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### RESEARCH ARTICLE

#### ATYPICAL CHEST PAIN: A CASE REPORT.

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#### Abstract

A 56-year-old female patient was referred to our institution for atypical chest pain and palpitations. Physical examination, resting ECG and transthoracic echocardiogram were unremarkable. Stress perfusion scintigraphy was positive for anterior and apical myocardial ischaemia. A subsequent coronary angiogram showed no signs of atherosclerotic coronary artery disease; however, it revealed a coronary arteriovenous fistula and multiple other fistulous connections between the proximal segment of the left coronary artery and the pulmonary artery trunk. We present a rare case of a symptomatic coronary fistula that was percutaneously closed using an Amplatzer Vascular Plug, which resulted in clinical improvement and late fistula occlusion. This case report underlines the importance of thinking beyond atherosclerosis in the evaluation of chest pain syndromes. Moreover, it describes some of the angiogram caveats in assessing the coronary fistula number and morphology, as well as the cardiac-catheter potential for multiple pathway coronary artery fistulae closer.

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#### Background:-

Coronary arteriovenous fistula (CAVF) varies widely in its morphological appearance, clinical presentation and long-term outcome. Fistulae are congenital or acquired coronary artery abnormalities in which blood is shunted into a cardiac chamber or other vessels, bypassing the physiological myocardial capillary network. The CAVF location and dimensions determine their haemodynamic significance and clinical presentation, which can include dyspnoea, angina, congestive heart failure, pulmonary hypertension, arrhythmias or myocardial infarction. A majority of CAVF describe a simple trail, arising from the right coronary artery or the left anterior descending coronary artery, and lack clinical significance. However, larger fistulae may lead to coronary artery steal, with resultant ischaemia of the myocardium or other sequelae. Treatment is indicated for symptomatic patients and for those asymptomatic patients who are at risk for future fistula related complications. Although percutaneous CAVF closure is currently being used more frequently, a surgical approach is still considered the most suitable technique for larger, multiple fistulae or those associated with ventricular tachyarrhythmias or myocardial ischaemia.

CAVF usually presents at an earlier age, and although coronary atherosclerosis is by far a more frequent diagnosis in middle-aged patients with chest pain, the possibility of congenital coronary fistula must not be under-rated.

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### Case presentation

A 56-year-old woman with a medical history of Hashimoto's thyroiditis and mitral prolapse was referred to our institution because of mild palpitations and long-term atypical chest pain (non-effort-related anterior chest discomfort with no radiation and no associated symptoms). The patient denied a history of arterial hypertension, hypercholesterolaemia or diabetes mellitus. Physical examination was unremarkable, with no audible murmurs on cardiac auscultation.

### Investigations:-

Basal ECG showed frequent supraventricular premature beats. The transthoracic echocardiogram presented mild mitral regurgitation, with non-dilated right and left ventricles/atria, no regional wall abnormalities and good left ventricle systolic function.

She had a previous stress perfusion scintigraphy, which was positive for significant anterior and apical myocardial ischaemia. As a result, a coronary angiogram was performed, which unveiled normal coronary origins and no signs of atherosclerotic coronary artery disease. However, it also revealed a coronary artery fistula between the proximal segment of the left coronary artery and the trunk of the pulmonary artery. Several other fistulae connections were also visualised in relation to the main CAVF. However, the smaller fistulae were difficult to characterise due to their complex three-dimensional (3D) geometry and reduced contrast media fill.

### Differential diagnosis:-

The differential diagnosis of patients presenting with chest pain is extensive, ranging from benign musculoskeletal aetiologies to life-threatening cardiac disease. Reports of the causes of chest pain include musculoskeletal (36–49%), cardiac (15–18%), gastrointestinal (8–19%), pulmonary (5–10%) and psychiatric disorders (8–11%).<sup>1</sup> However, coronary atherosclerotic disease is by far the most common aetiology of myocardial ischaemia, considering our patient's age. Concerning supraventricular premature beats, it can occur frequently in individuals with normal hearts, and it is thus difficult to establish a definite relationship with other disorders or to delineate the factors that predispose to this type of extra cardiac beats. Furthermore, the incidence of supraventricular premature beats is variable in different forms of structural heart disease, and is often found among patients with mitral valve prolapse.<sup>2</sup>

### Treatment:-

Since the angiogram findings were compatible with those of the previously performed stress perfusion scintigraphy (anterior/apical wall motion abnormalities in the myocardial region supplied by the left coronary artery), the patient was referred to CAVF closure. However, following a refusal (by the patient) of the standard surgical approach, she was subsequently called for percutaneous catheter CAVF closure.

