

Left Main Coronary Artery Arising From the Right Sinus of Valsalva in a Patient with Apical Hypertrophic Cardiomyopathy

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Abstract

Coexistence of left main coronary artery emerging from the right sinus of Valsalva with Coronary artery-left ventricular fistulae and apical hypertrophic cardiomyopathy in the same patient has not been reported before. An apparently healthy 55-year-old male was referred for evaluation of atypical chest pain. Electrocardiography, computed tomography angiography and echocardiography confirmed the diagnosis of apical hypertrophic cardiomyopathy. Coronary angiography and computed tomography angiography demonstrated left main coronary artery arising from the right sinus, with left main coronary artery taking a course anterior to the pulmonary artery. The patient received treatment using a calcium channel blocker and stayed asymptomatic until the end of six months of follow-up.

Keywords: Congenital anomaly; Coronary angiography; Computed tomography; Hypertrophic cardiomyopathy

Introduction

The anomalous origin of the left main coronary artery (LMCA) emerging from the right sinus of Valsalva accounts for only 1.3% of coronary anomalies and is found in 0.017–0.03% of patients undergoing coronary angiography [1,2]. However, it also has the potential for life-threatening presentations due to intermittent ischemia and sudden cardiac death (SCD). Apical hypertrophic cardiomyopathy (AHCM) is a unique form of hypertrophic cardiomyopathy (HCM) with left ventricular hypertrophy confined predominantly to the cardiac apex, and ≤ 25% of individuals can develop significant late cardiovascular morbid events [3,4]. Coexistence of these two clinical entities (both of which are known to independently increase the likelihood of SCD) in the same patient has been reported rarely [5].

Case Report

An apparently healthy 55-year-old male was referred for evaluation of discomfort on exertion and relieved by rest of one-month duration. Medical history included uncontrolled hyperlipidemia and smoking two packs of cigarettes a day for 20 years. He denied having a family history of heart disease.

Upon examination, his blood pressure was 110/80 mmHg and heart rate was 62 beats per minute (bpm) with a regular rhythm. His weight was 50 kg and height 160 cm. No extra heart sounds or murmurs were heard.

Laboratory evaluation was unremarkable, without elevation of troponin-I levels in serial measurements. Resting electrocardiography (ECG) showed sinus bradycardia with significant left ventricular hypertrophy (RV5 = 41 mm; RV5 + SV1 = 63 mm), giant T-wave inversions in V3–V4 (>10 mm), and diffuse absence of septal Q-waves in leads I, II, III, aVF, aVL, and V3 to V6. ST-segment depression (>1 mm) can also be observed in precordial leads V3 to V6 (Figure 1).

Subsequent coronary angiography demonstrated a dominant right coronary artery (RCA) and an anomalous left coronary artery arising from the right sinus of Valsalva with a separate ostium from the RCA, and no significant stenosis could be observed in any of the coronary arteries (Figures 2A and 2B). Coronary artery-left ventricle fistulae arising from a diagonal branch of the left anterior descending coronary

artery was identified (Figure 2C). Computed tomography angiography (CTA) revealed that the left coronary artery took a course anterior to the pulmonary artery (Figure 2D) and the typical diastolic spade-like configuration of the left ventricular cavity (Figure 2E). Upon two-dimensional echocardiography, an apical four-chamber view of the left ventricle revealed thickening of the wall of the left ventricle confined to the most distal region of the apex. Maximum apical wall thickness was 18.2 mm (Figure 2F) with an ejection fraction of 0.78. Follow-up 24-h Holter monitoring showed only isolated ventricular ectopic

