Case Study

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Congenital coronary artery anomaly simulating a ventricular septal defect

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Abstract

Anomalous origin of the circumflex artery of the right sinus of Valsalva is a rare finding which may be present with other cardiac malformations. A 19-year-old man presented with syncope. A transthoracic echocardiogram revealed discrete subaortic stenosis with a small defect just below the aortic valve, suggesting a ventricular septal defect. Transesophageal echocardiography showed anomalous origin of the circumflex artery from the right sinus of Valsalva. This was confirmed by coronary angiography. The patient underwent successful web resection without concomitant coronary surgery. Failure to demonstrate a coronary artery anomaly can be misleading for surgeons and perilous for patients.

Keywords

Aortic stenosis, coronary vessel anomalies, echocardiography, transesophageal, heart defects, congenital, sinus of valsalva

Introduction

Discrete fibromuscular subaortic stenosis is a ridge or fibrous ring at varying distances from the aortic valve, obstructing the left ventricular outflow tract. Various congenital heart defects are associated with subaortic stenosis, with a prevalence of 25%–60%. The more notable anomalies include ventricular septal defect (VSD) in 6%–25%, patent ductus arteriosus, coarctation of the aorta, bicuspid aortic valve, Shone syndrome, and persistent left superior vena cava.[1,2] We describe the case of a patient with exercise-induced syncope who was found to have subaortic stenosis and an abnormal coronary artery origin. Due to a diagnostic misinterpretation, the abnormality was initially reported as a VSD. We would like to alert cardiac surgeons and cardiologists to this unusual condition which may cause a perilous situation at surgery.

Case report

A 19-year-old man was referred to the emergency department with syncope during strenuous exercise; he was unconscious for nearly 3 minutes. He did not complain of any chest pain, dyspnea, or palpitations before unconsciousness. He had no history of cardiovascular disease or sudden cardiac death in his family. On physical examination, the only abnormal finding was a 3/6 mid-systolic murmur over the right and left sternal borders, without an ejection click. Electrocardiography demonstrated sinus rhythm with inverted T waves in leads II, III, and aVF. A chest radiograph was normal. In view of the systolic murmur on physical examination, 2-dimensional transthoracic echocardiography was performed. This revealed a discrete subaortic web and a tricuspid

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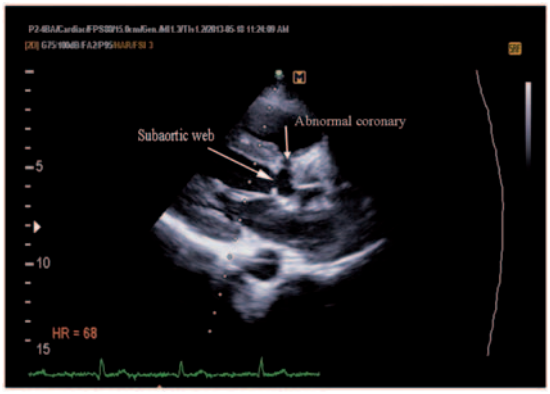

Figure 1. Parasternal transthoracic echocardiogram demonstrating discrete subaortic stenosis and a small dropout defect (abnormal coronary origin) just below the right coronary aortic cusp.

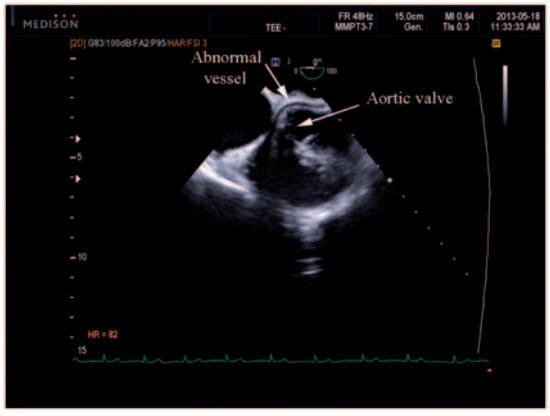

Figure 2. Mid-transesophageal echocardiogram revealing an abnormal vessel course, originating from the right coronary sinus and passing posterior the aortic root and continuing its pathway in left atrioventricular region.

