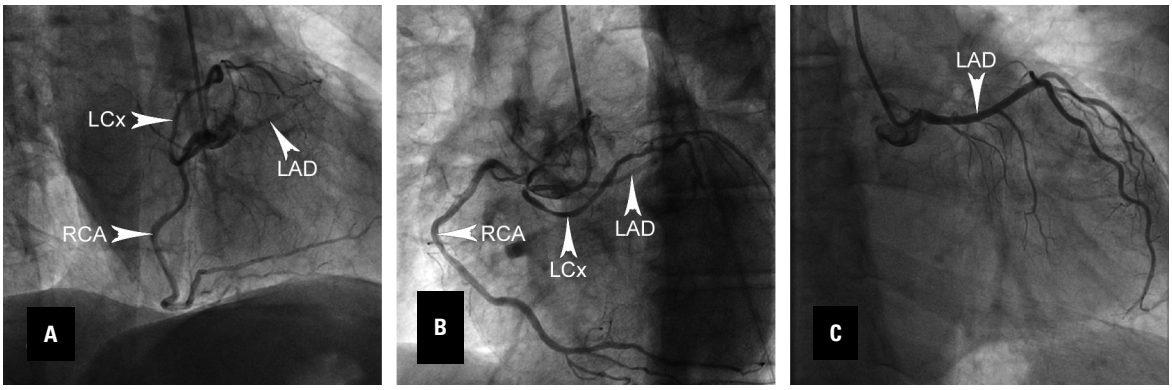

Figure 1. A–C. Coronary angiographic views showing three variant arteries: right coronary artery (RCA), left interventricular (LAD) (anterior interventricular artery) and left circumflex artery (LCx) as separate arteries arising from the right sinus of Valsalva.

a wide diversity of congenital variations in the origin, course and branches of the CA [2]. Such variations are rare, their incidence rate ranging between 0.3% [1] and 3.06% [9]. The vast majority of CAAs are benign and pose no increase in cardiovascular risk, but some rarer CAAs are associated with myocardial ischaemia, congestive heart failure, and sudden cardiac death. Angina can occur because of obstructive coronary artery disease, abnormal origin angulation of the CA, or compression between the aorta and the pulmonary artery. Detailed knowledge of any possible variation of the CA is paramount for all invasive cardiologists and cardiac surgeons to ensure the correct diagnosis and treatment of CA pathology [11].

The aim of the present report is to describe a rare CAA discovered during selective coronary angiography and to highlight the immense importance of recognising such possibilities to preclude complications and unfavourable diagnostic and therapeutic results.

CASE REPORT

A rare anomaly was registered when a 57-year-old male presented with stable angina pectoris for 2 years and no history of syncope. His risk factors were arterial hypertension, dyslipidaemia and smoking. The resting electrocardiography revealed sinus rhythm, left anterior hemiblock and negative T-waves in leads V5–V6. Echocardiography revealed mild symmetric left ventricular hypertrophy, ejection fraction –60%, and no valvular heart disease. The laboratory tests were normal. We considered that the patient had a high clinical likelihood for obstructive coronary artery disease because of typical angina at a low level of exercise that did not affect from optimal medical therapy, and coronary angiography was performed. Initially, the RCA was relatively easily engaged with a 5F JR3.5 diagnostic catheter, and no significant stenosis was visible. Catheterisation of the LCA was then attempted with a 5F JL3.5 diagnostic catheter. After several unsuccessful attempts, and no vessels originating from the left sinus of Valsalva visible, the contrast medium was ejected non-selectively into the RSV, leading to the visualisation of three separate vessels. No left main coronary artery (LCA) was demonstrated. Multiple attempts were made to cannulate the LAD (anterior interventricular artery) and LCX selectively, and we succeeded by using a JR catheter for the LAD (anterior interventricular artery) and an AR1 for the LCx. After thorough and careful catheterisation of each of the three vessels, one of them was found to course to the right margin of the heart and to supply blood to the right atrium and right ventricle; thus, this vessel was named the RCA (Fig. 1A, B). The second vessel descended into the middle of the heart toward the apex, supplying blood to the anterior portion of the interventricular septum; therefore, this vessel was labelled the LAD (anterior interventricular artery) (Fig. 1A–C). The third vessel curved left and posteriorly, surrounding the heart, and was deemed the LCx (Fig. 1A, B). The RCA was dominant and had no stenosis, the LAD (anterior interventricular artery) had moderate stenosis on the distal segment, and the LCx had non-significant stenosis proximally. The patient was discharged with medical therapy and planned for further investigations after 3 months.

DISCUSSION

Anomalies in the origin of the CA are rarely reported. Alexander and Griffith [1], in an autopsy study,

reported a 0.3% mean incidence of CAAs. Lipsett et al. [8], in another multicentred autopsy study, found a 0.5% incidence rate. In an angiographic study of 126,595 patients, Yamanaka and Hobbs [12] reported a 1.3% rate. Yildiz et al. [13] reported a 1% incidence rate in an angiographic study of 12,457 patients. According to the angiographic study by Sidhu et al. [9], the incidence rate of CAAs is 3.06%. Among the various CAAs described, the most common are anomalies in the origin of the LCx and LCA [8, 12, 13]. In contrast, CAAs involving multiple vessels arising from the RSV via separate ostia have been described in only a few case reports [4]. Yildiz et al. [13] reported all the major CAs arising from the RSV in 0.008% of the population. Origins of all three coronaries from the RSV, as in our case, were reported by Asciutto et al. [3] and Chan et al. [5]. Suspicion for this during angiography is based on the detection of an "avascular area" in the anatomical zone of the LCA and the absence of collaterals [3]. In some cases, this extraordinary origin of all three main arteries cannot be detected by angiography, and multidetector computed tomography can be used for correct evaluation of the coronary anatomy [3, 5].

Coronary artery anomalies are most commonly asymptomatic and are discovered by chance or during an autopsy postmortem [1, 6]. Nevertheless, they are clinically important owing to their association with higher cardiovascular risk and the threat of sudden cardiac death [11]. According to Yamanaka and Hobbs [12], CAAs can be separated into two groups according to their origin and course: benign, and potentially serious. An LCx originating from the RSV is classed as a benign variation. However, a LAD (anterior interventricular artery) originating from the RSV is deemed potentially serious [12], especially if it courses between the aorta and the pulmonary trunk, when it is associated with exercise-induced sudden cardiac death [11]. Serious complications could be provoked in such cases if angiography is conducted by an inexperienced interventional cardiologist; it is a technically and logistically demanding procedure. Moreover, the management of patients with such an anomaly is not clear because guidelines are lacking and cases are extremely rare.

Through the literature, several different types of classification have been proposed to provide a detailed and precise depiction of CAAs. Angelini et al. [2] proposed a thorough and sophisticated approach that categorised CAAs on the basis of their origin, course, intrinsic anatomy, termination site and anastomoses. According to this classification, our anomaly should be classified as A4b2c1 [2]. Another classification, more limited and nowhere near as complex, was proposed by Dollar and Roberts [7]. This classification considers only the number of ostia in the coronary sinuses. It has three categories, for one to three ostia in the coronary sinus [7].

CONCLUSIONS

Despite their rarity, anomalies in the origin of the coronary arteries can pose severe diagnostic, technical and therapeutic problems during coronary angiographies. Therefore, detailed knowledge of these variations is essential for the correct diagnosis and treatment of any pathology regarding the coronary arteries.

Conflict of interest: None declared

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