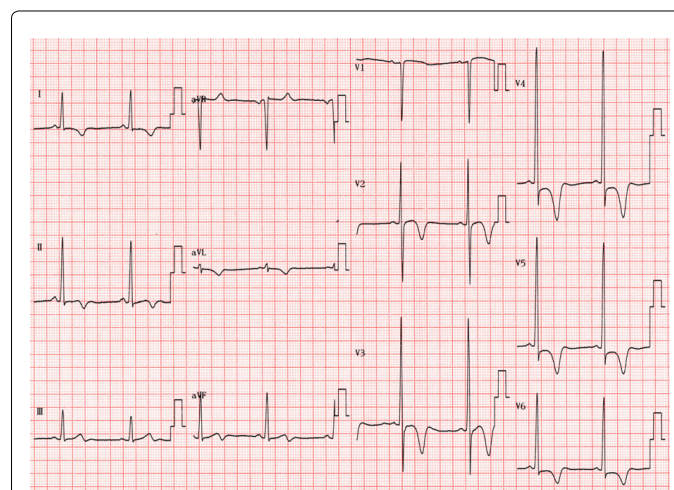


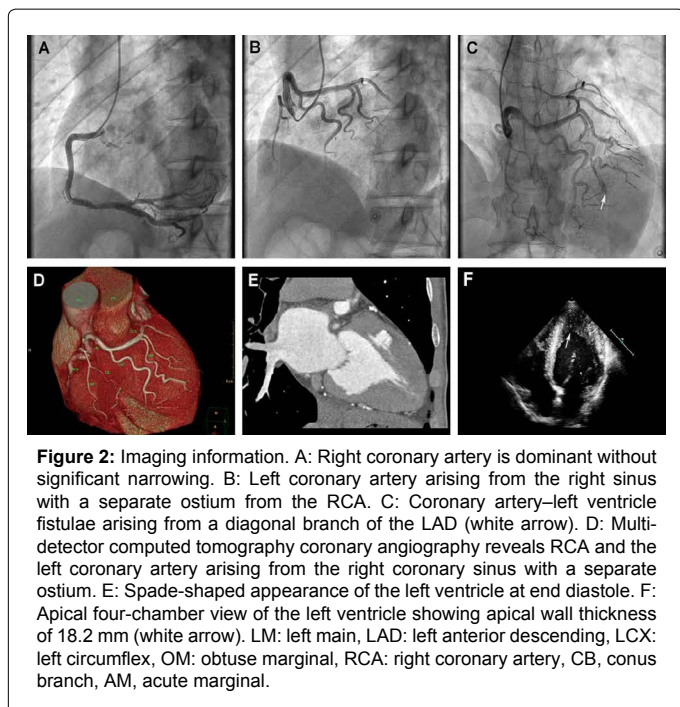
Figure 1: Resting electrocardiography. Resting 12-lead electrocardiogram demonstrating significant left ventricular hypertrophy (RV5 = 41 mm; RV5 + SV1 = 63 mm) and giant T-wave inversions in V3–V4 (>10 mm).

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beats. The patient received treatment using a calcium channel blocker (diltiazem hydrochloride sustained-release capsule, 90 mg/day). He stayed asymptomatic until the end of the sixth month of follow-up.

Discussion

This case illustrates an incidental finding of an anomalous origin of LMCA arising from the right sinus of Valsalva coexistent with coronary artery-left ventricular fistulae and AHCM.

An independent origin of the LMCA and RCA from the right aortic sinus is an exceedingly rare anomaly [1]. Anomalies associated with clinical sequelae involving coronary ischemia and SCD are dependent upon the course of the LMCA. A course of the LMCA between the aorta and pulmonary artery (Type 2A) can be responsible for myocardial ischemia and SCD, whereas a course anterior to a pulmonary artery (type I), such as in our case, is relatively benign [6].

AHCM, first described in Japan, constituted 13% to 25% of all cases of HCM in Japan [7,8]. However, it is not restricted merely to the Japanese population [9]. The proportion of patients with AHCM has been reported to range from 30% to 41% of the total HCM population in Asians [3,10,11]. With regard to morphology, AHCM can be divided into two subgroups based on whether asymmetric apical hypertrophy ("pure AHCM") is present or whether co-existent hypertrophy of the interventricular septum ("mixed AHCM") is present. Pure AHCM is the most common form [4,12].

Coronary artery-left ventricular fistulae are uncommon. Their coexistence with AHCM has been reported in only a few cases [12,13]. The clinical significance of coronary artery-left ventricular fistulae is dependent upon their size and location [12,13].

The main presenting symptom of our patient was chest pain. This symptom could be due to: increased demand for oxygen by the hypertrophied ventricular myocardium; increased diastolic filling pressures leading to impaired coronary flow; small-vessel disease

within the myocardium; coronary fistulae resulting in coronary steal and left-to-left shunt reducing ventricular perfusion and increasing diastolic volume overload [14].

Optimal treatment for AHCM is guided by clinical status. In general, calcium antagonists are the preferred therapy in AHCM patients with normal systolic function and impaired relaxation.

Unlike other variants of HCM, the prognosis of AHCM is relatively benign. The cardiovascular mortality and morbidity of pure AHCM (as in our patient) has been reported to be 0.8% and 17.1%, respectively, which are significantly lower than 15.0% and 56.7% for mixed AHCM after a follow-up of 7 years [10]. Some AHCM patients may develop potentially life-threatening complications such as myocardial infarction, arrhythmia and stroke. Hence, close long-term follow-up is essential to reveal and manage potentially fatal complications (particularly in the elderly) promptly.

Conclusion

We reported a rare case of LMCA arising from the right sinus and coronary artery-left ventricular fistulae in a patient with AHCM. This case seems worthy of notice due to its extreme rarity, and on account of it being less benign than suspected previously.

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