Anomalous Right Coronary Artery Arising from Left Coronary Sinus in Two Brothers

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We report two brothers aged 61 and 69 respectively with abnormal right coronary artery (RCA) originating from the left sinus of Valsalva. Both of them presented with exertional dyspnea and chest tightness in their seventh decade. Another younger brother of theirs suffered from sudden cardiac death at the age of 60. Other symptomatic family members were screened by multi-slice computed tomography (MSCT), but no coronary artery anomalies were found. Considering different risk factors of these two brothers, one patient underwent coronary artery bypass surgery, while another took medical therapy. Previously reported familial cases of anomalous coronary arteries are very rare.

Key Words: Anomalous coronaries • Computed tomography • Familial clustering

INTRODUCTION

Coronary artery anomalies are reported in 1.3% of patients undergoing coronary angiography, and may be associated with sudden death, myocardial ischemia, arrhythmia and syncope. Some anatomic presentations of coronary anomalies are considered to be high-risk group. However, many patients were asymptomatic before their presentation of sudden cardiac death, indicating that early detection of potential lethal cases is difficult. Familial cases of anomalous coronary arteries have been very rare. In this case report, we report two brothers with abnormal right coronary artery (RCA) origin from the left coronary cusp with identical courses of RCA, and we suggest clinicians pay attention to the possibility of familial clustering of coronary anomalies.

CASE REPORT

A 61-year-old man (patient 1) presented to our institution because of exertional dyspnea and chest distress. He had hypercholesterolemia and was a casual smoker. He received treadmill exercise test, which revealed ST segment depression of 1 mm at leads V4-V6 during the stage III exercise. Exercise thallium-201 myocardial perfusion scan with dipyridamole also demonstrated reversible perfusion defect in the proximal inferior walls of the left ventricle. Cardiac catheterization showed aberrant origin of the right coronary artery (RCA) from the left coronary cusp. There were also mild atherosclerotic changes in the coronary arteries (Figure 1A). Multi-slice computed tomography (MSCT) proved the diagnosis of anomalous RCA arising from the left sinus of Valsalva and taking an interarterial course between the aorta and pulmonary artery (Figures 1B&C). The patient received anti-atherosclerotic medication and his condition remained stable during 3 years of follow-up.

A 69-year-old brother (patient 2) of patient 1 was admitted to our hospital 2 years after his brother’s coronary examination. Another brother of the two had died suddenly 3 months previous to this hospitalization at his age of 60, but the diagnosis was uncertain. Patient 2 suf-
fered increasing shortness of breath and dyspnea on exertion for months. His past medical history were significant for hypertension and dyslipidemia. Treadmill exercise test showed marked 2 mm ST segment depression at inferior leads and leads V4-V6 during the stage III exercise and recovery phases. Exercise thallium-201 myocardial perfusion scan with dipyridamole demonstrated significant myocardial ischemia involving the left ventricular anteroseptal wall and apex. Cardiac catheterization showed total occlusion at the proximal portion of the left anterior descending artery (LAD) and tortuous atherosclerotic left circumflex artery. The RCA arose from the left coronary cusp, and a narrow orifice was suspected (Figure 1D). MSCT revealed abnormal orifice of the RCA from the left coronary sinus, with interarterial course and narrowed ostium (Figures 1E&F).

Coronary artery bypass surgery was performed and direct compression of the RCA proximal portion by the pulmonary artery was found during the operation. Therefore, the LAD and RCA were revascularized. The patient was in stable status during 6 months of follow-up. Other symptomatic family members were screened by MSCT, but no coronary artery anomalies were found.

DISCUSSION

Coronary artery anomalies were reported in 1.3% of patients undergoing coronary angiography by Yamanaka.1 Important clinical problems of coronary anomaly are sudden cardiac death, myocardial ischemia, arrhythmia and syncope. Sudden death mostly happens before 35

![Figure 1](image-url)
years of age. Increased risk of sudden cardiac death with this anomaly has been associated with four risk factors: interarterial course between the aorta and pulmonary artery; slit-like coronary orifice; acute angle of take-off of the anomalous coronary artery from the aorta; and the presence of aortic intramural coronary arteries. Symptoms premonitory to a fatal event, such as exertional syncope, chest pain or palpitations, are common in patients at risk, so surgical correction is indicated in symptomatic patients at any age. However, only about 20% of fatal anomalous coronary artery cases had prodromic symptoms. RCA arising from the left coronary sinus is more common than left coronary artery (LCA) from the right sinus. In Yamanaka’s report, RCA arising from the left coronary cusp was noted in 0.17% of coronary angiographies, while LCA from the right coronary cusp was seen in 0.047%. Data from Garg reported that 15 out of 4100 patients (0.37%) had abnormal orifice of RCA from the left sinus of Valsalva, but only 1 patient (0.02%) had LCA from the right sinus. However, the clinical course of LCA from the right coronary sinus is more malignant than that of RCA from left coronary sinus. Basso reported 27 sudden deaths in young athletes. Of these, 23 had LCA arising from the right coronary sinus and only 4 had RCA from the left coronary sinus. Thus, surgical repair is recommended in patients with anomalous LCA. Nevertheless, treatment in patients with anomalous RCA is still controversial because most conditions of this disease are benign. MSCT can be used to evaluate the orifice and the course of anomalous coronary artery, which can guide the treatment policy.

Familial clustering of anomalous coronary arteries was only described in five previous reports. Devanagondi reported one 10-year-old boy with aberrant origin of LCA from the right sinus of Valsalva presenting with sudden cardiac death, while his 8-year-old brother, who had aberrant RCA from the left coronary sinus with interarterial course and slit-like orifice, was asymptomatic. Un-roofing operation was performed for this 8-year-old boy, with good short-term outcome. Laureti demonstrated two brothers with anomalous LCA from the right coronary sinus. The junior was found to have interarterial course and was treated by reimplantation of LCA, while the senior with retroaortic course was treated medically. Horan described a father with abnormal origin of RCA from the circumflex artery and his daughter with abnormal LCA arising from the RCA. In a study of combined coronary and perfusion cardiovascular magnetic resonance imaging, Bunce demonstrated two sisters having anomalous coronary arteries. One had abnormal RCA arising from the left coronary cusp, and the other sister had single coronary artery arising from the right sinus of Valsalva. Rowe reported anomalous origin of the left circumflex artery in a father and his son and daughter. In these previous reports, only one mortality was noted. A brother of our index cases suffered sudden cardiac death, but no autopsy was done, so the definitive diagnosis was undetermined. Comparing to other reports, our patients also had atherosclerotic coronary disease in addition to coronary anomalies, making the cause of clinical symptoms uncertain. Patient 2 had several risk factors: severe atherosclerotic coronary artery disease, narrow RCA orifice, interarterial course of RCA, and suspected family history of sudden death. Surgical intervention thus is reasonable for him. The treatment of patient 1 is relatively questionable. Since most anomalous RCA with interarterial courses are benign, medical treatment was decided.

According to our cases and other previous reports, we believe that familial clustering of coronary anomalies does exist. Recent study demonstrated that gene Tbx1 defect is associated with anomalous coronary artery in mice, though definitive gene of humans is unknown yet. In this case report, we suggest that if family members of a patient with coronary anomalies present with exertional syncope or chest pain, the possibility of familial clustering of anomalous coronaries must still be kept in mind. Coronary angiogram or MSCT can be arranged in symptomatic family members with positive stress test.

REFERENCES

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