Triple trouble: single anomalous coronary artery origin and coronary artery disease in a patient with hypertrophic cardiomyopathy

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Abstract

Anomalous coronaries occur in <1% live births. Single coronary artery origin is rare. We present such a case in setting of hypertrophic cardiomyopathy and discuss the clinical implications. A 49-year-old African-American hypertensive male, with mild mental retardation presented with angina, was hemodynamically stable, had mild tachycardia. Initial electrocardiogram suggested left ventricular hypertrophy, antero-septal ischemia. Echocardiogram revealed normal valves, left ventricular ejection fraction (70%) without wall motion abnormality; though a small left ventricular cavity, asymmetrical left ventricular septal hypertrophy (posterior wall to interventricular septum ratio-1.3:2); systolic anterior motion of mitral valve, moderate eccentric mitral regurgitation; and restrictive pattern on mitral flow Doppler. Troponins peaked at 0.29ng/mL. Coronary angiogram showed single coronary artery arising from right sinus with 50% stenosis at diagonal origin. Fractional flow reserve was 0.95 and did not warrant angioplasty. Pullback pressure gradient in left ventricular mid-cavity was 19mmHg. He was managed conservatively for the noncritical coronary artery disease, hypertension, dyslipidemia, gastroesophageal reflux disease.

Both hypertrophic cardiomyopathy and single coronary artery can result in sudden cardiac death. Myocardial ischemia can further increase risk of sudden cardiac death. Angina is often a presenting symptom. Tailored management strategies must be pursued.

Keywords: Hypertrophic cardiomyopathy; Single coronary origin; Coronary artery disease; Chest pain; Sudden cardiac death

Introduction

Hypertrophic cardiomyopathy (HCM) and a single coronary artery are both relatively uncommon conditions; but each in itself has the potential to cause ischemia and sudden cardiac death. Their occurrence together in a single patient is extremely rare though reported in literature [1-5]. Hypertrophic cardiomyopathy can present with anginal chest pain or acute coronary syndrome due to demand-supply mismatch of coronary blood supply. This can be due to associated atherosclerotic coronary artery disease (CAD), small vessel disease [6,7] or secondary to coronary anomalies with potential for causing ischemia. Management of these patients can be challenging. Awareness of this possible rare association may be of use to the physicians. Here, we report such a case of HCM with a single coronary artery detected in a patient presenting with chest discomfort and associated coronary artery disease.

Case report

A 49-year-old African-American gentleman, with a history of mild cognitive impairment since childhood, hypertension on hydrochlorothiazide, presented with complaints of recent onset intermittent exertional chest discomfort and throat pain (7/10 severity) lasting for a few minutes. He denied shortness of breath, palpitations, dizziness, or any other symptoms. At admission, he was hemodynamically stable with mild tachycardia at 110s -120 beats per minute. Initial ECG suggested left ventricular hypertrophy, with antero-septal non-ST elevation myocardial infarction (NSTEMI) based on ST-T changes; there was sinus tachycardia with heart rate of 113bpm and QTc 477msec. Trans thoracic and transesophageal echocardiograms revealed normal valves. LV cavity was small with asymmetrical left ventricular septal hypertrophy with a posterior wall to Interventricular septum ratio of 1.3:2. Systolic anterior motion of mitral valve, moderate eccentric mitral regurgitation; and restrictive pattern on mitral flow Doppler. Troponins peaked at 0.29ng/mL. Coronary angiogram showed single coronary artery arising from right sinus with 50% stenosis at diagonal origin. Fractional flow reserve was 0.95 and did not warrant angioplasty. Pullback pressure gradient in left ventricular mid-cavity was 19mmHg. He was managed conservatively for the noncritical coronary artery disease, hypertension, dyslipidemia, gastroesophageal reflux disease.

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diameter on nitroglycerine injection. Fractional flow reserve of the lesion was 0.95 and it did not warrant angioplasty. Despite several attempts at various levels in all the sinuses, the right coronary artery could not be cannulated. There was no vessel filling in conventional right coronary artery course favored the diagnosis of single coronary. The pullback pressure gradient in LV mid-cavity was 19 mmHg, which was opined as not significant. Patient was managed conservatively for noncritical CAD, hypertension, dyslipidemia, gastroesophageal reflux. He

The serial troponins showed mild elevation from 0.09 to a peak of 0.29 ng/mL. Computerized tomography pulmonary angiogram with venous run-off was negative for pulmonary embolism. Coronary angiogram with left heart study showed a single left coronary artery origin arising from right sinus with a 50% discrete stenosis at the origin of Diagonal 2 branch and diffuse disease of left anterior descending artery (LAD) in its mid-course (Figure 2A and 2B). There was no change in lesion

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**Figure 1A.** Dimensional transthoracic echocardiogram showing septal hypertrophy, small left ventricular cavity.

