CASE REPORT

Coronary artery fistula and mid-cavitary obstructive hypertrophic cardiomyopathy: a rare association

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SUMMARY

We report a case of a 60-year-old man with a history of angina on exertion (New York Heart Association Class II) who was found to have mid-cavitary obstructive hypertrophic cardiomyopathy on two-dimensional echocardiography and whose coronary angiogram showed right coronary artery–left ventricular fistula, with no significant coronary atherosclerosis suggestive of ischaemic heart disease. The patient was started on beta-blocker therapy, and on follow-up his angina improved. The patient had a benign course and did not have any progressive heart failure or acute coronary syndrome on follow-up. This case report highlights a rare association of mid-cavitary obstructive hypertrophic cardiomyopathy with coronary artery fistula.

BACKGROUND

A coronary artery fistula is the most common congenital anomaly affecting the coronary perfusion. It is a communication between a coronary artery and a cardiac chamber, a great artery or the vena cava. Few case reports have reported the association of apical variant of hypertrophic cardiomyopathy with coronary artery–left ventricular fistula. However, coronary fistula to one of the cardiac chambers and a coexisting mid-cavitary obstructive hypertrophic cardiomyopathy have not been reported previously. Both coronary artery fistulae and hypertrophic cardiomyopathy can independently result in angina pectoris, and their association may aggravate the clinical condition.

CASE PRESENTATION

A 60-year-old man, with no risk factors for coronary artery disease and not on any medications, was admitted to our hospital with a 6-month history of angina on exertion (New York Heart Association Class II). There was no previous history or family history of any heart disease. On physical examination, his blood pressure was 140/80 mm Hg, and his pulse was 75 beats/min. His cardiovascular examination showed no abnormalities except for a fourth heart sound and a grade 3/6 systolic murmur at the left lower sternal border increasing on standing and Valsalva manoeuvre.

INVESTIGATIONS

ECG showed ST segment depressions associated with deep asymmetrical inversions in T wave in leads V2–V6 suggestive of left ventricular hypertrophy with strain. Chest radiography revealed

cardiomegaly while laboratory findings including cardiac enzymes were found to be normal. Two-dimensional transthoracic echocardiography showed asymmetrical septal hypertrophy involving the interventricular septum, apex and anterior wall with a severe mid-cavity gradient of 70 mm Hg with no left ventricular outflow tract gradient and no systolic anterior motion of mitral valve leaflet (figure 1). Left ventricular volume measured was within the normal range. No regional wall motion abnormality was seen, and left ventricular ejection fraction was normal. A coronary angiogram done to exclude the diagnosis of ischaemic heart disease revealed no significant stenosis, however a current of dye flow into the left ventricle was visualised after injecting the dye into the right coronary artery (figure 2 and video 1).

TREATMENT

Therapy with beta-blocker metoprolol succinate was initiated for the patient and his symptoms improved subsequently.

OUTCOME AND FOLLOW-UP

On follow-up his angina had reduced and so he was continued on beta-blocker therapy.

DISCUSSION

A coronary artery fistula is deemed as a rare entity, its minuscule incidence being approximately 0.1%–0.2%. The most common aetiology for coronary artery fistulae is congenital due to the persistence of the protrusions of endothelium or sinusoids into the intertrabecular spaces that connect the coronary veins, arteries and cardiac trabeculae components of the original microcirculation of embryonic myocardium). Normally, these embryonic structures degenerate leading to the thebesian vessels formation in the adult heart. A coronary artery fistula is commonly known to terminate inside low-pressure chambers like the right ventricle, right atrium and pulmonary artery. Majority of the patients with coronary artery fistulae have no symptoms. Among those who have symptoms, main presenting complaints include atypical chest pain, syncope, palpitation, angina, dyspnoea, arrhythmia, congestive heart failure and rarely sudden cardiac death because of aneurysmal rupture.¹

One study reported 2% prevalence in coronary artery fistula in patients undergoing a diagnostic angiogram. This study retrospectively studied 20 patients who had microcoronary artery fistulae and

