

中文題目：冠狀動脈瘻管併心尖肥厚型心肌病變：病例報告

英文題目：Brief Report: Coronary artery fistula with apical hypertrophic cardiomyopathy

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Introduction

Coronary artery fistula to the left ventricle with apical hypertrophic cardiomyopathy is a rare combination with very low incidence in the general population. When apical hypertrophic cardiomyopathy is combined with coronary artery fistula, it exacerbates myocardial ischemia and could present with symptoms like exertional angina.

Hypertrophic cardiomyopathy can be diagnosed with echocardiogram, however when combined with coronary artery fistula, coronary angiography is required for diagnosis. Medical treatment with beta-blocker, calcium channel blocker or nitrate are used to control symptoms. Beta-blocker and nitrate used for this symptomatic patient with both coronary artery fistula to the left ventricle and apical hypertrophic cardiomyopathy are shown to relief the symptom and maintain stable condition of the patient.

Case Presentation

A 68-year-old Taiwanese male presented to our hospital with exertional chest tightness. He has a history of hepatitis B, and has been smoking for 50 years. He has no family history of cardiovascular disease. His initial heart rate was at 68 beats per minute with a blood pressure of 119/71. Physical examination findings are unremarkable.

Electrocardiogram showed T wave inversion of anterior leads. Chest X-ray showed increased lung markings, interstitial infiltration in bilateral lung fields and borderline cardiomegaly. Echocardiogram showed dilated aortic root, left ventricular apical hypertrophy with normal ejection fraction. SPECT Thallium-201 stress scan showed mixed myocardial ischemia and scarring at the anterior, basal septal and inferior walls. Coronary angiogram revealed left coronary fistula to the left ventricle (Figure 1 and Figure 2) and left ventriculogram showed apical hypertrophic cardiomyopathy (Figure 3

and Figure 4). He was treated with beta blocker carvedilol and nitroglycerin. His symptoms improved after that and continued to follow up at the outpatient department.

Discussion

Coronary artery fistulas(CAF) are rare defects of the circulatory system. CAF are atypical connections between coronary arteries and other structures like other branches of arteries or heart chambers. Although they are mostly asymptomatic, they can still lead to several life-threatening complications. Therefore coronary angiography is necessary for appropriate diagnosis, and treatment is carried out by catheterization closure in cases with such needs. A past study has shown prevalence of CAF to be 0.9% with coronary angiographic findings. Apical hypertrophic cardiomyopathy(AHCM) is an uncommon finding amongst hypertrophic cardiomyopathy patients, first described by Sakamoto in 1976. Studies have found AHCM prevalence to be especially high in Asian countries. According to a Taiwan study, men had a three-fold prevalence of AHCM.

A study published on February 2021 presented a case of a 72-year-old Caucasian female who was brought to medical attention due to syncope, demonstrating upon coronary angiography a distal left circumflex artery (LCx) to LV fistula from the apical hypertrophy but no coronary artery disease. Whereas her echocardiography showed AHCM but with good systolic function. Another study done in Korea in 2015 revealed multiple coronary artery microfistulas associated with hypertrophic cardiomyopathy, with left and right coronary artery to the left ventricle. The patient was symptomatic with chest pain. Similarly, our patient presented with mild exertional chest tightness. There are a few reported cases of coronary fistula with apical hypertrophy, however, there are still rare cases of presentation of coronary fistula combined with AHCM.

Myocardial ischemia is common and of multifactorial origin to hypertrophic cardiomyopathy. It could act alone is pathological mechanisms or along with others synergistically to present with angina in 50% of patients. CAF are more commonly found to terminate in the right ventricle, whereas only 3% terminate in the left ventricle. CAF draining into the left ventricle could hemodynamically compromise myocardium that

leads to myocardial ischemia. This could cause the “steal” phenomenon. Symptoms of angina could occur. As hypertrophic cardiomyopathy and CAF individually could cause angina and myocardial ischemia, the presence of both could exacerbate the severity of symptoms presented clinically.

Treatment would take into consideration the size and anatomic features of the fistula, the patient’s age, presence of other cardiovascular disease or other comorbidities and the presence of symptoms. Patients may undergo surgery or only be treated medically, this decision depending on severity of symptoms and other factors, especially when fistula closure in patients without symptoms remain controversial and that specific guidelines for its management have not been established. A single-center case series found that watchful waiting comprised 80% of treatment approaches, being the main method to treat CAF. Surgical or a transcatheter approach are adopted to close the fistula in a patient presented with symptoms. Patients who are treated medically are usually administered beta-blockers, calcium channel blockers or nitrates. Our patient was treated medically with carvedilol and nitroglycerin. He was advised to follow up at outpatient department annually for echocardiogram.

