CASE REPORT

The dilemma of evaluating a continuous murmur in a patient of aneurysm of sinus of Valsalva and coronary cameral fistula presenting with supraventricular tachycardia

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SUMMARY

A 39-year-old hypothyroid woman on thyroxine replacement therapy presented with an unresolving episode of palpitations (narrow-complex tachycardia). Clinical examination, after reversion to normal sinus rhythm revealed a precordial continuous murmur. Initial transthoracic echocardiogram showed an unruptured aneurysm of left sinus of Valsalva (LSOV), however, because a continuous murmur could not be explained by this condition, a repeat colour Doppler study was made, revealing a communicating tract from the left main coronary artery (LMCA) and terminating in the right atrium (RA). A transesophageal echocardiogram revealed an aneurysmal LMCA and LSOV, with similar colour Doppler findings. A further CT scan and coronary angiogram confirmed a coronary cameral fistula opening into RA. In conclusion, the relevance of a diligent clinical examination and imaging after conversion to normal sinus rhythm in picking up such anomalies cannot be over-emphasised, as previous routine echocardiograms on the same patient had been reported as normal.

BACKGROUND

Coronary cameral fistulas are rare anomalies seen in 0.08–0.3% of patients undergoing diagnostic coronary angiograms.^{1–2} The natural course of untreated coronary artery aneurysms with coronary artery fistulas is obscure³ and patients commonly present with arrhythmias including supraventricular tachycardia (SVT).⁴ A coronary cameral fistula is associated with a low intensity continuous murmur clinically, and sometimes masquerades as an unruptured aneurysmal left sinus of Valsalva (ALSOV) on echocardiography. Thus clues gathered from both a thorough cardiovascular examination (especially after control of heart rate) in patients presenting with a tachyarrhythmia, as well as from imaging studies, are essential in arriving at a diagnosis.

CASE PRESENTATION

A 39-year-old woman presented with an unresolving sensation of rapid and regular heart beating for 2 h and exertional shortness of breath for 3 months. She had a history of three similar episodes of palpitations over the past 2 years (6 months, 1 year and 2 years ago) and of Grave's disease, medically managed for 1 year; with recent conversion to a hypothyroid state, treated with thyroxine replacement for the past 3 months. Her

previous episodes of palpitation were treated symptomatically at a remote suburban facility where three echocardiograms taken over the past 2 years had been reported as normal.

At presentation, the patient's examination revealed a heart rate of 170/min, blood pressure of 100/60 mm Hg, pallor, bilateral exophthalmos and a large nodular goitre. Her ECG revealed narrow complex SVT with a rate of 170/min, which reverted to sinus rhythm with an intravenous bolus of 6 mg adenosine.

Re-examination of her cardiovascular system after reversion to normal sinus rhythm revealed a non-radiating, grade 2/6, continuous murmur localised to her right second intercostal space with no respiratory variation and a normally split second heart sound. There were no additional heart sounds or signs of heart failure. In the coronary care unit (CCU) she was started on diltiazem orally (60 mg three times a day).

INVESTIGATIONS

Her blood work revealed haemoglobin of 9 g/dL and euthyroid state with thyroid-stimulating hormone (TSH) of 1.9 mIU/L (0.5–4 mIU/L). Three previous echoes (performed at suburban facilities) were reported as normal. An initial transthoracic echocardiogram (performed at presentation) was repeated (once rate was controlled) to show an ALSOV but because of the presence of a clinically inexplicable continuous murmur, a repeat colour Doppler was performed. This showed a fistulous communication from an aneurysmal left main coronary artery (LMCA) coursing the interatrial septum, ending in the right atrium (RA) and an unruptured ALSOV (figure 1).

A transesophageal echocardiogram (TEE) confirmed a coronary cameral fistula (between an aneurysmal LMCA and RA) and an aneurysmal LMCA as well as ALSOV (figure 2). A diagnostic cardiac catheterisation and angiogram were performed to define the anatomy and visualise its openings (figure 3). A full saturation run was performed, which revealed a step-up in oxygen saturation in the RA (table 1) with a left to right shunt of Qp/Qs=1.75.

