Anomalous Origin of the Right Coronary Artery: Report of a Case

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Summary: A 63-year-old man was admitted with a complaint of dyspnea. Echocardiography showed severe aortic regurgitation (AR), and moderate mitral regurgitation (MR). Coronary angiography revealed that the right coronary artery (RCA) arose from the ascending aorta with a high takeoff and a significant stenosis at the distal segment of the RCA. Scintigraphy with Thallium showed a transient perfusion defect on the inferior wall. The diagnosis of AR and MR associated with anomalous origin of the RCA and myocardial ischemia was made. After successful catheter intervention for stenosis of the RCA, an operation was performed on the aortic and mitral valve. At surgery, the orifice of the RCA was located above the commissure of the right and left coronary cusps and the shape was obliquely elliptical. The RCA originated at an acute angle from the ascending aorta, and its proximal segment was incorporated in the wall of the aorta. After aortic valve replacement and mitral valve repair, a neo-ostium without unroofing of the intramural segment of the RCA was created at the proximal RCA, and the intima of the RCA was fixed to the intima of the aorta. The patient recovered uneventfully and is doing well without findings of myocardial ischemia at present 40 months after operation.

Key words anomalous origin of the right coronary artery, anomalous origin of the coronary artery, myocardial ischemia

INTRODUCTION

Although congenital bicuspid aortic valve is often complicated with anomalous origin of the right coronary artery (RCA), the RCA that arises ectopically from a different sinus of Valsalva with a tricuspid aortic valve is relatively rare [1,2]. This condition of the RCA was initially regarded as a benign variant of normal anatomy [3]. In 1982, however, Roberts and colleagues [4] demonstrated its clinical significance, showing that anomalous origin of the RCA frequently causes fatal cardiac dysfunction, particularly, sudden death, as is the case with the left coronary artery (LCA). Since then, several reports have also emphasized the possibility of sudden death and myocardial ischemia, and the necessity of surgical treatment in this anomaly.

In this paper, we describe a patient who had anomalous origin of the RCA associated with tricuspid aortic valve and discuss surgical management for anomalous origin of the RCA.

CASE REPORT

A 63-year-old man was admitted to our hospital for a close examination of dyspnea on exertion. The patient had become aware of a cough at night and epigasric discomfort 9 months prior to this admission, and 4 months later, he became orthopneic. On admission, the patient was not dyspneic at rest. His pulse rate was 51/min with a regular rhythm and blood pressure was 120/70 mmHg. A grade 2 to and
Ao murmur at the 4th intercostal space on the left sternal border and a grade 2 pansystolic murmur at the apex were heard. A chest X-ray showed mild cardiomegaly with a cardiothoracic ratio of 0.53 and no pulmonary venous congestion. An electrocardiogram revealed normal sinus rhythm and left ventricular hypertrophy with depression of ST segments on the V5 and V6 leads. Echocardiography showed dilatation of the left ventricle with moderately impaired systolic function, shortening of the aortic cusps with severe regurgitation, and moderate mitral regurgitation (MR) due to annular dilatation. The aortic valve was tricuspid on echocardiograms. Cardioangiography demonstrated severe aortic regurgitation (AR) and moderate MR with moderately impaired LV wall motion, and the RCA arose from the left lateral wall of the ascending aorta with a high takeoff (Fig. 1A). Cannulation of the catheter to the RCA was difficult, and selective coronary angiography revealed that the proximal segment of the RCA ran obliquely for several centimeters until it reached the right atrioventricular groove, and a significant stenosis was found at the distal segment of the RCA (Fig. 1B). The intact left coronary artery was also delineated (Fig. 1C). The patient was asymptomatic but cardiac scintigraphy with Thallium indicated a transient perfusion defect on the inferior wall of the LV. On the basis of these findings, the diagnosis of AR and MR associated with anomalous origin of the RCA and myocardial ischemia caused by a significant stenosis of the RCA was made. After successful percutaneous transluminal coronary angioplasty for stenosis of the RCA, aortic valve replacement and mitral valve repair were scheduled.

At surgery, with the aid of cardiopulmonary bypass, the aortic root was transversely opened. Myocardial protection was achieved by cold blood cardioplegia. The orifice of the LCA was situated in the normal position with a round shape and that of the RCA was located about 2 cm above the commissure of the right and left coronary cusps and the shape was obliquely elliptical. The RCA originated at an acute angle from the ascending aorta and its proximal segment was actually incorporated in the wall of the aorta. The RCA came out to the aortic surface at the midpoint of the right coronary sinus,

Fig. 1. (A) An aortogram showing severe AR and the RCA with a high takeoff (arrow) from the ascending aorta. (B) A selective right coronary arteriogram showing the proximal RCA that runs obliquely until it reaches the right atrioventricular groove and a significant stenosis (arrow head) of the distal RCA. (C) A selective left coronary arteriogram showing the intact LCA.
Fig. 2. A schema of operative findings. (A) The orifice of the RCA (OR) was located about 2 cm above the commissure of the right (R) and left coronary (L) cusps and the shape was obliquely elliptical. The RCA originated at an acute angle from the ascending aorta and its proximal portion (dot lines) was actually incorporated in the wall of the aorta. The orifice of the LCA (OL) was situated in the normal position. (B) After aortic valve replacement with a mechanical valve, the neo-ostium (Or) without unroofing of the intramural segment of the RCA was created at the proximal RCA, and the intima of the RCA was fixed to the intima of the aorta.