Nuclear stress test can be used to screen for coronary artery disease and allow the physician to determine whether invasive procedures such as catheterization are needed to examine patients. A past research presented exercise myocardial perfusion test revealing a medium-sized completely reversible apical anterior perfusion defect suggestive of stress-induced ischemia, he was described as having coronary steal phenomenon. Therefore, he underwent coronary angiography and discovered the coronary fistula to left ventricle. EKG of our patient showed diffuse T inversion, therefore treadmill exercise test would not be suitable due to its limitation. SPECT Thallium-201 stress test rather than exercise stress test was conducted for our patient as simulation of the patient in an exercise state to conduct the test for evaluation. Our patient’s SPECT Thallium-201 stress test showed mixed myocardial ischemia and scarring at the anterior, basal septal and inferior walls. Therefore, we carried out coronary angiography which discovered the CAF to the left ventricle.

Conclusion

In conclusion, CAF to the left ventricle combined with AHCM is yet a rare case with very low incidence in the general population. When AHCM is combined with CAF, it exacerbates myocardial ischemia and could present with symptoms like exertional angina. Hypertrophic cardiomyopathy can be diagnosed with echocardiogram, however when combined with CAF, coronary angiography is required as a diagnostic tool. In most cases, medical therapy with beta-blockers are sufficient for treatment and control the symptoms.

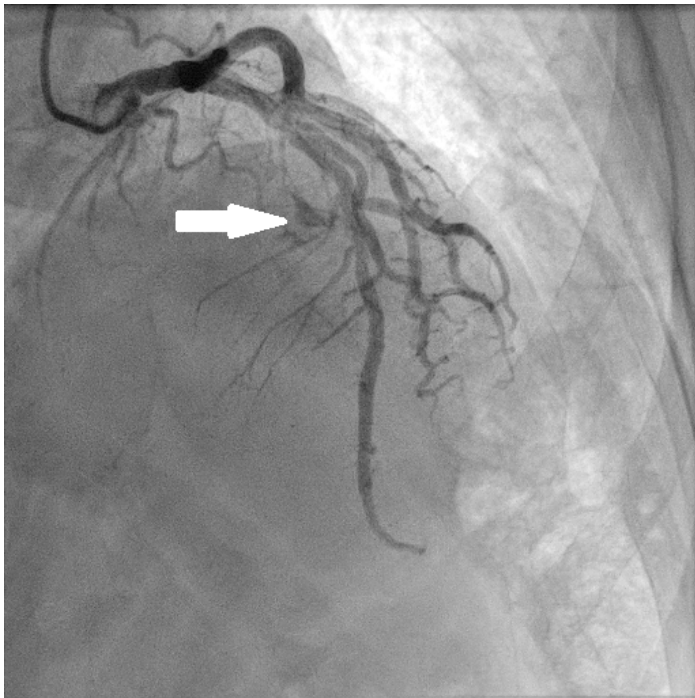


Figure 1. Coronary angiogram revealed left coronary fistula to the left ventricle

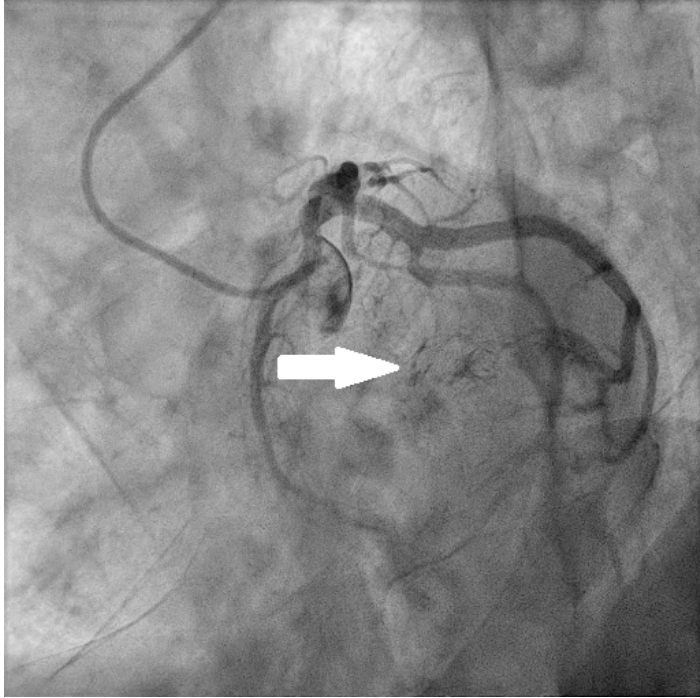


Figure 2. Coronary angiogram revealed left coronary fistula to the left ventricle

