CASE REPORT

Aneurysmal coronary cameral fistula

Gohar Jamil,¹ Asad Khan,² Azhar Malik,² Anwer Qureshi¹¹Department of Medicine, Division of Cardiology, Tawam Hospital, Al Ain, United Arab Emirates²Department of Medicine, Division of Medicine, Tawam Hospital, Al Ain, United Arab Emirates

Correspondence to

Dr Gohar Jamil, goharjamil@gmail.com

SUMMARY

A 26-year-old asymptomatic man, being medically managed for ventricular septal defect since childhood, presented to the outpatient clinic for a second opinion. Clinically, he was well built with normal vital signs. Cardiac auscultation was significant for a diastolic murmur over the praecordium. An ECG showed non-specific ST changes, and a subsequent transthoracic echocardiography performed revealed diastolic flow from the left ventricular (LV) anteroseptal wall into the LV cavity. A diagnosis of coronary–cameral fistula was confirmed by a multidetector CT which showed a 2.5×2 cm aneurysmal left anterior descending artery fistula to the LV. In addition to starting aspirin, transcatheter closure with occlusion device was considered knowing the potential risk of thrombus formation in the aneurysm and subsequent systemic embolisation. The patient however refused any percutaneous or surgical intervention. He remains asymptomatic 1 year after returning to his home country.

BACKGROUND

Coronary artery anomalies can be classified based on their origin, course or termination. Anatomically, coronary arteries arborise into capillaries and penetrate the myocardium. Coronary artery fistulas (CAF) are rare forms of coronary artery anomalies in which either a direct communication exists between the coronary artery and cardiac chamber (coronary–cameral fistula) or a segment of pulmonary and systemic circulation (coronary arteriovenous fistula). Embryologically, CAF are thought to be due to persistence of primitive intratrabecular spaces.¹ More than 90% of CAF drain into the venous circulation,² whereas 3% drain into the left ventricle (LV).³ The association of CAF with huge coronary aneurysms draining into the LV is extremely rare, with no specific management recommendations. Addition of each such a case to literature will enlighten the physician community about different management strategies.

This case also illustrates the importance of a good bedside examination in this high-tech era. As, had the diastolic murmur been properly auscultated the fistula would not have been confused with muscular ventricular septal defect (VSD) on the subsequent transthoracic echocardiogram.

CASE PRESENTATION

A 26-year-old active, asymptomatic man diagnosed and medically managed as VSD since childhood was visiting UAE. Insisted by his family, he presented to our hospital for a second opinion. There was no significant medical or surgical history. In addition, he was not on any regular medication and

did not consume tobacco, alcohol or recreational drugs. Family history was non-contributory.

Examination revealed a well-built man with normal facies. His blood pressure was 116/72 mm Hg and heart rate 76 bpm. The patient's body temperature and oxygen saturations were normal. The bounding peripheral pulses were normal, and no cyanosis or clubbing was noted. There were no carotid bruits, and the jugular venous pressure was not elevated. Chest was clear to auscultation with no added sounds. A cardiac examination revealed a regular rate and rhythm with a normal first and second heart sounds. In addition, an early diastolic murmur was audible over the whole praecordium, which was more prominent in the aortic area. There was no specific change in the quality of murmur with different cardiac manoeuvres. The abdomen was soft without any hepatosplenomegaly and normal bowel sounds. The remaining physical examinations were non-contributory.

INVESTIGATIONS

Transthoracic echocardiogram showed normal left and right ventricular sizes and functioning. Both atria were normal with no significant valvular pathology. Parasternal views showed diastolic flow from a circumscribed cavity in the anteroseptal wall to the LV, suspicious for a coronary artery to LV fistula (figure 1A,B).

Cardiac CT confirmed the presence of a single large 2.5×2 cm aneurysmal fistula from the left anterior descending (LAD) artery to mid-LV cavity (figure 2A,B). The coronary arteries were free of any atherosclerotic disease.