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Figure 1 Two-dimensional echo showing asymmetrical septal hypertrophy involving the interventricular septum, apex and anterior wall with left ventricular mid-cavity gradient.

found that 90% of them had a concentric left ventricular hypertrophy. They concluded that persistent volume overload due to the fistulous shunt led to a compensatory hypertrophy of the myocardium.²

Nevertheless, in our case, the left ventricular volume measured was normal, and a localised asymmetrical hypertrophy was noted involving the interventricular septum, apex and anterior wall. Therefore, the probable reason behind the association between hypertrophic cardiomyopathy and coronary artery fistula in this case may be congenital due to disarray of myocardial cells due to cardiomyopathy itself. MRI or cardiac biopsy can aid in finding out the mechanism of this relationship between these two distinct clinical entities of cardiomyopathy and coronary artery fistula. However, it was not done, as it does not influence our further management in any way.

Few case reports in literature indeed have discussed the rare association of coronary artery–left ventricle fistulae with the apical type of hypertrophic cardiomyopathy.^{3–6} However, the association of coronary artery fistula with mid-cavitary obstructive type of hypertrophic cardiomyopathy has not been reported thus far.

Figure 2 Arrow shows stream of dye into the left ventricle via a tiny plexus of intramural vessels, from the posterior descending branch of the right coronary artery during systole and diastole in LAO cranial angiographic projection. LAO, left anterior oblique.

Video 1 Coronary angiogram of the right coronary artery in anterior-posterior cranial projection showing dilated right coronary artery and a stream of dye ejecting from its posterior descending branch into the left ventricle during systole and diastole.

Compared with a more common termination into low-pressure chambers like the right ventricle, right atrium and pulmonary artery, the terminal drainage of a coronary artery fistula into a high-pressure chamber like the left ventricle is comparatively much less common, its incidence ranging from 3% to 17%.⁷

The coronary artery fistulae can be classified anatomically based on the Sakakibara angiographic classification, depending on the site of origin of the fistulae.⁸ Type A fistulae originate from the proximal one-third of the parent coronary whereas type B fistulae are known to arise after the proximal one-third or signify the final connections of the parent coronary artery. The coronary artery segment proximal to a type B fistula is known to be dilated. The right coronary artery to left ventricle fistula in this patient was hence a type B Sakakibara fistula as it originated beyond the proximal one-third of the parent right coronary artery, and the right coronary artery diameter proximal to the fistula was increased. This anatomical classification is important during the management of fistulae. One such example is seen in type B fistulae which are prone to cause coronary slow flow and subsequent risk of coronary thrombosis especially after transcatheter closure by vascular coil. This is because before closure of the fistula, a high volume of blood flows through the fistula rapidly that comes after travelling through the entire length of the native coronary, but after closure of the fistula, there is a sudden fall in the volume flow rate leading to stasis in the parent coronary.

Once this slow flow phenomenon is diagnosed, anticoagulation must be started immediately, beginning with low-molecular weight or unfractionated heparin and then bridging to warfarin for achieving international normalised ratio goal of 2.0–3.0. If there is no bleeding, anticoagulation should be advised for 6 months. This allows adequate time for the remodelling of the parent coronary and for its flow to become normal, following which anticoagulation can be terminated.

It is not clear whether the left ventricular hypertrophy is a compensatory response to the persistent volume overload caused by this shunt operating between the coronary artery and the left ventricle or if it is the myocardial hypertrophy in hypertrophic cardiomyopathy causing disarray of myocardial cells, triggering the formation of a coronary artery fistula.⁹

Both hypertrophic cardiomyopathy and coronary artery fistula can independently contribute towards a patient developing angina pectoris. When they both coexist, their association can worsen the symptoms.

Pressure gradient in diastole caused by blood run-off from the vasculature of the coronary artery to a low pressure left ventricular cavity leads to shunting of blood away from the coronary artery via the low resistance fistulous tract. This mechanism of 'Coronary Steal' leads to attenuation of blood flow distal to the fistula. Due to the reduced blood supply, the myocardium that is distal to the origin of the coronary artery fistula is always at an increased risk of ischaemia whenever the demand for oxygen rises, for example, during exercise, due to oxygen demand–supply mismatch.¹⁰

Similarly, in patients with coronary artery fistula with hypertrophy, an increased demand for oxygen occurs due to the hypertrophied myocardium. This together with reduced coronary blood flow due to shunting causes oxygen demand–supply mismatch in the myocardium leading to ischaemia and hence, chest pain.

