

Coronary artery fistulae in patients with hypertrophic cardiomyopathy

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Abstract

Myocardial ischemia with multifactorial pathogenesis is frequently noted in hypertrophic cardiomyopathy (HCM). Coronary artery fistulas (CAF) may be found in HCM but their contribution to myocardial ischemia has not been studied. From 1984 to 2022, 43 patients with HCM and CAF (age 6–82 years, mean 58; 58% male; 77% apical variant; 12% obstructive) were reported in medical literature. Single and multiple CAF were reported in 24 (56%) and 19 (44%), respectively. Overall, a total of 70 major epicardial coronary arteries were affected. CAF arose from left main (LMCA) [1 (1%)], left anterior descending (LAD) [35 (50%)], left circumflex (LCx) [13 (19%)], right (RCA) coronary artery [20 (29%)], and either LAD or LCx [1 (1%)]. The receiving site for CAF was left ventricle (LV) [51 (73%)], pulmonary artery (PA) [10 (14%)], right ventricle (RV) [6 (9%)], both PA and RV [1 (1%)], both left atrium (LA) and PA [1 (1%)] and unspecified in 1. Overall, 28 (65%) patients presented with chest pain, and 14 (33%) had objective evidence of myocardial ischemia (troponin release or positive stress test). Medical therapy was offered to 26 (60%) and 2 (5%) underwent percutaneous or surgical interventions. Follow-up was available for 24 (56%) patients, of whom 23 improved symptomatically while one continued to have episodic chest pain. CAF may be present in patients with HCM, particularly the apical phenotype, and may potentially contribute to myocardial ischemia in this disease. Symptomatic improvement is generally expected in most cases after medical therapy or intervention.

Keywords: Hypertrophic cardiomyopathy, Apical hypertrophic cardiomyopathy, Coronary fistula, Myocardial ischemia

Abbreviations: CAD: Coronary Artery Disease; CCAA: Congenital Coronary Artery Anomalies; HCM: Hypertrophic Cardiomyopathy; SCA: Sudden Cardiac Arrest

Introduction

Hypertrophic cardiomyopathy (HCM) is the most common congenital heart disease affecting approximately 1 in 500 individuals [1]. HCM is an umbrella term divided into smaller morphologic variants including asymmetric septal, concentric, reverse septal, neutral, and apical [2]. Independent of the HCM variant, patient presentation will vary from asymptomatic found incidentally to symptomatic with exertional dyspnea, angina, palpitations or syncope. Myocardial ischemia is a relatively common finding in HCM and is often attributed to microvascular disease, extravascular compressive forces, reduced capillary density, reduced coronary flow reserve, and concomitant atherosclerotic coronary artery disease [3]. Other potential etiologies of myocardial ischemia in HCM such as congenital coronary artery anomalies including coronary artery fistulae (CAF) have not received much attention [4]. In this brief report, we aimed to review and summarize the potential contribution of CAF to myocardial ischemia in adults with HCM as presented in case reports in the world literature.

Methods

Relevant literature on HCM and coronary fistula was found using online databases, including PubMed, Google Scholar, and the Temple University Library search engine. We identified 38

published case reports and series, highlighting 43 patients with HCM and CAF. Cases involving the formation of coronary fistula from traumatic injuries or surgical procedures were excluded. The patient demographics, clinical descriptions, physical examination findings, diagnostic methods, management strategies, and outcomes of each case were reviewed in detail.

Results

Apical HCM accounted for 77% (n=33) of the 43 patients with CAF while 12% (n=5) demonstrated either mid-cavitary (n=2), left ventricular outflow tract (n=2) or unspecified (n=1) obstructive phenotypes. The remaining 5 patients were broadly categorized as non-obstructive HCM. The mean age of the patients was 57.8±15 years, 58% were male and 65% presented with the chief complaint of anginal chest pain. The presence of ischemia, as shown by typical troponin rise and fall and/or a positive stress test, was demonstrated in 14 (33%). One patient experienced sudden cardiac arrest, while another reported a family history significant for sudden cardiac death. Family history of HCM was present in 2 patients.

Multiple CAF, defined as more than one fistula either in the same or different epicardial coronary artery, was noted in 44% (n=19) while 56% (n=24) had a single CAF. The total number of major epicardial coronary arteries with at least 1 CAF was 70. CAF origin and termination locations were graphically reproduced. CAF was most commonly present in the left anterior descending artery (LAD) [n=35] followed by the right coronary artery (RCA) [n=20], the left circumflex (LCx) [n=13], and the left main coronary artery (LMCA) [n=1]. One fistula originated from either the LAD or the LCx, but it was not explicitly described in the case report.

