CASE REPORT

Woman with chest pain and shortness of breath since age 18

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SUMMARY

We present an interesting case of a 38-year-old woman who presented with a history of left-sided chest pain, dyspnoea and palpitations. The symptoms have been occurring since age 18, and were previously diagnosed as costochondritis. Because of the suspicious history, the patient underwent further cardiac investigations. The echocardiogram demonstrated an abnormal structure adjacent to the aortic valve. A diagnostic coronary angiogram revealed a large left circumflex artery fistulating into the right atrium. Surgery was performed to ligate the fistula. The patient recovered well and has been asymptomatic since.

BACKGROUND

This case was written to highlight the rare disease of coronary artery fistulae. It is also meant to educate the readers about the importance of taking a thorough history in patients with chest pain, especially the younger age patients, who we tend to overlook when it comes to diagnosing cardiac chest pain.

CASE PRESENTATION

A 38-year-old woman presented to the emergency department with ongoing left-sided chest pain, dyspnoea and palpitations. On further questioning, she described the chest pain as a tightness sensation that occurs intermittently. She was having this pain since the age of 18 and previously diagnosed as costochondritis. The chest pain has been progressively worsening and did not improve with the use of analgesics or non-steroidal anti-inflammatory drugs. The patient noted that her chest pain worsened at times of stress. She was a lifelong non-smoker and had no cardiac risk factors. She worked in a retail grocery store. Physical exam was essentially normal.

INVESTIGATIONS

ECG showed T-wave inversion in leads III, aVF, V4, V5 and V6 (inferior and lateral wall). Full blood count, urea and electrolytes were all within normal limits. Troponin T levels taken on admission and repeated 12 h later were both normal. Thyroid function tests were also normal. A 24-h Holter monitor did not reveal any arrhythmias. An exercise stress test was performed, at which the patient lasted 8 min and 19 s reaching Stage 3 Bruce protocol. The stress test was stopped because the patient developed chest pain, but there was no significant changes noted on the ECG. A transthoracic echocardiogram was performed which demonstrated an abnormal structure at the level of the left

coronary artery beside the aortic valve (figure 1 and video 1).

Because of the typical history of progressive chest pain with a typical character, a diagnostic coronary angiogram was performed. This revealed a large left circumflex artery that was fistulating into the left atrium (figure 2 and video 2). The other coronary arteries were normal.

The patient had a CT coronary angiogram performed. Just distal to the origin of the left circumflex artery, there was a large fistula which courses between the root of the aorta and the roof of the left atrium. The artery had a tortuous path before draining to the posterior aspect of the upper right atrium (figure 3). In addition, there appeared to be a small communication with the anterior wall of the left atrium at the distal aspect of the fistula.

A right heart catheterisation was performed with saturation measurements in different areas of the right heart (table 1). It demonstrated a step-up in oxygen saturation in the right atrium.

TREATMENT

Based on the above findings, the patient underwent open surgical fixation of the fistula. The fistula was found to drain into both the left and right atrium.

Figure 1 Still image of transthoracic echocardiogram in short-axis view showing the trileaflet aortic valve with the abnormal structure adjacent to it (circled).

To cite: Lobo R, Lim RY, Meany B, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-010118

Figure 2 Still image of left coronary angiogram cine showing the large left circumflex artery (LCx) and its relation with the left anterior descending artery (LAD) and right atrium (RA).

These were ligated. The patient was also found to have a secundum atrial septal defect, which was also closed.

OUTCOME AND FOLLOW-UP

The patient had recovered well and her symptoms had completely resolved. One year after surgery, the patient continued to do well and has not had any further chest pains.

DISCUSSION

Coronary artery fistula is defined as an anomalous connection between a coronary artery and a major vessel or a cardiac chamber. It was first described in 1841.¹ Communications between the coronary arteries and the cardiac chambers (coronary-cameral fistulae) or other vessels (coronary artery arteriovenous malformations) are often due to deviations from normal embryological development. They may also be acquired from trauma (stab, gunshot or projectile injuries), or from invasive cardiac procedures such as pacemaker implantation,

Figure 3 Coronal slice of the CT coronary angiogram showing the outline of the left circumflex artery (LCx) coursing from its origin across the root of the aorta (Ao) towards the right atrium (RA).

Table 1 Blood oxygen saturation measurements in various parts of the right heart during catheterisation

Location	Oxygen saturation (%)
Femoral artery	98
Superior vena cava	72.7
Right atrium	80.3
Right ventricle	76.5

endomyocardial biopsy or coronary angiography.² Its incidence in the general population is about 0.002%.³ Some cases are identified because of an associated murmur, which is loud, superficial and continuous.⁴

Video 1 Transthoracic echocardiogram in short-axis view showing the trileaflet aortic valve with the abnormal structure adjacent to it.

Video 2 Left coronary angiogram cine showing the large left circumflex artery (LCx) and its relation with the left anterior descending artery (LAD) and right atrium (RA).

The major sites of origin are the right coronary artery (55%), the left coronary artery system (35%) and both coronary arteries (5%). The major termination sites with right and left coronary artery origins are the right ventricle (40%), right atrium (26%) and pulmonary arteries (17%).⁴

Most coronary artery fistulae are small, myocardial blood flow is not compromised and the patient is asymptomatic. However, there may be coronary artery steal, with resultant ischaemia of the segment of myocardium perfused by the coronary artery distal to the fistula. If untreated, haemodynamically significant fistulae may result in clinical symptoms or sequelae in approximately 19% of patients under the age of 20 and 63% percent of those over the age of 20.⁵ Possible sequelae include angina, myocardial ischaemia, pulmonary hypertension, endocarditis, thrombosis of the fistula or an associated aneurysm.⁴

Treatment involves surgical ligation of the fistula. There are also various percutaneous catheter techniques that have been employed for the closure of these fistulae.

Learning points

- ▶ Coronary artery fistulae are a rare but important cause of cardiac chest pain in young patients.
- ▶ Be wary of a young person reporting recurrent chest pain. Thorough history taking is important.
- ▶ If a patient provides a convincing history of chest pain, more advanced cardiac investigations should be undertaken, regardless of the age.

Acknowledgements Authors would like to thank Dr Abid Hussaini who was involved in the management of the patient in the hospital. The University Hospital Limerick Echocardiography department.

Contributors RL was involved in research and writing of the manuscript. RYL was involved in writing of the manuscript, BM and TJK contributed by editing the manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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