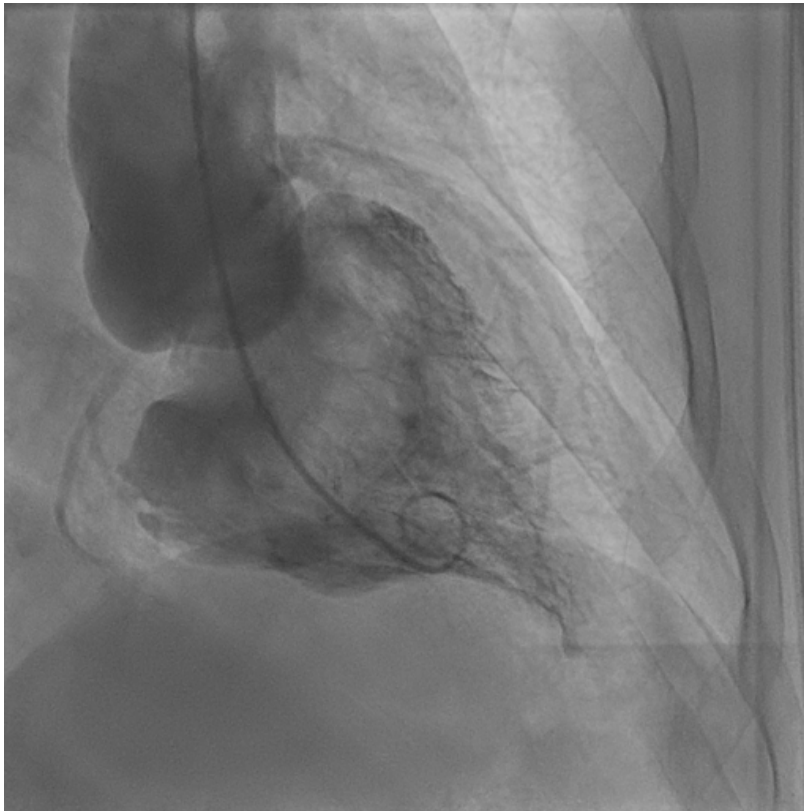


Figure 3. Left ventriculography on diastolic phase

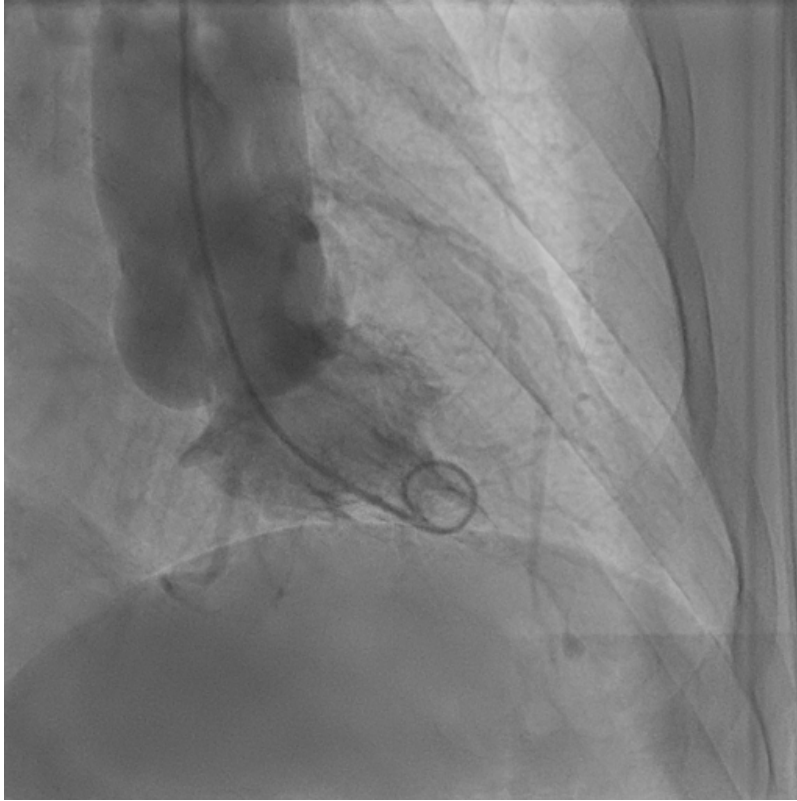


Figure 4. Left ventriculography on systolic phase