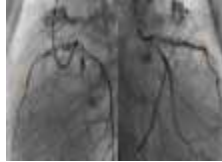
Access was obtained with a femoral arterial catheter. Hand injection of contrast allowed visualisation of coronary anatomy and location of fistulae including the origin and drainage sites. The main fistula access was done using a Choice Extra Support guidewire and a previously sized Amplatzer Vascular Plug 4 was successfully deployed, with no complications. After main fistula embolisation, the other fistulous vessels were clearly visualised. Figures 1–3 show the coronary fistula before, during and after the closure procedure. Access to the remaining fistulae collaterals was attempted using several coronary guidewires; however, their small diameter and tortuosity precluded entry for a coil or other embolic devices. Although complete CAVF occlusion was not possible, the main fistula branch was successfully closed and the small collaterals seemed not to be clinically significant.



**Figure 1:-** Coronary angiogram before fistula closure. \*, Coronary arteriovenous fistula; Cx, circumflex coronary artery; LAD, left anterior descending coronary artery; LM, left main coronary artery.



**Figure 2:-** Coronary angiogram during main fistula closure. A, Amplatzer Vascular Plug.



**Figure 3:-** Coronary angiogram after closure of main fistula. \*, Coronary arteriovenous fistula; A, Amplatzer Vascular Plug; Cx, circumflex coronary artery; LAD, left anterior descending coronary artery.

### Outcome and follow-up:-

At 1-month of follow-up, the patient was asymptomatic and 1 year later, she underwent a multidetector CT (MDCT) angiography that confirmed complete closure of the main CAVF, no signs of the fistula collaterals and no coronary arterial density in the pulmonary artery.

### Discussion:-

CAVF is the most frequent congenital coronary abnormality, with a prevalence of 0.002% in the general population and 0.25% in patients undergoing a coronary angiogram.<sup>3 4</sup> 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 are the right ventricle (40%), the right atrium (26%) and the pulmonary arteries (17%).<sup>5</sup> Most coronary artery fistulae are small and produce no clinical manifestations. However, some might produce ischaemia of the segment of the myocardium perfused by the coronary artery distal to the fistula (through a coronary artery steal) and/or the left-to-right shunt leading to progressive right ventricle dilation and heart failure. Other reported cardiac complications are pulmonary hypertension, endocarditis, malignant arrhythmias and rupture of the fistula.

Although the diagnosis of CAVF may be challenging, many are identified following the detection of a continuous murmur at the lower or midsternal border, depending upon the site of drainage. The chest x-ray and ECG are usually normal, but may show cardiac chamber enlargement and ischaemia. Significantly enlarged coronary arteries can be detected by 2D echocardiography, and a transoesophageal echocardiogram has been used to detect the entrance and termination sites of the shunt.<sup>6</sup> Nonetheless, the standard procedure to evaluate CAVF is coronary angiography, which establishes the size and anatomical features of the fistula.

The American College of Cardiology/American Heart Association (2008) guidelines<sup>7</sup> for the management of adults with congenital heart disease recommend the closure of all large coronary artery fistulae regardless of the symptomatology using transcatheter or surgical techniques. The guidelines also support the closure of small to moderate fistulae in the presence of symptoms, such as ischaemia, arrhythmia, endarteritis and unexplained systolic or diastolic dysfunction. Conversely, an asymptomatic small fistula with no haemodynamic significance does not require further treatment.

Some controversy persists regarding the management of medium-sized/small-sized clinically silent CAVF. One must consider what is known about the risks of non-intervention and the long-term effect of any available intervention. There are several factors associated with CAVF prognosis; the most important ones may be the relative size of the fistula, the presence of symptoms or complications related to the fistula, the age of the patient and the fistula's anatomy/complexity. Symptoms and complications of coronary artery fistulae are less common in children and become more significant in adults.<sup>8</sup> It is acknowledged that coronary fistulae tend to get larger over time, which may lead to potential complications, with cardiac problems developing in approximately 19% of patients under the age of 20 and in 63% of those over 20 years.<sup>9</sup> Conversely, spontaneous closure has been also reported in patients with small-size or medium-size fistulae, particularly in neonates.<sup>10 11</sup> Furthermore, Sherwood *et al*<sup>12</sup> investigated the prognostic significance of clinically silent coronary artery fistulae, and found that none developed symptoms over time and 23% had spontaneous closure based on either echocardiogram or angiography. However, there are no good

large and long-term comparative studies of a significant cohort with coronary artery fistulae treated by specific strategies or left untreated.<sup>13</sup>

Although the symptoms and complications of CAVF are markedly affected by size and the great emphasis on fistula diameter, evaluation has been made by the current guidelines, coronary artery size definition is still controversial and has not been done systematically. Moreover, the absolute CAVF size seems to be a poor measure approach, since a relatively small diameter fistula may have no significant effect in an adult, though it may have a more detrimental effect in a neonate or child.<sup>14</sup> Accordingly, Latson proposed calling coronary artery fistulae 'small' if the fistula was not larger than twice the proximate normal coronary artery diameter, 'medium' if bigger than twice but smaller than three times the size of the proximate and 'large' if of even bigger dimensions.<sup>10</sup> This is the standard measure method used in our practice.

