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"Double Steal Phenomenon": A rare case of coronary fistula from LAD to main pulmonary artery in the presence of SVG to LAD

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Introduction

Coronary artery fistula (CAF) is a rare form of congenital cardiac anomaly. A few embryological postulations have been associated to explain the origin but the exact mechanism is still unknown. The prevalence of CAFs seen at computed tomographic (CT) angiography is reported to be as high as 0.9% and 0.002%–0.3% at invasive angiography [1-4]. In a review by Papadopoulos DP et al, 50% of the fistulas were found to arise from the RCA, 42% from the left coronary artery, and 5% from both coronary arteries. The most common site of drainage is the right ventricle (41%), followed by the right atrium (26%) and the pulmonary artery (17%) [5]. Coronary steal phenomenon is the term used to explain angina or myocardial ischemia during exercise in patients with CAF. This is due to reduced perfusion distal to the fistula. The extent of the shunt is determined on the basis of the fistula size and the pressure gradient between the coronary artery and drainage area[6].

Case Summary

54 year old lady with Diabetes Mellitus and Hypertension since 2014. She had Coronary artery bypass graft (CABG) surgery on 23/1/2009 at another private hospital where single vessel grafting i.e saphenous vein (SVG) to LAD was done. She first saw me in November 2015 to continue her care after her previous doctor passed away. She was well until July 2017 when she was admitted with chest pain. Her Troponin was negative. ECG was normal. Dobutamine stress echo was positive at inferior wall and she had chest pain during peak dose but no ECG changes. Coronary angiogram and graft study was then performed on 20th July 2017. Coronary angioplasty was performed to mid LAD after anastomosis (70-80% stenosis) and mid LCx (70-80% stenosis) with Biofreedom 2.75 x 18 and Biofreedom 3.0 x 14 stents respectively. Large coronary fistula was noted from proximal LAD into pulmonary artery. At the time decision was made not to intervene as symptoms was thought to be due to LAD and LCX diseases. SVG to LAD was widely patent with some retrograde flow into the fistula.

She presented as outpatient on 15th August 2021 with progressive worsening of shortness of breath since early July 2021.In early Stage II of Treadmill ECG test, she had severe dyspnoea until she let go the treadmill handle bar and fell down. No syncope and no ECG changes were noted. CT coronary angiogram was done on 16th August 2021 which revealed patent stents in LAD and LCX arteries as well as SVG to LAD. Fistula networks seen from LAD connecting to main pulmonary artery (Fig 1). We then proceeded to coronary angiogram on 20th August 2021 which revealed patent stents in mid LAD and mid LCX. Large fistula was noted from proximal LAD into main pulmonary artery. Flow and size of fistula vessel network were larger than that in 2017(Figure 2a-b). SVG to LAD was patent but showed progressive higher retrograde flow in the proximal LAD fistula into pulmonary artery (Figure 2c) in comparison with the fistula flow in 2017 (Figure 3: images in 2017). In antegrade injection, it appeared as proximal LAD was totally occluded as blood flow into the fistula(Figure 4). This phenomenon was likely due to combined factors of competitive flow from the SVG to LAD plus the existence of the large fistula. She decided against having another open heart surgery thus we proceeded with percutaneous intervention. Coiling procedure was performed with two Penumbra Ruby coils 4.0 mm x 15 cm and 3.0 mm x 15 cm which were delivered through 2.6F PX Slim microcatheter. Post coiling angiogram showed much reduced flow in the fistula (Figure 5a-c). Immediately after the procedure patient felt better. She described it as new 'feeling relieved' in term of her breathing since many years ago.

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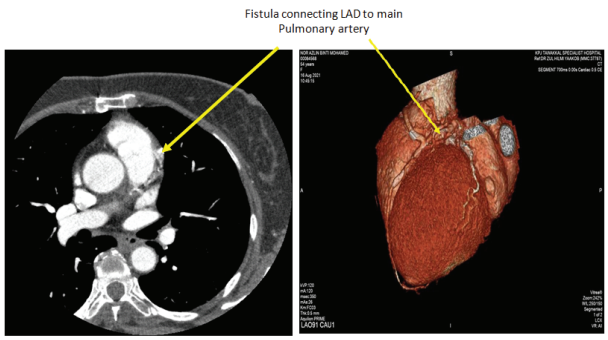