A preoperative CT coronary angiogram showed a large coronary cameral fistula (20 mm wide) with an 11 cm long tract between the left main artery and the RA, starting from the left main at the

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Figure 1 Transthoracic echocardiogram in short-axis view, at the level of the aortic valve, showing an aneurysmal left main coronary artery (LMCA) and left sinus of Valsalva aneurysm (LSOV), right atrium (RA), right ventricle (RV), aorta (Ao), pulmonary artery (PA), left atrium (LA) and interatrial septum (IAS).

coronary sinus, traversing in between the aortic root and left atrium, and terminating at the RA (figure 4).

TREATMENT

A successful surgical patch closure was performed under general anaesthesia, after correcting the patient's anaemic status. At surgery, the fistula was incidentally found to have two separate openings into the RA, both of which could be closed with a patch. As the two openings were not detected on any imaging modality performed by us, it was well that we opted for surgical closure, which served the patient's requirement better than a closure device would have.

OUTCOME AND FOLLOW-UP

The patient's postoperative course was uneventful and her continuous murmur disappeared postsurgery. She was discharged on dual antiplatelet therapy. At 4-week follow-up, a repeat TEE showed obliteration of the fistulous tract, without any post-operative leaks. The patient continues to be asymptomatic and is doing well at 6 months of follow-up.

DISCUSSION

Coronary artery fistulas are communications between the coronary arteries (right coronary (55%), left coronary (35%), both

Figure 2 Transesophageal echocardiogram showing a modified five-chamber view revealing CCF opening into the RA. Right atrium (RA), right ventricle (RV), aorta (Ao), left atrium (LA), coronary cameral fistula (CCF) and left ventricle (LV).

Figure 3 Coronary angiogram showing a dilated aneurysm of left sinus of Valsalva (LSOV) and a coronary cameral fistula (CCF) opening into the right atrium (RA).

coronaries (5%)) and a cardiac chamber (right ventricle (40%), RA (26%), left ventricle (1.2%)) or a major vessel such as pulmonary artery (17%) and less frequently superior venae cavae, or coronary sinus.⁵ Coronary cameral fistulas are rare and seen in 0.08–0.3% of patients undergoing diagnostic coronary angiograms.^{1 2}

Their haemodynamic consequences depend on their size and the communicating chamber.⁶ Up to 50% of patients remain asymptomatic and have excellent long-term prognoses,⁶ however, the remainder may present with dyspnoea on exertion, fatigue, congestive heart failure, pulmonary hypertension, bacterial endocarditis, arrhythmias and myocardial ischaemia secondary to coronary steal phenomenon.⁴

Our patient presented with arrhythmia (paroxysmal SVT, PSVT),⁴ a common presentation of coronary arteriovenous fistulas (CAVF), which was unlikely secondary to hyperthyroidism, as her TSH levels indicated a euthyroid state. Her symptoms of breathlessness on exertion were in part due to her severe anaemic state rather than the cardiovascular aetiology alone. However, the exertional dyspnoea continued even after correction of the anaemia by blood transfusion.

The presence of continuous (systodiastolic) murmurs are not always audible in association with coronary cameral fistulas^{7 8} and in their presence it is essential to rule out the rare possibility of a ruptured sinus of Valsalva, a likely differential diagnosis in our case in view of an associated ALSOV. Sinus of Valsalva aneurysms (SVAs) are commonly found arising from the right sinus (94% of the cases) and rarely in the other two sinuses (non-coronary in 5%, and left in 1%).⁹ Further, unruptured SVAs are usually clinically silent.¹⁰ Occasionally, they can precipitate arrhythmias or obstruct coronaries resulting in myocardial ischaemia, or disrupt normal haemodynamics. Ruptured aneurysmal sinus of Valsalva is more common among Asian women.¹¹ Shah *et al*¹² describe their centre's experience, revealing that a majority of their patients presented with an asymptomatic murmur (44.4%), 22.2% with acute chest pain, 22.2% with mild heart failure, 11.1% with severe heart failure and 11.1% with cardiogenic shock. The provisional diagnosis in our case of a ruptured ALSOV into the RA was unlikely due to the rarity of this anomaly, the lack of acute symptoms and signs of heart failure.¹³

Coronary vessels try to compensate for a reduction in myocardial blood flow distal to the site of connection of the fistula by

Table 1 Details of a full saturation run performed in our patient with a coronary cameral fistula

Chamber	Superior vena cava (SVC)	Inferior vena cava (IVC) (%)	Right atrium (RA)	Right ventricle (%)	Pulmonary artery (%)	Pulmonary artery wedge (%)	Aorta (%)	Pulmonary vein (%)
Oxygen saturation (PaO ₂ %)	High SVC: 67%, low SVC: 68%	73	High RA: 84%, mid RA=85.5%, low RA=78%	81	80	96	96.50	96

progressive enlargement of their ostia and the feeding artery causing aneurysmal malformation, as noted in the LMCA of our case.¹⁴

Coronary artery aneurysms occur in approximately 19%¹⁵ of patients with coronary artery fistulas. With time, they may be complicated by intimal ulceration, intimal rupture, atherosclerotic deposition, calcification and mural thrombosis.¹⁶

Although cardiac catheterisation with coronary angiography has been considered the gold standard for the diagnosis of coronary artery fistulas, recently, multidetector CT (MDCT) has gained acceptance as a better alternative, especially when dealing with fistulas coursing between vascular structures, similar to the situation of our case. The MDCT's superior spatial resolution and ability to show relationship of anatomic structures has led to its increased popularity.^{15 17}

However, a trans-thoracic echocardiogram is easily available and advised; it is commonly performed in almost all patients admitted to the CCU, especially in those with murmurs. In this case, perhaps most previous echocardiograms may have been done with the patient in active PSVT, hence the CAVF was probably missed (in the setting of tachycardia).

Again, in our case, the continuous murmur was hard to appreciate in the presence of a fast heart rate but a repeat clinical

examination after conversion to normal sinus rhythm was key to final diagnosis; it prompted for a repeat echocardiogram, a practice not commonly performed once patients admitted to the CCU stabilise, unless there is a new cardiovascular symptom.

All symptomatic patients with a coronary artery fistula should undergo closure by either a surgical or transcatheter approach. Catheter closure techniques have been performed to treat coronary fistulas with devices, including detachable balloons, stainless steel coils, controlled-release coils, controlled-release patent ductus arteriosus (PDA) coils, and Amplatzer PDA plug.¹⁸

Our patient's symptoms and associated aneurysmal LMCA favoured a surgical patch closure. An incidental discovery of two openings of the fistula into the RA further supported this line of management as being the best decision for this patient.

The natural course of untreated coronary artery aneurysms with coronary artery fistulas is obscure and their rupture rate is still unknown, but ruptured cases seem to be very rare.³

In conclusion, a case of coronary cameral fistulas presented as a rare cause of PSVT. However, a transthoracic and TEE, cardiac catheterisation with coronary angiography and a CT coronary angiogram helped confirm the diagnosis and plan definitive management; the importance of a diligent clinical examination in picking up a continuous murmur to identify anomalies such as coronary arteriovenous fistulas cannot be overemphasised. Further, echocardiograms should also be repeated after the patient converts to normal sinus rhythm or after controlling the heart rate to avoid missing such anomalies. Successful surgical correction as recommended for symptomatic large coronary cameral fistulas rendered our patient asymptomatic.

Figure 4 CT coronary angiogram showing a coronary cameral fistula (CCF) leading from the left main coronary artery (LMCA) into the right atrium (RA), traversing in between the aortic root (Ao) and the left atrium (LA).

Learning points

- ▶ Patients presenting with arrhythmias, especially tachyarrhythmias, must have a diligent clinical examination at presentation as well as after control of tachycardia.
- ▶ Patients with unidentified causes for arrhythmias and an unclear picture on preliminary echocardiograms should undergo repeat imaging studies once they are further stabilised, especially after control of tachycardia.
- ▶ Ruptured sinus of Valsalva aneurysm may be a rare differential diagnosis for coronary arteriovenous fistulas with an aneurysmal left main coronary artery, however, the lack of signs of acute heart failure would go against the former.

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