In the present case, the patient complained of long-term atypical chest discomfort and palpitations, which were thought to be a likely manifestation of a mitral prolapse syndrome. Physical examination and ancillary tests were unremarkable, with the exception of the stress perfusion scintigraphy. The presence of angina/myocardial ischaemia related to CAVF at such a late stage is a very uncommon form of presentation.

There are several approaches that can be used to manage coronary artery fistulae. Procedural options and planning can be optimised by careful identification of the number of fistulous connections, site of the fistula's origin and termination, and ischaemic burden or left-to-right shunt severity. However, as the present case depicted, it is not always possible to accurately establish CAVF complex morphology using a conventional coronary angiogram. As reported elsewhere,<sup>15-17</sup> in the case of complex CAVF anatomy, coronary angiography may not delineate the course of the anomalous vessel due to its complicated 3D geometry that is fluoroscopically displayed in two dimensions. On the other hand, MDCT angiography not only allows an accurate and non-invasive depiction of the origin, course and termination of the fistulae, but also it has been shown to be superior to conventional angiography in the delineation of the ostial origin, the proximal path of the anomalous coronary arteries and the drainage site of CAVF. It more accurately displays the complex vascular relationships with 3D and multiplanar reformations (3D reconstructions). In the present case, MDCT angiography was only used to confirm the long-term closure of CAVF. Although this method can accurately evaluate the main fistula channel occluded with the Amplatzer device, it may be inappropriate in assessing the other residual small fistulae that persisted after CAVF closure.

The first reported successful attempt of surgical closure of a coronary artery fistula was by Biorck and Crafoord.<sup>18</sup> Surgical correction might be indicated when CAVF is characterised by high fistula flow, multiple communications, very tortuous pathways, multiple terminations and/or significant aneurysmal formation and for those undergoing operative repair of other cardiovascular problems.<sup>15</sup> The surgical approach can be also indicated for reduction in the size of very large aneurysmal dilations of either the fistula or the proximal coronary artery.<sup>19</sup> Overall, mortality related to the surgical closure of isolated coronary artery fistulae is low (<1%) and recurrence or incomplete closure may occur in about 10% of the cases.<sup>19, 20</sup> However, most of the recurrent or residual shunts have been small and repeated surgical procedures are rare.

Transcatheter closure was first reported by Reidy *et al*<sup>21</sup>, and has been emerging as an alternative in patients with favourable anatomy (non-tortuous vessel, single fistula and easily accessible).<sup>22</sup> Overall, the early outcomes have been similar to surgery with low morbidity and mortality. A variety of devices have been used including coils, detachable balloons, covered stents, vascular plugs (as in this case—Amplatzer Vascular Plug 4) and atrial septal defect closure devices. The most frequent complication associated with catheter-based closure<sup>23</sup> includes myocardial infarction and migration of the devices to extracoronary structures or coronary artery branches (the latter occurring in 7 of 40 patients, as per Qureshi and Tynan<sup>24</sup>). Although most of the series reported an excellent procedural success rate, data are lacking on the short-term and long-term recanalisation rates of percutaneously treated patients with coronary artery fistulae. Recently, Jama *et al*<sup>13</sup> reported a rare series of 29 patients who underwent CAVF percutaneous closure and had a follow-up angiogram at a median of 1.5 years after the initial procedure. These authors reported that 56% of patients had no recanalisation, 22% had trivial recanalisation and 22% had significant recanalisation. These recanalisation rates are comparable to those reported by Cheung *et al*<sup>8</sup> in the surgical literature. Thus, the importance of routine follow-up after fistulae closure is unequivocal, even in asymptomatic patients. Compared with other modalities such as an echocardiogram or a CT scan, a coronary angiogram has the advantage of allowing direct device visualisation and sizing of recanalisation (none, trivial and significant).<sup>13</sup>

**Learning points:-**

1. We have presented a case of symptomatic multiple pathway fistulae associated with myocardial ischaemia. Although the percutaneous closure approach is not the first therapeutic option in patients with multiple fistulae or drainage sites due to procedure complexity/inappropriateness, we should bear in mind that the occlusion of the main fistula may improve symptoms and prevent disease progression and life-threatening complications, especially in those patients reluctant to go in for standard cardiac surgery.
2. Although coronary atherosclerotic disease is by far the most common aetiology of myocardial ischaemia, physicians should look beyond atherosclerosis in patients with atypical presentations and low overall coronary risk.
3. While coronary artery fistulae are usually congenital abnormalities and most often present during infancy or childhood, they may occasionally present in adulthood or even in elderly patients. Myocardial ischaemia, right ventricular dilation and dysfunction, arrhythmias and infective endocarditis are potential associated complications.
4. Percutaneous closure of the coronary artery fistulae is a safe and effective therapy in small to middle-sized coronary fistulas without marked tortuosity or multiple fistulous connections.

**Footnotes:-**

**Competing interests:-** None.

**Patient consent:-** Obtained.

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