aortic valve with mild aortic regurgitation. Continuous-wave Doppler echocardiography showed a mean left ventricular outflow tract gradient of 60 mm Hg. The left ventricular wall displayed the upper limit of normal thickness. A small defect was observed just below the aortic valve, which was initially suspected to be a VSD (Figure 1). For detailed echocardiographic examination of the left ventricular outflow tract, VSD, and aortic valve, transesophageal echocardiography (TEE) was performed. This demonstrated an anomalous coronary artery arising from the right sinus of Valsalva and passing posterior to the aortic root (Figure 2). Two other coronary arteries were shown to originate from the left and right sinuses of Valsalva. TEE also indicated that the anomalous coronary origin was most probably related to the left circumflex coronary artery (LCX). Further evaluation by coronary angiography revealed that the LCX was not

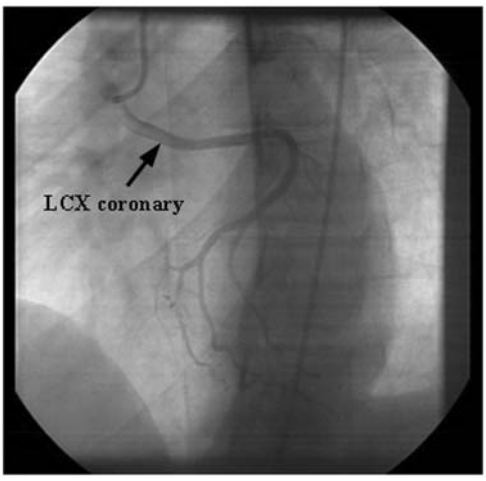

Figure 3. Left anterior oblique projection illustrating the left circumflex artery (LCX) arising from separate ostia in the right sinus of Valsalva.

located at its normal site. The LCX originated from the right coronary sinus, passed posteriorly from the aortic root, and continued to the left atrioventricular region (Figure 3). The right coronary artery originated from the right coronary sinus and coursed its natural track. The patient was operated on successfully and the fibromuscular ring was thoroughly excised. Because the LCX passed posteriorly and there was no compression effect, it was left intact.

Discussion

Depending on concomitant lesions, subaortic stenosis has various presentations, but in isolated forms, patients may be referred with a systolic murmur or signs and symptoms of fatigue, dyspnea, chest pain, or syncope.[3] In this case, the patient was referred with syncope. Coronary artery anomalies are rare, with an incidence of 0.2%–1.2%. Although this anomaly is classified as asymptomatic, it has been reported to provoke myocardial ischemia and even a few cases of sudden death, myocardial infarction, and angina pectoris.[4–6] The increased risk of sudden death may be due to a slit-like ostium, a bend with an acute takeoff angle in the aberrant coronary artery, or arterial compression between the pulmonary trunk and aorta when blood flow through these vessels increases with exercise and stress. Origin of the right coronary artery from the left coronary artery or left aortic sinus with passage between the aorta and the right ventricular outflow tract is also associated with myocardial ischemia and sudden death. In rare cases of anomalous origin of the left coronary artery from the right sinus, myocardial ischemia may occur even if the left coronary artery passes anterior to the right ventricular outflow tract or posterior to the aorta.[7,8]

Although anomalous origin of the LCX from the right sinus of Valsalva is rare and usually has no clinical significance, accurate recognition is very important, as demonstrated in our case, particularly in patients undergoing cardiac valve replacement or subaortic stenosis surgery. The coincidence of fibromuscular subaortic stenosis with a left-sided heart abnormality has been reported. However, its association with abnormal origin of a coronary artery has not been noted so far. Failure to demonstrate the coronary artery anomaly can be misleading, with a perilous outcome for the patient.[9] Knowledge of the abnormal route of a coronary artery helps surgeons during valve replacement and when performing deep sutures, because they should be cautious of damage to the coronary arteries. TEE has been shown to be a valuable technique for determining the anomalous origin of the right coronary artery from the left sinus and the LCX from the right sinus of Valsalva, and their association with the great vessels. In this case, TEE was preformed for better evaluation of the subaortic defect, subsequently identifying an abnormal coronary origin. Coronary angiography is an excellent method for determining coronary artery anomalies and concomitant coronary atherosclerosis, but TEE may play an important role in younger patients.[10] Because the LCX was very close to the web and suspected to pass through the interventricular septum, a coronary angiogram was also performed. Careful evaluation of patients is necessary to determine these concomitant disorders, since overlooking them can prove hazardous for patients.

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Conflicts of interest statement

None declared.

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