DIFFERENTIAL DIAGNOSIS

The different causes of a diastolic murmur should be considered in the differential diagnosis of such patients. These could be mainly related to aortic and pulmonary valvular insufficiency in a vast majority of cases. Similarly, stenosis or increased flow across the atrioventricular valves can cause diastolic murmurs, and rarely rupture of the sinus of Valsalva to the right atrial fistula can mimic findings similar to CAF.

TREATMENT

The pathophysiological consequences of an LAD to LV fistula are similar to volume overload caused by aortic valve insufficiency. Fistulas may progressively enlarge overtime, and have been associated with myocardial ischaemia, congestive heart failure, bacterial endocarditis, premature atherosclerosis, arrhythmias and sudden death.

In case of our patient aspirin was started for its antiplatelet effect in preventing microemboli.

To cite: Jamil G, Khan A, Malik A, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008649

Figure 1 (A and B) Parasternal long-axis view showing abnormal diastolic colour flow signal (arrow) from the cameral coronary fistula to left ventricle (LV, left ventricle; LA, left atrium; CCF, cameral coronary fistula).

Prophylactic antibiotic prior to surgical procedures was recommended. A percutaneous closure of the fistula was considered, but not performed because of various factors, including patient's reluctance, the giant size of the aneurysm and concerns about thrombus formation in the aneurysm and subsequent systemic embolisation. The patient declined any surgical intervention.

OUTCOME AND FOLLOW-UP

The patient remains asymptomatic and event free 1 year since the correct diagnosis.

DISCUSSION

Coronary cameral fistulas (CCF) are rare and fascinating anomalies found in approximately 0.08–0.3% of unselected patients

undergoing coronary angiography.^{4 5} Ninety per cent of these communicate with right-sided chambers. Most CCFs are small and consequently the patients are asymptomatic. Symptoms usually depend upon the size and location of the fistula, as well as the resistance of the communicating chamber. Distal fistulas may cause massive dilation in the proximal native coronary artery,^{6 7} resulting in stagnation and thrombosis of the proximal vessel. Spontaneous closure has been reported in children but is uncommon in adults.^{8 9} CCF from LAD to LV associated with an aneurysmal sac is limited to a few case reports in literature.^{10 11} Depending upon the termination of the CAF, a systolic, diastolic or a continuous murmur is audible. A CCF communicating with the LV would produce a diastolic murmur. The importance of an accurate clinical exam in these cases is vital and cannot be

Figure 2 (A and B) Three-dimensional CT shows the left ventricle side (arrow) and left anterior descending artery side (curved arrow) of the coronary cameral fistula (arrow head). (LV, left ventricle; LAD, left anterior descending artery).

overemphasised. Clinical assessment in conjunction with multimodality imaging, that is, echocardiography and multidetector CT¹² can reliably localise the fistula, outline its anatomical course and confirm the diagnosis. In addition, modern imaging techniques can eliminate the need for invasive angiogram prior to any planned surgical or percutaneous procedure if needed.

Management decisions for CCF at times can be challenging, owing to the rarity of this condition. Conservative management has been recommended in patients with small, asymptomatic and incidentally discovered fistulas.¹³ Sherwood *et al*⁹ showed patients with silent fistulas remained asymptomatic for a mean follow-up of 2 years. However, data regarding the long-term risk of intervening in asymptomatic patients and the long-term effects of interventions are presently lacking.¹⁴ Patients who are symptomatic are either treated surgically, or by transcatheter closure (TCC) device.^{11–15} TCC, a relatively recent approach, associated with a shorter hospitalisation and quicker recovery may have an apparent advantage over open heart surgery. Results from transcatheter embolisation and surgical ligation show that both approaches have comparable early effectiveness, morbidity and mortality.¹⁴ TCC is not recommended for distally located fistulas, those with significant aneurysmal dilation or multiple openings. Gowda *et al*¹⁶ have recommended conservative management for small CAF and percutaneous or surgical closure of moderate to large proximal CCF, while the approach to distal

CCF still remains controversial. The duration of antiplatelet therapy depends on the size and location of the CCF.¹⁶ Small CCF does not require antiplatelet therapy. Moderate-to-large-sized CCF which are proximally located, require a year of antiplatelet therapy, while those located more distally, may need lifelong treatment. An additional 6–12 months of warfarin therapy is advised for symptomatic patients with large distal CCF.