Because this association is rare, full natural course of coronary artery fistula to the left ventricle in the presence of hypertrophic cardiomyopathy has not been adequately reported in literature.

Ischaemia in this setting commonly has a benign progression with no case of myocardial infarction been reported thus far.

Delineating the fistula's anatomical origin, course and relation to adjacent structures is sometimes difficult, especially in a large and tortuous fistula and a fistula overlapping its own site of beginning and end. In such cases, non-invasive techniques such as CT or MRI imaging with three-dimensional coronary reconstruction are useful. However, due to cost constraints and low socioeconomic status of the patient, it was not feasible to do these imaging tests in this patient.¹¹

Nuclear stress test can be helpful in the assessment of myocardial ischaemia caused by a coronary artery fistula. It can help decide the need for invasive approach to the management. If the patient is asymptomatic and has a reduced territory (<10% left ventricular surface area) showing myocardial ischaemia, drug therapy is the first line of management.¹² However, due to cost constraints and low socioeconomic status, it was not feasible to do nuclear stress test in this patient.

There has been no guideline fixed for the management of patients with coronary artery fistulae in the presence of hypertrophic cardiomyopathy. According to American Heart Association guideline, closure of the fistula by percutaneous or surgical means is a Class I indication for closing a large fistula irrespective of symptoms. Closure for small-sized to moderate-sized fistula is however indicated when there is evidence of myocardial ischaemia, ventricular dysfunction or enlargement, arrhythmia or endarteritis.¹³

Percutaneous transcatheter closure is less invasive and avoids complications of surgery, so it is a preferred method. However, if anatomy is unfavourable to transcatheter closure, especially if the vessel is tortuous or in cases of a large fistula, surgical closure is a better choice. Asymptomatic patients having small fistulae with non-significant shunt should only undergo close follow-up. However, elective closure of an asymptomatic fistula is indicated in children who are older than 3–5 years, as they are at a higher risk of developing complications due to the fistula. Closure is also indicated for a fistula originating from the proximal portion of coronary arteries because of their higher susceptibility to aneurysm formation and rupture.¹⁴

However, since angina was the only evidence for ischaemia and there was a coexisting hypertrophic cardiomyopathy that can independently contribute towards a patient developing angina pectoris, angina could not be solely attributed to coronary artery fistula alone. So surgical closure in this patient was not considered as the first option.

Medical management with drugs like beta-blockers (metoprolol) and nicorandil addresses the problem of demand–supply mismatch that is responsible for myocardial ischaemia in this patient. Metoprolol reduces the heart rate, thus decreases the oxygen consumption by the heart, and so reduces the demand problem for a hypertrophied heart, as in this patient. Nicorandil is a potassium channel opener and a coronary vasodilator that increases the coronary blood flow and thus helps to increase the supply of coronary blood flow which is reduced by the coronary steal caused by arterio-venous fistula.

In this case, metoprolol was given to the patient, and his symptoms improved suggesting that treatment with beta-blockers seems to be the preferred choice of management in cases of coronary artery fistula coexisting with hypertrophic cardiomyopathy.

Learning points

- ▶ This case report depicts a rare association of coronary artery to left ventricular fistula and mid-cavitary obstructive hypertrophic cardiomyopathy.
- ▶ Both hypertrophic cardiomyopathy and coronary artery fistula can individually contribute towards a patient developing myocardial ischaemia and angina pectoris. However, when they both coexist, their association can worsen the symptoms.
- ▶ Ischaemia in the setting of coronary artery to left ventricular fistula coexisting with hypertrophic cardiomyopathy normally has a benign progression with myocardial infarction unlikely to occur.
- ▶ Symptomatic treatment with beta-blockers seems to be the preferred choice of management in cases of coronary artery fistula coexisting with hypertrophic cardiomyopathy.

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