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CASE REPORT

Coronary Artery Fistula Presenting as a Rare Cause of Heart Failure

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Abstract

Coronary artery fistula (CAF) is an abnormal connection between one of the coronary arteries and a heart chamber or another blood vessel. The communication of the coronary artery could be with the lumen of a cardiac chamber (right ventricle - 41%, right atrium - 26%, left atrium - 5%, left ventricle - 3%), coronary sinus (7%), superior vena cava (SVC) (1%), pulmonary artery (17%) or the pulmonary vein. Here we present a rare case of CAF draining in SVC, which presented as heart failure, but on subsequent workup showed presence of fistula which was then surgically corrected successfully.

Key Words

Coronary artery fistula, Heart failure, Superior vena cava

Introduction

Coronary artery fistula is an abnormal connection between one of the coronary arteries and a heart chamber or another blood vessel. Many literatures report the key role of invasive coronary angiography in CAF assessment. This modality could not provide a clear fistula visualization due to contrast medium dissipation in high-pressure vessels and should be recommended only for patient treatment (1).

The right coronary artery (RCA) is the most common artery involved in fistulous formation (50% of cases) and is often symptomatic usually in 5th decade, presenting as either ischemic or fluid overload states. Left coronary artery (LCA) accounts for 42% of fistulas and is often asymptomatic. Both RCA and LCA are involved in 5% of cases (2). Majority of fistulas are single, but multiple and complex fistulas have been reported. Cardiovascular anomalies are associated in 5%-30% of cases (3). Here we present a rare case of CAF draining in SVC.

Case Report

A 72-year-old female presented with dyspnea on exertion (New York Heart Association class IV) for 1 month and gradually developed orthopnea. She had history of hypertension for 3 years. On presentation her vitals were normal, grade IV/VI pansystolic machinery murmur best heard in the left parasternal area radiating towards right chest, and mild pedal edema. She was managed in emergency as a case of acute LVF. TTE revealed normal valves, normal left ventricular ejection fraction (>60%) with moderate to severe PAH with mild turbulent flow at RCA ostium, then TEE was done which showed RCA aneurysm with high velocity flow, bicaval view showed 39*34 mm aneurysmal dilatation of RCA at opening of SVC (*Figure 1*). Conventional angiography was done which showed normal coronary arteries with a fistula arising from LCX and marked aneurysmal dilatation of RCA was seen. Computed tomographic coronary

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Figure 1: TEE View Showing Aneurysmal Dilatation