Overall, 51 of the 70 CAF (73%) terminated in the left ventricular cavity while the remaining 19 terminated in pulmonary artery in 14% (n=10), right ventricular cavity in 9% (n=6), both pulmonary artery and the right ventricular cavity in 1% (n=1) or in both left atrial cavity and pulmonary artery in 1% (n=1). The termination point of 1 CAF was not clearly specified.

CAF was treated medically in 26 (60%) patients while 2 (5%) required either transcatheter (vascular occlusion coil) or surgical closure. Follow-up was available for 24 patients (56%). Among these, 17 (71%) were completely asymptomatic, 6 (25%) showed substantial improvement in their symptoms, and 1 (4%) continued to experience symptoms after medical, interventional, or surgical treatment.

Discussion

Coronary artery fistulae, defined as abnormal connection of a coronary artery to other blood vessels or cardiac chambers by bypassing the myocardium. It is often congenital but could be acquired following trauma or cardiac procedures. It is estimated that it affects 0.1% to 0.2% of the population [5]. Advances in cardiovascular imaging has resulted in an increasingly higher rates of detection of CAF in recent years with prevalence rates as high as 0.9% [6]. CAF can be identified by invasive or non-invasive (computed tomographic) coronary angiography, echocardiography with Doppler interrogation, or less commonly with cardiac magnetic resonance imaging [4,7].

Once identified by imaging, the classification of these fistulae is based on their respective vessel of origin and their termination points. The RCA is the most common vessels of origin of CAF in

patients without HCM (50-60%), followed by the LAD (25-42%), and the LCx (18.3%) [7]. In those without HCM, the most common termination point for CAF is the right ventricle (14-40%) followed by the right atrium (19-26%), pulmonary artery (15-20.2%), and left ventricle (2-19%) [8]. Isolated CAF is generally asymptomatic, however, if sizable, they may result in myocardial ischemia, arrhythmias, or congestive heart failure [9].

Our study showed that 77% of HCM patients with CAF had apical variant of the disease. Among all patients with HCM, the apical variant generally accounts for <25% and may vary with ethnicity [10,11]. The higher prevalence of CAF in apical variant of HCM remains unexplained. Whether CAF is truly congenital in this setting or acquired as part of the development of apical hypertrophy remains uncertain. CAF has been reported in other cardiomyopathies that preferentially involve the left ventricular apical segments [12,13]. One may speculate that apical HCM and CAF may share certain common genetic etiologies. Angina and objective evidence of myocardial ischemia is a common feature of AHCM. Apical myocardial perfusion defects are frequently found in such patients [2] and are attributed to microvascular dysfunction that is augmented by prolonged contraction of the apical segments that extend into diastole [12]. Presence of CAF may contribute to myocardial ischemia in AHCM by reducing blood flow to distal adjacent myocardium through coronary steal phenomenon. Eventually, reduced blood flow may lead to development of myocardial fibrosis and apical aneurysms [14].

Calcium channel blockers are frequently used to manage microvascular ischemia in AHCM. It is likely that, at least in part, the effectiveness of these medications is related to their negative inotropic properties. Whether or not newer, more specific medications such as myosin inhibitors would provide additional benefit in such settings is not yet studied. In addition, it is not known whether either medication would be efficacious in the presence of CAF. We believe awareness of potential coexistence of AHCM and CAF may be important in better understanding of myocardial ischemia in this subtype of hypertrophic cardiomyopathy. The most common indications for CAF closure include cardiac chamber enlargement, ventricular dysfunction, myocardial ischemia in the feeder artery territory, arrhythmia related to CAF, vessel rupture, and endocarditis [5]. Closure of these fistulas can be accomplished with either a percutaneous or surgical approach involving the use of covered stents, coils, surgical closure or surgical bypass [5]. It is likely that these general approaches would be followed in patients with CAF and AHCM for the time being.

Conflicts of Interest

The authors declare that we have no conflicts of interest.

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Author Contributions

LM: Conceptualization, Data curation, Formal analysis, Investigation, Visualization, Writing-original draft, Writing-review & editing. MS, KG, NR, BC: Writing-original draft, Writing-review

and editing. JS: Conceptualization, Formal analysis, Supervision, Validation, Writing-original draft, Project administration, Writing-review & editing. All authors read and approved the submitted version.

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