Case Report

Large coronary arteriovenous fistula presenting with infective endocarditis and regurgitation of the aortic valve – an atypical presentation of a rare condition

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Abstract

Coronary arteriovenous fistulae are rare congenital anomalies which usually present in adulthood with congestive cardiac failure. Infective endocarditis is an infrequent complication of the condition. Further, aortic valve involvement, leading to aortic regurgitation is a rare occurrence. We report an atypical case of a 40 year old male whose initial presentation was pyrexia of unknown origin which was later found to be due to infective endocarditis of the aortic valve complicated by aortic regurgitation. Echocardiography and further imaging with CT revealed a large left coronary arteriovenous fistula which was the primary pathology leading to the infective sequelae. The patient underwent surgery for replacement of the aortic valve and closure of the fistula.

Introduction

Coronary arteriovenous fistula is a rare anomaly, which by definition involves a sizable communication between a coronary artery and one of the four cardiac chambers or a segment of either the pulmonary or systemic vasculature, bypassing the myocardial capillary bed [1,2]. This usually occurs in isolation and is found in approximately 0.002% of the general population [1,3]. Usually these fistulas are of congenital origin, while rarely they may be iatrogenic. Krause was the first to describe these in 1865 and Bjork and Crafoord performed the first surgical management in 1947[4].

Case report

We report a case of a 40-year-old male who was referred to the cardiothoracic surgical unit III of the National Hospital of Sri Lanka with aortic infective endocarditis with severe aortic regurgitation. He had been investigated for pyrexia of unknown origin in a medical unit and echocardiography revealed severe aortic regurgitation with vegetations in all three aortic valve cusps.

The patient was in New York Heart Association Class I. On examination, he was afebrile with a tachycardia of 106 beats/minute and blood pressure of 110/50 mm Hg. He also had a grade 4 high pitched early diastolic murmur best heard in the left 3rd intercostal space. Laboratory investigations revealed mild normochromic normocytic anemia, ESR of 80 mm and CRP of 85 mg/dl. Although blood culture was sterile, Gram stain revealed chains of Gram positive cocci.

Transthoracic echocardiogram (TTE) revealed a mildly dilated left ventricle with 55% ejection fraction and severe aortic regurgitation with vegetations in all three aortic valve cusps. Trans-oesophageal echocardiography (TOE) localized an aneurysmal structure close to left coronary sinus with features of an anomalous coronary. Invasive cardiac catheterization was deferred initially as the aortic vegetations were fragile carrying a significant risk of embolization. Contrast enhanced cardiac tomography revealed that there was a large coronary AV fistula arising from the left main coronary artery draining to the right atrium (Figure 1).

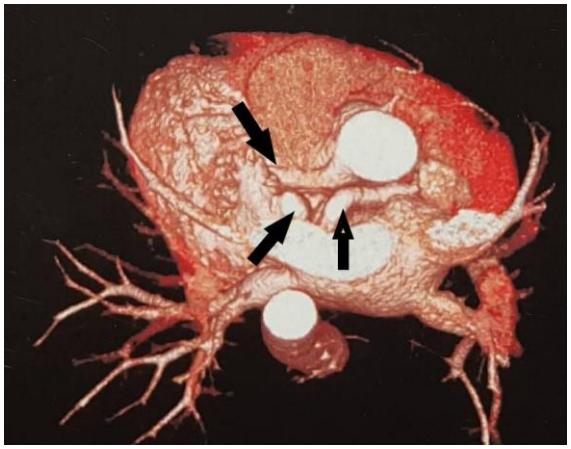

Figure- 1: Coronary CT angiogram reconstructed view
Arrows show the arteriovenous fistula

A limited coronary artery angiogram was done to delineate the exact anatomy of the AV fistula and the configuration of coronary arteries (Figure 2). It confirmed the finding of the coronary fistula arising at the origin of the left circumflex artery. The patient underwent surgery on cardiopulmonary bypass and cardioplegic arrest. At surgery the left coronary ostium was found to be massive and the large epicardial arteriovenous fistula was identified (Figure 3).

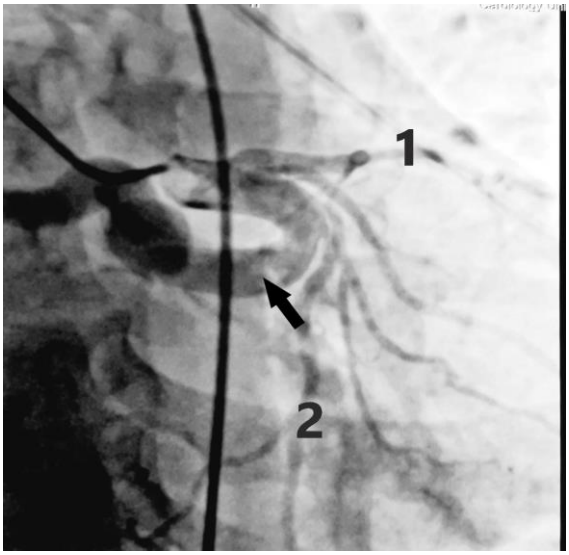

Figure- 2: Coronary angiogram; Arrow – Left AV fistula, 1 – Left anterior descending artery (LAD), 2 – Left circumflex artery

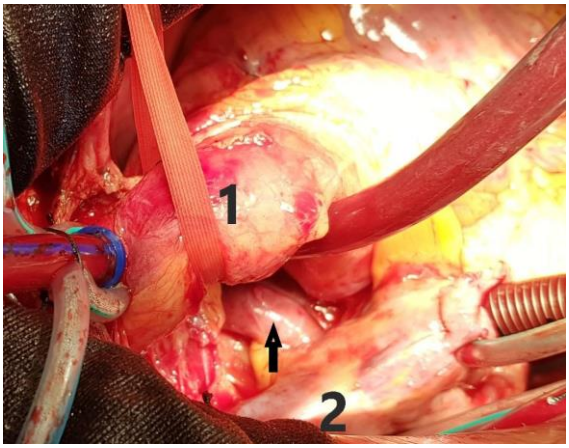

Figure- 3 : Coronary AV fistula in the transverse sinus; Arrow – AV fistula, 1 – Aorta, 2 – Superior vena cava

It was travelling posterior to the main pulmonary artery and the aorta through the transverse sinus to drain in to the right atrium - superior vena cava junction. AV fistula was clamped to prevent cardioplegia shunting through the fistula.