**Figure 1B.** Mild mitral regurgitation shown in transthoracic echocardiogram in color mode imaging.

**Figure 1C.** Transthoracic echocardiogram in M mode showing systolic anterior motion of the mitral valve.

**Figure 2A.** Coronary angiogram in antero-posterior cranial view showing single artery origin; from the right coronary cusp. Non-critical coronary artery disease at origin of Diagonal 2 and mid left anterior descending artery.

**Figure 2B.** Coronary angiogram in right anterior oblique caudal view showing the same findings.
was given extended release metoprolol for HCM. Genetic testing and counseling was performed as an outpatient, and he was tested negative for Noonan’s syndrome. Although magnetic resonance imaging (MRI) was planned as an outpatient, it could not be done due to patient non-compliance.

**Discussion**

Occurrence of anomalous coronaries has been reported in <1% live births [8]. Coronary anomalies may result in myocardial ischemia due to the course in relation to the aorta and pulmonary arteries. Single coronary artery is a potentially serious anomaly [9] that refers to the common origin of left and right coronary arteries from a single aortic ostium. In an analysis of 50,000 angiograms by Desmet et al, the incidence of isolated single coronary artery was 0.066% [10]. Forty percent of single coronary artery cases are associated with congenital heart diseases, such as Fallot tetralogy, transposition of great arteries, persistent truncus arteriosus, and pulmonary atresia [11]. Though rare, association of HCM with single coronary artery has been reported [1,2].

Hypertrophic cardiomyopathy is also an uncommon disease estimated to occur in 0.2% of a general population by echocardiographic screening. Ischemic chest pain is not an uncommon presenting symptom in HCM. Angina in HCM can occur from severe systolic narrowing of epicardial coronaries or major branches (myocardial bridges) [12], and coronary demand-supply mismatch. Associated atherosclerotic coronary artery disease can further complicate the situation. There can be significant obstructive atherosclerotic coronary artery disease (19%), small vessel disease or coronary anomalies with potential for causing ischemia [6,7].

Myocardial ischemia is an important contributor to sudden cardiac death (SCD) in patients with HCM [13]. Our patient presented with a NSTEMI and was found to have HCM, single coronary artery as well as atherosclerotic coronary lesion in the culprit vessel. Given the non-critical atherosclerosis in a distal location in LAD, angioplasty or myomectomy were not warranted. Non-significant atherosclerotic lesions in presence of NSTEMI in the presence of HCM is not surprising, as it has been reported by others [14,15]. Ten-year survival in CAD in HCM patients has been reported as 46.1%, 70.5% and 77% for severe, mild-moderate and no CAD, respectively.16 Cardiac computed Tomographic angiography or MRI may be utilized to delineate the precise coronary anatomy noninvasively and help understand the mechanism of coronary ischemia in setting of coronary artery anomalies.

Both HCM and single coronary artery origin can independently increase risk of SCD. The role of Implantable cardiac defibrillators (ICD) in such situations is unclear. Guidelines recommend ICD implantation in HCM if there was a prior cardiac arrest or sustained ventricular tachycardia. It is reasonable to consider ICD in those with HCM and unexplained syncope, massive LV hypertrophy or positive family history of SCD. Patients with HCM may be referred for septal modification therapy with alcohol septal ablation or surgical myectomy for symptomatic obstruction if medical therapy fails. However, anomalous coronary artery origin may increase the procedural risk of alcohol septal ablation. Depending on the coronary artery anomaly type, the management strategies include observation, coronary angioplasty with stent deployment and surgical repair (unroofing/bypass). Need for revascularization of anomalous coronary depends on presence of high risk features such as slit like orifice, acute angulated origin, intramural course within aorta, or interarterial course due to compression between aorta and right ventricular outflow tract or pulmonary vasculature.

This rare case highlights the need to assess the resting LV mid cavity gradient at rest, detailed coronary anatomy and atherosclerotic burden when a patient with HCM presents with chest pain. A tailored approach in management of such patients is mandatory. It is worth looking at any possible association of single coronary artery and some forms of HCM.

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**Conflict of Interest**

None to declare by any of the authors

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**References**