Fig. 3. Postoperative right coronary arteriogram showing no anastomotic stenosis between the proximal RCA and aorta and a 25% stenosis of the distal RCA.

and then it continued to the atrioventricular groove. In addition to the significant stenosis of the distal RCA, these anomalies appeared to be the causes of myocardial ischemia in this patient. Therefore, after aortic valve replacement with a mechanical valve and mitral valve repair, a neo-ostium without unroofing of the intramural segment of the RCA was created by excising the common wall between the aorta and RCA at the proximal RCA, and the intima of the RCA was fixed to the intima of the aorta with fine monofilament suture [5], as shown in Fig. 2. The patient recovered uneventfully, and postoperative coronary arteriography demonstrated opacification of the whole RCA through the neo-ostium without anastomotic stenosis (Fig. 3). In a recent follow-up, 40 months after operation, the patient is doing well with no evidence of myocardial ischemia.

DISCUSSION

Although congenital bicuspid aortic valve is often complicated with anomalous origin of the coronary arteries, the RCA that arises ectopically from a different sinus of Valsalva with a tricuspid aortic valve is relatively rare [1,2]. Among anomalous origins of the coronary arteries, ectopic origin of the RCA from the left sinus of Valsalva is a relatively rare coronary anomaly. The incidence of patients with the RCA that arises from a different aortic sinus is only 0.2% of the angiographic population [2].

Based on a study of 32 necropsy cases [6], anomalous origin of the coronary arteries has been classified into two groups. Anomalous origin of the LCA is classified as group 1 and that of the RCA is included in group 2. Since the origin of the RCA was located above the commissure of the right and left coronary cusps in the present patient, this case is classified into group 2C.

Initially, these variations of origin of the coronary artery were generally considered to be minor congenital anomalies [3], in 1982, however, Roberts [4] reported clinical and autopsy findings in patients in whom anomalous origin of the RCA had caused sudden death, and emphasized that this condition is not such a benign congenital anomaly as had been
supposed. Since then, several reports have described cases of sudden death and myocardial ischemia in this anomaly [5,7,8]. While sudden death is clearly more common in the group 1 anomalies than the group 2 anomalies, the latter condition should be recognized as a potential cause of fatal myocardial ischemia.

The detailed mechanism of cardiac dysfunction caused by anomalous origin of the RCA from a different sinus of Valsalva has been a topic of considerable controversy. Three theories, such as compression of the RCA between the aorta and pulmonary trunk during exercise [9], acute angled origin of the RCA from the aorta resulting in a flap-like mechanism at the coronary ostium [8], or coronary dominance rather than the shape of the ostium [6], have been proposed. Among these theories, the acute angle at the ectopic origin with flap-like mechanism is most favorably accepted. Cheitlin and colleagues [8] suggested that in cases of the RCA with an acute angle at ectopic origin, expansion of the aorta during exercise in combination with the intramural course of the ectopic artery could easily lead to obstruction of the already slit-like ostium. In the present patient, a slit-like ostium with valve-type ridge was not confirmed at the operation, but the RCA left the aorta at an acute angle and the proximal RCA was incorporated in the aortic wall. In addition to stenosis of the distal RCA, these anomalies of the proximal RCA appeared to be the causes of myocardial ischemia in this case.

Treatment for the RCA that arise ectopically from a different sinus of Valsalva has varied. Surgical approaches have included coronary artery bypass grafting [10], reimplantation into the correct sinus from outside the aorta [10], and modification of the orifice of the anomalous RCA at the aorta by excising the common wall between the RCA and coronary artery [5]. This unroofing procedure was originally employed for repair of the LCA that arises from the right sinus by Mustafa and colleagues [11], and has been adopted by many surgeons with good immediate results. In 1997, Jacques and colleagues [12] operated on 2 patients with anomalous origin of the coronary artery with the modified unroofing technique and emphasized the simplicity and effectiveness of this modified technique. This modified unroofing technique includes creation of the neo-ostium without unroofing of the intramural segment of the coronary artery in the appropriate sinus. We also used the modified unroofing technique for surgical treatment of this patient.

In conclusion, we reported a case of anomalous origin of the RCA in which the RCA arose from above the commissure of the right and left cusps, and also describe surgical treatment of this anomaly. We believe that the creation of the neo-ostium from inside the aorta is very simple and effective for repairing this condition because this procedure widens the coronary orifice and prevents compression of the intramural coronary artery segment.

REFERENCES