Figure 1: Fistula Connecting LAD to main pulmonary artery

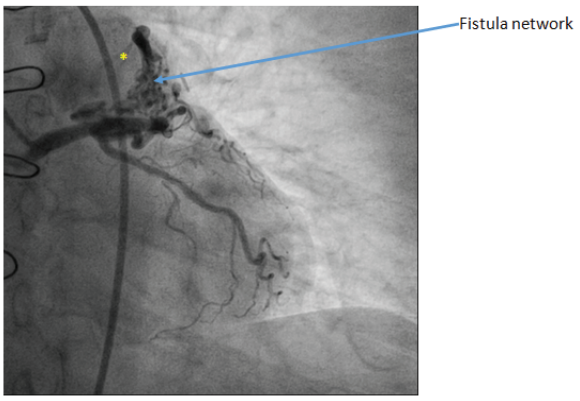

Figure 2a: Angiogram images in 2021(*flow from fistula into pulmonary artery)

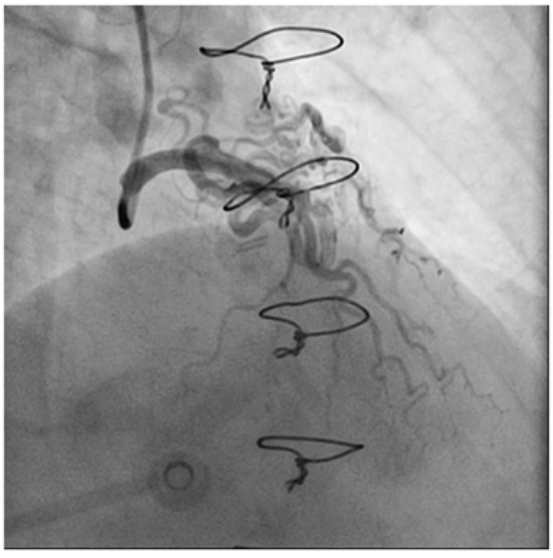

Figure 2b: Angiogram images in 2021

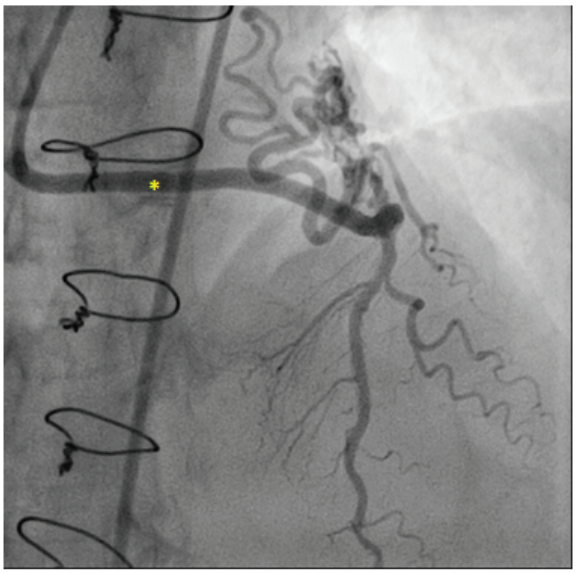

Figure 2c: SVG to LAD (*)

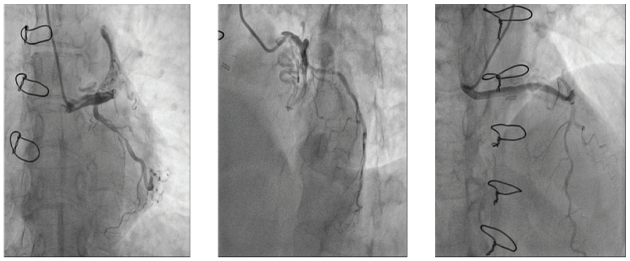

Figure 3: Images in 2017

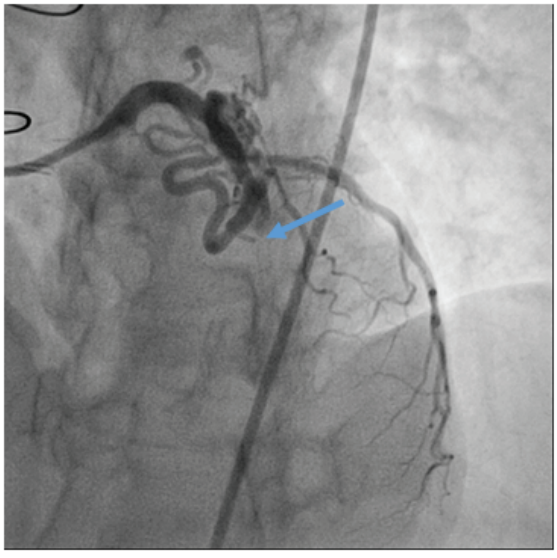

Figure 4: No antegrade flow from proximal LAD distal to fistula (arrow)

Discussion

This patient had coronary bypass surgery with single SVG to LAD. The indication and decision for the bypass surgery remained controversial. The author would like to state that it was done more than 10 years ago in another hospital. Nevertheless, the highlights of this case are:

I. This case illustrated a very rare case of a fistula between the left anterior descending artery and the main pulmonary artery [7,8].