Longitudinal studies are warranted to clarify these issues. However routine follow-up with non-invasive multimodality imaging should be the standard of care in all the cases. Our patient's refusal of surgery and major concerns of thromboembolism limited his options to medical management alone, and he opted for assiduous follow-up and long-term aspirin therapy in his native country.

Learning points

- ▶ History-taking and clinical examination still remain the cornerstone in the diagnosis and subsequent management of patients.
- ▶ Coronary–cameral fistula from left anterior descending into LV associated with a giant aneurysm is an extremely rare condition. Comparative outcomes of conservative medical, percutaneous or surgical interventions are unknown. The management of each reported case adds to our pool of knowledge.
- ▶ Multimodality imaging obviates the need for invasive angiogram in diagnosing and follow-up of these patients.

Contributors GJ was involved in performing the cardiac angiography and writing up the case report. AQ was also involved in patient care, performing the echocardiogram and approved the final draft. AK and AM were involved in researching the topic, writing and editing the manuscript and Pubmed research.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Barbosa MM, Katina T, Oliveria HG, *et al*. Doppler echocardiographic features of coronary artery fistula: a report of 8 cases. *J Am Soc Echocardiogr* 1999;12:149–54.
- Fujimoto N, Onishi K, Tanabe M, *et al*. Two cases of giant aneurysm in the coronary-pulmonary artery fistula associated with atherosclerotic changes. *Int J Cardiol* 2004;97:577–8.
- Lin FC, Chang HJ, Wen MS, *et al*. Multiplane transesophageal echocardiography in the diagnosis of congenital coronary artery fistula. *Am Heart J* 1995;130:1236–42.
- Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Catheter Cardiovasc Diagn* 1990;21:28–40.
- Vavuranakis M, Bush CA, Boudoulas H. Coronary artery fistulas in adults: incidence, angiographic characteristics, natural history. *Catheter Cardiovasc Diagn* 1995;35:116–20.
- Gowda ST, Latson LA, Kutty S, *et al*. Intermediate to long term outcome following congenital coronary artery fistulae closure with focus on thrombus formation. *Am J Cardiol* 2011;107:302–8.
- Valente AM, Lock JE, Gauvreau K, *et al*. Predictors of long term adverse outcomes in patients with congenital coronary artery fistulae. *Circ Cardiovasc Interv* 2010;3:134–9.
- Nakamura M, Matsuoka H, Kawakami H, *et al*. Giant congenital coronary artery fistula to left brachial vein clearly detected by multi-detector computed tomography. *Circ J* 2006;70:796–9.
- Sherwood MC, Rockenmacher S, Colan SD, *et al*. Prognostic significance of clinically silent coronary artery fistulas. *Am J Cardiol* 1999;83:407–11.
- Arat N, Gurel OM, Biyikoglu FS, *et al*. Coronary artery to left ventricular fistula demonstrated by transthoracic echocardiography. *Eur J Echocardiogr* 2008;9:121–2.
- Jo S, Choi Y, Oh D, *et al*. Coronary artery fistula with a huge aneurysm treated by transcatheter coil embolization. *Circ* 2006;114:e631–4.

- 12 Rittenhouse EA, Doty DB, Ehrenhaft JL. Congenital coronary artery-chamber fistula: review of operative management. *Ann Thorac Surg* 1975;20:468–85.
- 13 Latson LA. Coronary artery fistulas: how to manage them. *Catheter Cardiovasc Interv* 2007;70:110–16.
- 14 Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. *Int J Cardiol* 2006;107:7–10.
- 15 Jama A, Barsoum M, Bjarnason H, *et al*. Percutaneous closure of congenital coronary artery fistulae. Results and angiographic follow-up. *JACC Cardiovasc Interv* 2011;4:814–21.
- 16 Gowda ST, Forbes TJ, Singh H, *et al*. Remodelling and thrombosis following closure of coronary artery fistula with review of management. *Catheter Cardiovasc Interv*. Published Online First: 14 Feb 2013. doi:10.1002/ccd.24699

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow