A 72-year-old man presented to the outpatient clinic with symptoms of chest pain and dyspnoea on exertion (NYHA III) for past two months. Physical examination was unremarkable with normal first and second heart sounds. Unfortunately, the valsalva maneuver was not performed as no murmur was appreciated on clinical examination. Echocardiogram showed small sized LV cavity with asymmetrical septal hypertrophy in the mid segments measuring 22 mm, normal LV systolic function, producing mid cavity gradients of 50 mm of Hg, suggestive of hypertrophic cardiomyopathy. Cardiac magnetic resonance imaging (CMR) was performed that confirmed hypertrophic cardiomyopathy (HCM). A contrast enhanced CT angiogram (CTA) was done that showed coronary artery fistula arising as multiple small vessels from the proximal left anterior descending artery (LAD) and draining into the pulmonary artery (PA). Initially, the patient was managed medically considering HCM as the cause of his symptoms but subsequently, with the findings of coronary artery fistula by CTA, the surgical correction of the fistula was advised. However, the patient opted for conservative treatment. At one year follow-up, he was well on optimal medical treatment with improvement in functional class from NYHA III to II. This describes a rare case of coronary artery to pulmonary artery fistula in a patient with hypertrophic cardiomyopathy that presented late in life.

Keywords: Case report, Coronary artery-pulmonary artery fistula, Hypertrophic cardiomyopathy, Cardiac MRI (CMR), Chest pain.

Introduction

Anomalous coronary arteries are an uncommon but important clinical finding. This is a rare case of a patient who presented at an old age with a left proximal LAD to pulmonary artery fistula along with hypertrophic cardiomyopathy. We review briefly the etiology of coronary artery fistulae, clinical presentation, diagnostic tests used to establish diagnosis and the current treatment modalities. To the best of our knowledge, this kind of rare case has never been reported from Pakistan.

Case Report

A 72 year old man presented to the clinic in August 2014; at Aga Khan University Hospital, with complaints of chest pain and dyspnoea for two months. The dyspnoea was progressive, occurred on climbing one flight of stairs (NYHA III) and was associated with mild chest discomfort. His physical examination was unremarkable with normal first and second heart sounds. Unfortunately, the Valsalva maneuver was not performed as no murmur was appreciated on clinical examination. Biochemical and haematological investigations were normal while chest X-ray was also unremarkable. ECG showed features of left ventricular hypertrophy. Transthoracic echocardiogram showed small sized LV cavity with normal left ventricular systolic function. There was asymmetrical septal hypertrophy in the mid segments measuring 22 mm, the anterior wall measured 16mm, whereas the inferior wall measured 14mm; producing mid cavity gradients of 50 mm of Hg, suggestive of hypertrophic cardiomyopathy. The echocardiogram was performed without doing Valsalva maneuver. CMR was done (Figure 1 A & B) which showed left ventricular hypertrophy mainly involving the mid segments; with thinned out and akinetic small apical portion, along with obliteration of LV cavity in the mid region during systole. The LV volumes were normal with normal systolic function (EF 64%). Delayed enhancement of gadolinium was present only in the small apical portion. In addition, there were small focal areas of hyperenhancement of internal papillary muscles. This case of coronary artery to pulmonary artery fistula was confirmed by CTA. At one year follow-up, he was well on optimal medical treatment with improvement in functional class from NYHA III to II.
enhancement in the left ventricle at the right ventricular insertion site. No stress testing was performed in our case. These findings were consistent with hypertrophic cardiomyopathy (mid cavity type).

Due to history of chest pain on exertion, coronary CT angiogram (CTA) was performed to rule out coronary artery disease (CAD). CTA (Figure 2 A & B) showed bunch of small tortuous vessels arising from the proximal LAD and opening into the main pulmonary artery through a relatively bigger channel. The left ventricle appeared normal in size with significant left ventricular hypertrophy mainly involving the mid segments. There were calcified plaques in the proximal and mid LAD causing mild narrowing. Circumflex and ramus were free of disease while RCA had mild non-obstructive calcified plaques.

