Abnormal Origin of Right Coronary Artery From Left Ventricle With Bicuspid Aortic Stenosis

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Abnormal coronary arteries are rare but can lead to serious complications during cardiac operations if not recognized. We report a 55-year-old woman with anomalous origin of the right coronary artery from the left ventricle, and bicuspid aortic valve stenosis. Coronary angiography and computerized tomographic angiography indicated the diagnosis preoperatively. She underwent aortic valve replacement and coronary artery bypass grafting subsequently, with good recovery. One adult and two children with the same right coronary artery anomaly have been reported in the literature. Preoperative diagnosis is essential in these cases, and long-term follow-up is warranted.


Anomalous coronary arteries are rare and therefore often are found only incidentally during conventional coronary angiography, with an incidence of 0.3% to 0.8%. The commonest anomaly is an aberrant origin of the main left or right coronary artery (RCA) from the wrong sinus of Valsalva. Rarely there is a fistula draining into one of the cardiac cavities or a displaced connection, as is seen in an anomalous origin of the coronary artery from the pulmonary artery [1]. However, if not recognized the condition can lead to serious complications during cardiac operations. We are aware of only three previous reports of anomalous origin of the RCA from the left ventricle, in one adult and two infants [2, 3].

We report a 55-year-old woman with a known history of aortic valve stenosis and other comorbidities including hypertension, hyperlipidemia, and depression. Echocardiography showed a calcified and thickened aortic valve with severe aortic stenosis (mean pressure gradient 47 mm Hg, peak pressure gradient 78.1 mm Hg, aortic valve area 0.7 cm²) and mild aortic regurgitation. The remaining valves were normal, as was the left ventricle ejection fraction. Coronary angiography showed a normal left main coronary artery and left anterior descending (LAD), which supplied collaterals to the RCA. There was absence of the RCA ostium (Fig 1). Computed tomographic angiography indicated that the RCA originated from the right coronary sinus, which was separated by a roof of tissue continuous with the right aortic cusp (Figs 2 and 3).

The patient continued to experience mild symptoms at home, with shortness of breath and chest tightness when doing daily chores. Hence, she was electively admitted for aortic valve replacement, with coronary artery bypass grafting being kept in mind. Repeated coronary angiography before her operation showed a flap between the origin of the RCA with retrograde filling of the RCA from LAD through collaterals. There was no connection between the aorta and the RCA.

Eventually, she underwent aortic valve replacement (St. Jude #19 Master series mechanical aortic valve) and coronary artery bypass grafting (saphenous vein graft, RCA). We used retrograde cold blood cardioplegia through the coronary sinus from the beginning. During the operation we found a calcified bicuspid aortic valve, lack of a noncoronary cusp, and origin of the RCA from the left ventricle just below the aortic annulus and sealed up by a membrane but with normal caliber. Postoperatively, the patient recovered uneventfully. Repeated echocardiography showed a normal prosthetic valve function and a left ventricle ejection fraction of 50%.

Comment

One Turkish population study that reviewed seven different studies in seven races showed the incidence of anomalous RCA in patients undergoing coronary

Fig 1. Coronary angiogram showing right coronary artery retrograde supplied by left anterior descending collaterals.
angiography to be between 0.07% and 0.46%. Most of the anomalies were RCA originating from the left sinus of Valsalva [4]. One early autopsy study in 1978 confirmed the association of left dominant coronary system dominance with congenital bicuspid aortic valve. It was suggested that a left dominant coronary system might arise as a consequence of disproportionately decreased blood flow in the left heart chambers, one cause of which was aortic valve stenosis during early cardiogenesis [5]. In 1975, Schang and colleagues [6] first emphasized the association of anomalous RCA origin with a bicuspid aortic valve and the possible clinical consequences of this combination of malformations.

Anomalous origin of the RCA seems to be more common in patients with aortic valve pathologic features, especially bicuspid aortic stenosis. However, most reports have shown the RCA arising anomalously from the left main coronary artery, the left sinus of Valsalva, or the pulmonary artery or as a high takeoff from the ascending aorta [6–8]. To our knowledge, the diagnosis of RCA originating from the left ventricular cavity, just inferior to the aortic valve annulus, has been presented in only three previous case reports. One case was in a 64-year-old woman whose diagnosis was an incidental finding at the time of aortic valve replacement for aortic stenosis. She subsequently underwent bypass grafting [2]. The other two cases were in children. One was a 4-month-old infant with congenital valvular aortic stenosis who underwent surgical aortic commissurotomy in 1995. She died on the operating table when her left ventricle failed to recover at the end of her operation, and the coronary anomaly was discovered at autopsy [3]. Another patient was a 22-month-old boy, who was asymptomatic with a heart murmur that was evaluated by echocardiography and cardiac catheterization. No surgical intervention was performed [7].

Fatal or nonfatal myocardial infarction and sudden death occur in up to 30% of cases of anomalous origin of the RCA in the absence of coronary atherosclerosis [4]. The in vivo identification of anomalous coronary artery disease is still a challenge because of insufficient warning signs. Therefore, liberal criteria for preoperative angiography may be justified. Computed tomographic angiography with three-dimensional reconstruction is helpful to demonstrate the spatial relationship of the anomalous arteries to the aortic root. Cardiac magnetic resonance imaging might be another helpful modality.

Tavaf-Motamen and colleagues [8] advocated RCA reimplantation or bypass grafting with ligation of the RCA proximally to the bypass graft in the repair of anomalous origin of RCA from the left sinus of Valsalva, to prevent graft failure that might be caused by the competitive flow in the native RCA. In our case, the RCA was retrogradely filled by left coronary collaterals, and its ostia was completely sealed by a membrane below the aortic annulus. So far there is a lack of evidence in the literature to show the effectiveness of surgical intervention in this extremely rare RCA origin anomaly; therefore, long term follow-up for this case is warranted.

Fig 2. Computed tomographic angiogram showing origin of right coronary artery from right coronary sinus, separated by a roof of tissue continuous with the right aortic cusp (arrow). (A) Sagittal view and (B) coronal view.

Fig 3. Computed tomographic angiogram with 3-dimensional reconstructions: right coronary artery originating from left ventricle below aortic annulus. (A) Right anterior oblique 10°, cranial 30°. (B) Left anterior oblique 14°, cranial 62°. (AM1 = acute marginal 1; D1 = diagonal; D2 = diagonal 2; LAD = left anterior descending; RCA = right coronary artery.)
References