The fistula was isolated and closed at the origin taking care not to narrow the circumflex artery. Damaged aortic valve leaflets with the vegetations were excised and the aortic valve was replaced with a size 21 St Jude mechanical prosthetic valve. The patient had an uneventful postoperative recovery and was started on anticoagulation therapy with warfarin.

Discussion

Coronary arteriovenous fistulae can have variable morphology depending on the site of origin, laterality, site of drainage and branching pattern. The right coronary artery is the origin in about 52% of cases, followed by the left anterior descending coronary in approximately 30% and the circumflex coronary artery in 18%. Over 90% of the fistulas drain to the right side of the heart[5].

Fistulae draining to the right sided cardiac chambers cause volume overload of the right heart, the pulmonary vascular bed, the left atrium and the left ventricle. This results in dilation of cardiac chambers. The size of the fistula and the pressure gradient between the coronary artery and the draining chamber determine the size of the shunt [1]. The shunting of blood may lead to congestive cardiac failure and rarely, ‘steal’ of blood from adjoining myocardium may lead to myocardial ischaemia.

A patient with a small fistula and minimal shunting may remain asymptomatic. A moderate sized fistula may slowly increase in size with time and subject the left ventricle to volume overload. Patient will typically develop symptoms of cardiac failure from third decade of life. In rare cases the presentation may be due to a complication of the fistula, like intraluminal thrombosis, infective endocarditis, angina due to ‘steal’ and very infrequently rupture of an aneurysmal fistula.

In the illustrated case the cardiac functions were relatively preserved and not significantly affected by the shunt at the time of diagnosis, which is interesting considering the wider diameter of the shunt and the expected shunt volume [6]. In the reported case the fistula presented with infective endocarditis, which is a rare but known complication of a coronary AV fistula. The incidence of infective endocarditis as a complication of coronary arteriovenous fistulae is approximately 5% [7].

The pathophysiology is attributed to the turbulence near the coronary sinus at the coronary ostium which is likely to be the substrate of endocarditis in this patient.

In an analysis of 25 reported cases of coronary arteriovenous fistulae complicated with infective endocarditis by Said et al, echocardiographic evidence of valvular vegetations were detected in 16. Aortic valve was the site of infection in 4 of the cases [8].

Suspicion of a possibility of a fistula was first raised on TOE. Further imaging was needed for confirmation and to define the exact anatomy of the fistula. Although coronary angiography remains the gold standard imaging study for the coronary arteries it doesn't readily visualize the relations of other cardiac structures to the vessel [2]. CT coronary angiography was a good non-invasive alternative in this situation.

In the current literature it is widely recommended by most of the authors that symptomatic coronary arteriovenous fistulas should be treated percutaneously or surgically [4]. According to the American College of Cardiology and American Heart Association Guidelines for the 'Management of Adults with Congenital Heart Disease', it is a Class I recommendation to close large fistulae regardless of symptoms. Small- to moderate-sized fistulae require surgical intervention if they are associated with complications [9]. Accordingly, urgent surgical intervention was indicated for this case considering the size of the fistula, the risk of systemic embolization carried by the fragile vegetations, severe aortic regurgitation and poor response to intravenous antibiotics.

At surgery the location, size and the pathological state of the fistula should be noted [6]. It is advised by most authors to go on cardiopulmonary bypass in patients with dilated fistulae that can cause severe bleeding, in inaccessible ones and in fistulae with aneurysms [6].

In our patient, administering direct cardioplegia in to the coronary ostia was necessary due to the aortic valvular incompetence. Furthermore, occluding the fistula with digital pressure and a clamp distal to the origin of the left circumflex artery was needed to maintain pressure of the cardioplegia, by preventing its direct leakage in to the right atria.

The best technique suitable to close a fistula depends on its origin, distal end, length, flow status, size, tortuosity, aneurysmal dilation [10]. A narrow fistula opening in to a low pressure chamber such as the right atrium can be exposed at the distal end and closed from within the chamber. In contrast, when the fistula is wide and giving out major coronary branches as in the patient discussed here, closing from the distal end may cause thrombotic occlusion of the coronaries. Furthermore, it is essential to prevent recurrence due to recanalization [6,10] .

We isolated and closed the fistula with plageted sutures at the distal end through the right atriotomy approach. Further, it was ligated at the transverse sinus distal to the origin of the circumflex artery.

A good prognosis is indicated for both surgical and percutaneous management strategies of coronary AV fistulae. Life expectancy following surgical correction remains normal while about 4% of patients may need further surgery for recurrence [10].

Conclusion

Coronary arteriovenous fistula is a rare yet, significant clinical entity due the high morbidity and mortality of the disease sequelae and associated complications. Atypical presentations should prompt further investigations and possibility of infective and other complications should be considered. Due to its high morbidity, surgical or transcatheter interventions should be considered in all patients with clinical presentations and asymptomatic patients with large fistulae.

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