The patient was managed medically with aspirin, betablockers and statin considering hypertrophic cardiomyopathy as the cause of his symptoms. But subsequently, with the finding of coronary artery fistula on CTA, surgical resection was considered. However, the patient opted for conservative treatment. The patient was also advised to maintain good fluid status, avoiding dehydration. He was also advised to avoid certain medications which included nitrates, diuretics and ACE-inhibitors. Since the patient tolerated beta blockers and was doing fine with good symptomatic improvement in NYHA class, calcium channel blockers and disopyramide were not considered. At one year follow up visit, significant improvement in his symptoms was observed with reduction in NYHA class from III to II.

**Discussion**

A coronary artery fistula (CAF) is an abnormal vessel communication between the coronary artery and cardiac chamber, great vessels or other vascular structures, with an estimated incidence of 0.002% in the general population. The condition is found with equal incidence in males and females and is frequent below 10 years of age. Approximately 20% of patients with coronary artery fistulae have other cardiac anomalies, most frequently aortic and pulmonary atresia and patent ductus arteriosus. However, the co-existence of left anterior descending artery to pulmonary artery fistula and hypertrophic cardiomyopathy (HCM) has been rarely reported in the literature.

In more than 50% of the cases, the fistulae arise from the right coronary artery, and in 14-17% they terminate in the pulmonary artery. In our patient, the fistula arose from LAD and terminated in the pulmonary artery. The pathogenesis of CAF is congenital in most cases. Embryologically, they seem to represent persistent junctions of primordial epicardial vessels with intramyocardial sinusoidal circulation.

Approximately half of these patients are asymptomatic and undergo cardiac catheterization because of a continuous precordial murmur. The clinical presentations in symptomatic patients can vary from angina, atypical chest pain, syncope, dyspnoea, palpitation, congestive heart failure, arrhythmia, and can even lead to sudden cardiac death. Our patient had symptoms of chest pain and dyspnoea which could be due to hypertrophic cardiomyopathy or coronary fistula.

The mechanisms of myocardial ischaemia in HCM with CAF include increased oxygen demand by the hypertrophied ventricular myocardium, impaired coronary circulation, demand supply mismatch and increased diastolic filling pressures. CAF can result in coronary steal and left-to-right shunt which may further reduce the ventricular perfusion and increase diastolic volume overload. The factors influencing the clinical presentation and prognosis of the coronary to pulmonary artery fistula (CPAF) are the size of the communication, the amount of blood drained through it, the resistance of the recipient chamber, and development of myocardial ischaemia or infarction.

The clinical diagnosis of coronary artery-pulmonary artery fistulae is difficult because clinical presentation, laboratory and ECG manifestations are usually nonspecific. Coronary CT angiogram is a good tool to diagnose coronary artery fistula as in our case.

A study done by Salah in 2011 showed that most patients (80%) had unilateral fistulae, 18% presented
with bilateral fistulae and 2% with multilateral fistulae. CAFs originated from the left coronary artery in 69% and from the right coronary artery (RCA) in 31% of the subjects. Termination into the pulmonary artery (PA) was reported in unilateral (44%), bilateral (73%) and multilateral (75%) fistulae.7

There is a lack of robust evidence with regards to the management of these patients, especially if asymptomatic. Treatments range from none, medical therapy to coronary intervention or surgical re-implantation in high-risk cases. This has become an area of interest recently due to the increased use of cardiovascular CT, which has led to a larger number of these anomalies being identified as incidental findings.8 The coexistence of HCM and CAF, both of which are well known to independently increase the likelihood of sudden cardiac death, has been reported rarely and leads to complexities in management.

Conservative management with continued follow-up of patients with coronary artery to pulmonary artery fistula appears to be appropriate. Surgical treatment is usually considered only in severe forms with refractory medical treatment and goal is to obliterate the fistula while preserving normal coronary blood flow. Indications for surgery include the presence of a large fistula, increasing left-to-right shunt, left ventricular volume overload, myocardial ischaemia, left ventricular dysfunction, congestive cardiac failure and for prevention of endocarditis.9

The prognosis in asymptomatic patients is good and a conservative clinical follow up is recommended because of the low incidence of an adverse outcome.10

Informed Consent: Written informed consent was obtained from the patient for their anonymized information to be published in this article.

Disclaimer: This case report has never been published or presented in a conference previously.

Conflict of Interest: None to declare.

Funding Disclosure: None to declare.

References