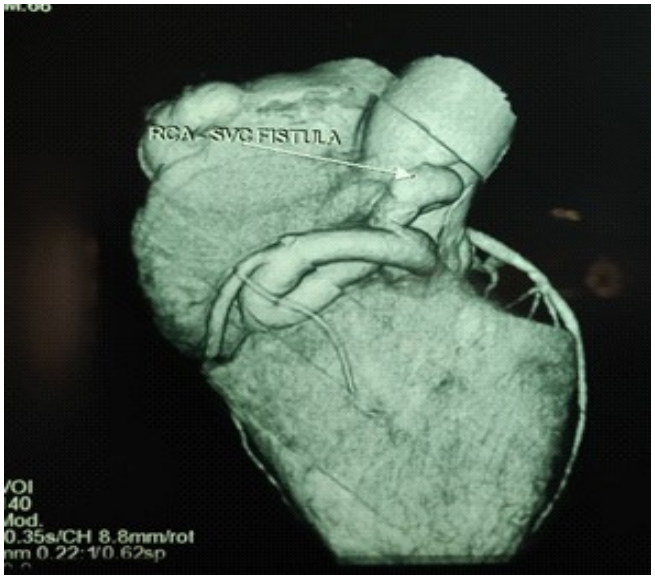

Figure 2: CT Angiography Showing Fistula Draining into SVC

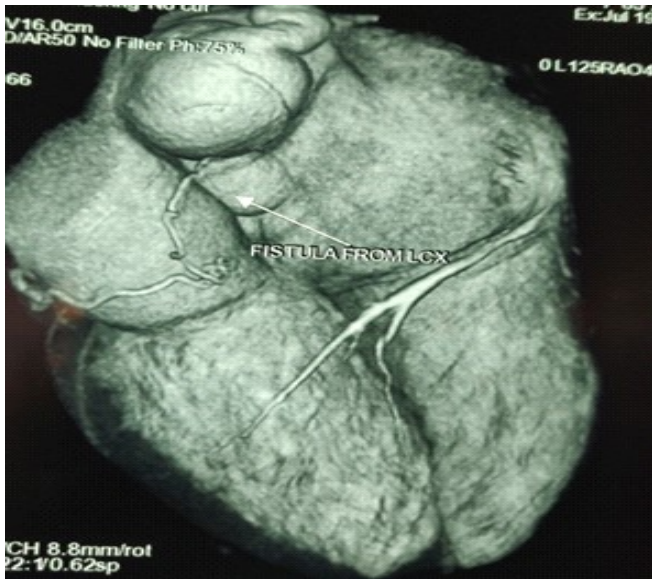

Figure 3: Fistula Arising from Left Circumflex Artery

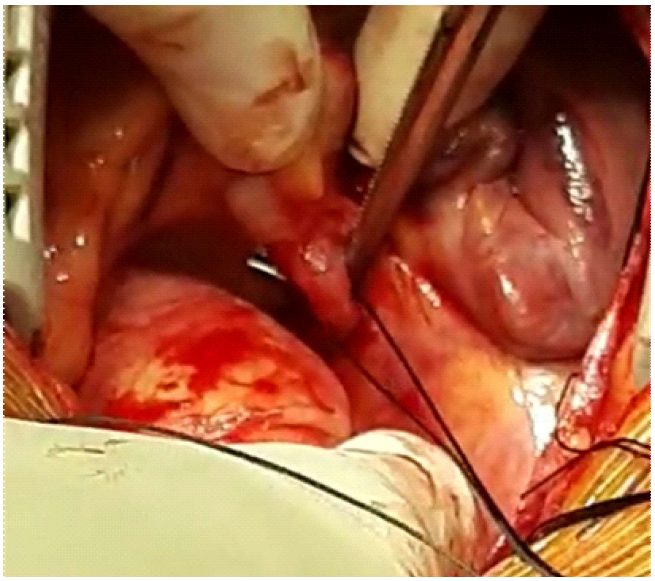

Figure 4: Aneurysmal Sac Ligation

angiography was done to look for drainage of fistula, which showed no significant obstructive disease, but showed significant dilation of RCA, with fistula draining into SVC (*Figure 2*), also small tortuous fistula was seen arising from small branch of LCX (*Figure 3*).

All the fistulae drained into the SVC on lateral surface at RA junction. The patient was taken up for surgery in view of her clinical symptoms. Patient was put on CPB, intraoperative findings signified that main aneurysm had a common wall with SVC (*Figure 4*). After giving cardioplegia the large RCA fistula was dissected going towards RA then towards SVC on lateral surface (aneurismal sac bulging present). Tapes of SVC and IVC was snuggled tightly, the SVC and RA were opened away from SA node. Large RCA fistula opening into SVC at lateral surface seen. Size 6-8 mm round opening which was closed with Teflon pledgeted 4-0 prolene suture and also ligated externally large RCA fistulous tract going in

medial portion of RA with silk tie in 2-3 places (*Figure 4*).

Large aneurismal sac opened which was bulging in SVC, RA inside portion. There was small left circumflex opening. Probe was inserted and cardioplegia was given, even then blood was coming through fistula opening, which was ultimately ligated with 4-0 pledgeted prolene suture. The aneurismal sac was repaired with 4-0 prolene in two layers, bulging was not there. Patient was rewarmed, RA/SVC incision closed in layers, and tapes released, heart filled, aortic venting done, aortic cross clamps released. Repeat TEE done, shows no collection in aneurismal sac. Patient's hemodynamics remained stable; closure of chest done after successful weaning from bypass. Patient had an uneventful post op. period. She was symptomatically better, is now being followed up on OPD basis.

Discussion

CAFs can be classified according to their origin, drainage site, or complexity. The right coronary artery (RCA) is the most common origin site of CAFs accounting for 50% to 55% of cases. The left anterior descending artery (LAD) is involved in about 35% to 40% of cases, while the LCX, as for the reported case, for the 5% to 20% (4,5).

Our patient had a very uncommon presentation showing RCA aneurysmal dilatation with fistulous tract draining in SVC. Such an anomaly, however, may exist, as demonstrated by this report, and may easily be confused with patent ductus arteriosus, aortopulmonary window, sinus of Valsalva aneurysm, pulmonary arteriovenous fistula, and even rheumatic valvular disease. While predominantly congenital, CAFs can be acquired in the setting of cardiovascular disease such as coronary atherosclerosis and MI, or be iatrogenic in origin and occur after procedures such as PCI, cardiac implantable electronic device insertion, and endomyocardial biopsy (6,7).

Small to moderate fistulas are associated with myocardial ischemia, arrhythmia, endarteritis, unexplained ventricular diastolic or systolic dysfunction, and ventricular dilatation (8). Early and prompt management of such conditions can result in significant decrease in morbidity and improves quality of life.

Conclusion

Heart failure should be evaluated extensively, especially the ones with normal ejection fraction and normal coronaries on conventional angiography with no significant stenosis. Early and prompt diagnosis with such scenarios where coronary artery fistula is the cause behind heart failure can be operated and managed in the golden hour and life-threatening condition can be resolved, thereby adding new presentation scenario to the literature.

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Nil.

Conflicts of Interest

There are no conflicts of interest.

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