Case reports

Single coronary artery associated with perimembranous ventricular septal defect

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The authors report the case of an adult male patient with a congenital ventricular septal defect who underwent an invasive assessment of his heart disease. Here, the septal defect was identified in the perimembranous area and a pulmonary-to-systemic flow ratio of 1.4:1 was calculated. At coronary angiography, a single coronary artery was found. This particular anomaly consisted of a proximal right coronary artery originating from a septal branch of the left anterior descending coronary artery and a distal right coronary artery arising from the distal left circumflex artery. Such an association constitutes an exceedingly rare congenital condition.

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Introduction

A single coronary artery is a rare congenital anomaly[1-4], described as an isolated finding or in association with other congenital cardiovascular abnormalities[3,5-7]. We report the case of a patient presenting with a single coronary artery and a perimembranous ventricular septal defect (VSD).

Case report

A 49-year-old male with a history of borderline hypertension, cardiac murmur since childhood and an echocardiographic diagnosis of VSD was evaluated at our center because of recent onset of palpitations. At chest auscultation, the patient, with a normal body mass index, was found to have splitting of the second heart sound and a 3/6 holosystolic murmur. A 12-lead electrocardiogram showed normal sinus rhythm and incomplete right bundle branch block. The arrhythmic pattern recorded at Holter monitoring consisted of both supraventricular and ventricular premature beats. Two-dimensional echocardiography showed mild left ventricular hypertrophy with a normal biventricular function and a perimembranous ventricular septal aneurysm with echo drop-out; at Doppler examination, a left-to-right shunt was found at the level of the septal aneurysm, and a pulmonary-to-systemic flow ratio of 1.4:1 was calculated. Cardiac catheterization confirmed the echocardiographic data (Fig. 1). The pulmonary artery pressure and pulmonary resistance were normal. Coronary angiography was performed as part of routine evaluation for male patients > 40 years old. Contrast injection in the left coronary artery opacified the entire coronary vascular bed. The right coronary artery (RCA) consisted of two separate segments. The proximal segment arose from the first septal branch of the left anterior descending coronary artery (LAD) and supplied the upper part of the right ventricle, while the distal segment arose from the distal left circumflex artery (LCx) (Fig. 2). These two vascular conduits both had a blind ending. Contrast injection in the right sinus of Valsalva did not show a right coronary ostium (Fig. 3). There was no angiographic evidence of atherosclerotic lesions. Exercise testing was normal with no evidence of ischemia. The patient was discharged on beta-blockers as symptomatic treatment for the premature beats and prophylaxis against endocarditis was recommended. At 6 months of follow-up he is asymptomatic and in good general conditions.

Discussion

A single coronary artery is a rare congenital anomaly consisting of one coronary artery arising from the aorta through a sin-

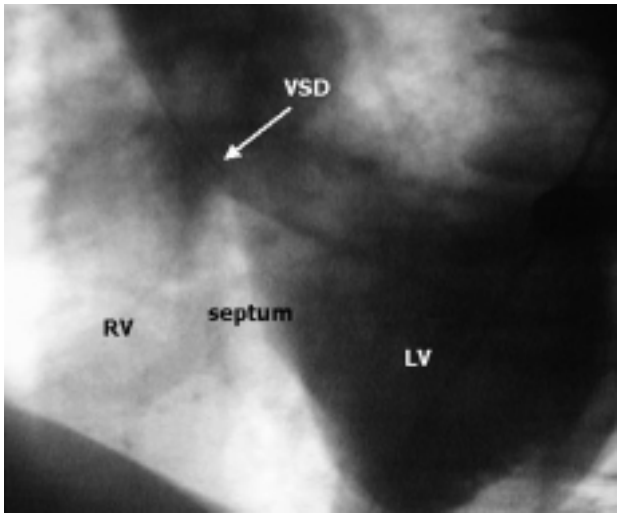

Figure 1. *Left ventriculography in the left anterior oblique cranial view showing the ventricular septal defect (VSD). LV = left ventricle; RV = right ventricle; septum = interventricular septum.*

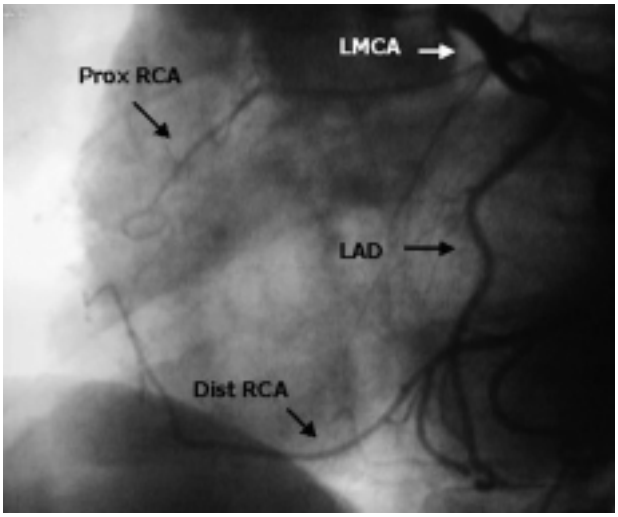

Figure 2. *Left coronary arteriogram obtained in the left anterior oblique view. The mid and distal segments of the right coronary artery (RCA) arise from the distal left circumflex artery. The proximal segment of the RCA arises from the first septal branch of the left anterior descending coronary artery (LAD). The proximal and mid-distal segments of the RCA are not in continuity. There is no evidence of atherosclerotic lesions or truncated stumps. LMCA = left main coronary artery.*