II. Although rare, CAFs is an important cause of myocardial ischaemia especially in the presence of large fistulas.

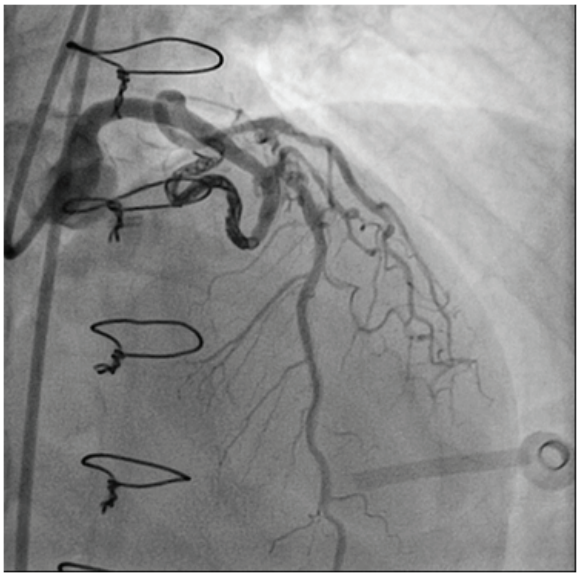

Figure 5a: Post coiling

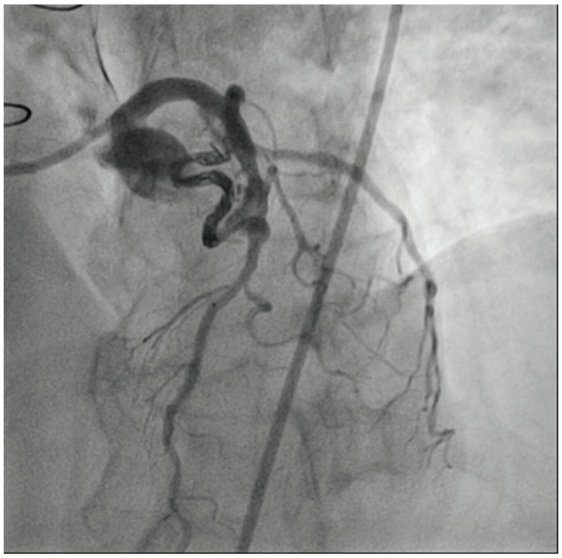

Figure 5b: Post coiling

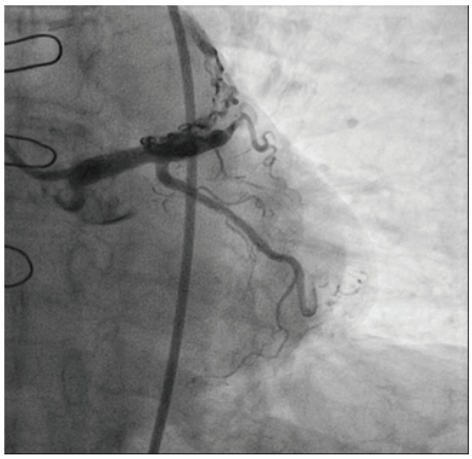

Figure 5c: Post coiling. Some residual fistula flow

III. The presence of SVG to LAD in this case did not reduce her symptoms as large volume of retrograde flow from SVG-LAD went into the fistula. In fact this may have contributed to the progression of the fistula network of vessels due to increased flow from LAD both antegradely and retrogradely (from SVG) thus explaining the worsening of her symptoms over time. This was evident in the comparison of angiogram images of the CAF flow between those in 2021 and 2017.

IV. In term of management, the optimal management of symptomatic patients with coronary artery fistulae still remains controversial.This patient had open heart surgery in 2009 (CABG) and from the beginning had expressed firm reluctance for another surgery. Thus surgical ligation of fistula was out of option. This case illustrated the success of percutaneous closure of fistula using coiling device.

Conclusion

Coronary artery fistula albeit rare remains an important cause of myocardial ischaemia. This case is unique where it showed all antegrade flow from LAD went into the fistula with the 'help' of competitive flow from SVG to LAD. Although the SVG to LAD was widely patent, she was still symptomatic since there significant flow into the fistula retrogradely in LAD from her SVG. This illustrated the unique 'double steal phenomenon'. Immediate improvement in symptoms demonstrated the success of percutaneous method of fistula closure in this case.

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