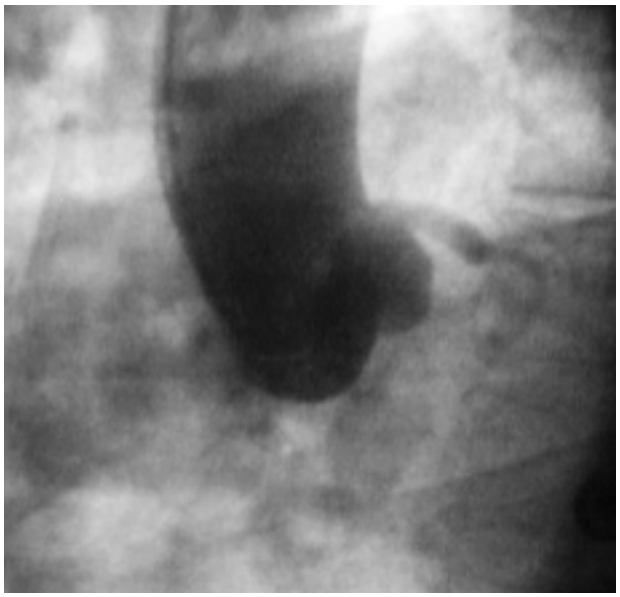

Figure 3. *Aortography in the left anterior oblique cranial view. No truncated stump or ostial calcification of the right coronary artery are identifiable.*

gle coronary ostium and supplying the entire heart, with an estimated incidence in the population undergoing coronary angiography ranging from 0.02 to 0.06%[4,8]. Previous reports found a similar prevalence for right and left ostium absence and no sex-related differences[4,8,9]. In the present report, we found the very rare occurrence of a single coronary artery in which the RCA consisted of two independent and separate vascular segments arising from the left coronary artery. Such an anatomical picture corresponds to class LI of the classification scheme proposed by Lipton et al.[4] and, to our knowledge, has been previously reported in very few cases[10,11]. This angiographic finding raised the suspicion of an acquired, atherosclerotic RCA occlusion at the ostium, with the subsequent development of a double collateral circulation from the left coronary system. Although this issue could not be solved on the basis of angiographic data alone, a few hints suggested that this was not the case. First, the patient had no history of coronary artery disease, no other coronary lesion and no wall motion abnormality of the right ventricular free wall or of the infero-posterior segments of the left ventricle. Second, the right coronary ostium was absent, with a smooth right sinus of Valsalva (Fig. 3). This latter finding was against an acquired RCA occlusion. Third, the angiographic aspect of the left-to-right system was one of a large epicardial vessel, again uncommon in acquired occlusions (Fig. 2).

In about 40% of cases[12], a single coronary artery is associated with other anomalies of the heart and great vessels, such as coronary arterio-venous fistula, bicuspid aortic valve[7] or transposition of the great vessels[13]. The association of VSD and single coronary artery has been reported only in a few cases[5,6,14-18]. In most of them, the single coronary artery was a RCA arising from the left sinus of Valsalva or from the left main stem[5,6,14,17]. In 2 cases, the single coronary artery and the VSD were the only cardiac anomalies present[5,15]; in the remaining, they were associated with other cardiovascular defects, such as an aneurysm of the sinus of Valsalva[14,16], subaortic and subpulmonary stenosis[16,18], tetralogy of Fallot[17], and annuloaortic ectasia[6]. A single coronary artery, alone or in combination with other cardiac anomalies, may present with the clinical features of myocardial ischemia, heart failure and sudden death[3,19,20]. In the absence of other cardiac anomalies,

the risk of severe adverse cardiac events is increased when the single coronary artery consists of a single vessel arising from the right sinus of Valsalva and when the left main stem runs between the aorta and the pulmonary artery (Lipton class RII-B), while the risk is very low when the left main stem is anterior or posterior to the great vessels (Lipton class RII-A and RII-B respectively)[4,5,21]. Conversely, when the RCA originates from the left coronary artery (Lipton class LII-A, LII-B, LII-P), the risk of sudden death or myocardial infarction is generally considered to be negligible[22], although some reports cast doubts on this belief[23-25]. One such report described a case of a single coronary artery free from atherosclerotic disease in which the RCA partly arose from the proximal LAD and partly from the distal LCx (Lipton class LI), as the only abnormal finding in a patient with unstable angina[10]. In this particular case, the mechanism proposed by the authors to explain the occurrence of myocardial ischemia was the "relatively" small size of the single coronary artery, rendering the vessel functionally unable to sustain the blood flow for the whole myocardium[26]. In our case, the absence of symptomatic ischemia and a normal exercise stress test argue against this hypothesis.

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