Introduction

Coronary artery anomalies are relatively rare in the general population with prevalence about 1% (1). Coronary artery fistula (CAF) is a type of coronary artery anomaly in which an abnormal vessel originating from a coronary artery bypasses the normal capillary bed and terminates within the great vessels or cardiac chambers. The prevalence of CAFs on CT coronary angiogram (CAG) is 0.9% (2-4). Bilateral coronary pulmonary artery fistulas is an extremely rare occurrence. We report a case of acute coronary syndrome with bilateral coronary pulmonary artery fistulas.

We present the following case in accordance with the CARE reporting checklist (available at http://dx.doi.org/10.21037/jxym-20-82).

Case presentation

A 65 years diabetic male patient with no other comorbidities and no significant past and family history presented with anterior wall ST elevation myocardial infarction (STEMI). Cardiac examination was clinically normal. Echocardiogram showed regional wall motion abnormalities in the anterior wall with ejection fraction of 48%, no chamber dilatation and normal pulmonary arterial pressure. Chest X-ray was normal. Patient was treated with fibrinolytic agent. Subsequently CAG was done which showed significant lesion in the proximal left anterior descending (LAD) artery with bilateral CAF from left main (LM), proximal LAD artery and right coronary artery (RCA) to main pulmonary artery. Percutaneous coronary intervention (PCI) with drug eluting stent (DES) to LAD artery was performed. CT CAG was done following PCI which showed bilateral CAF. Bilateral CAF was hemodynamically insignificant (normal pulmonary artery pressure and Qp/Qs:1.2) hence conservative management was advised. This is a rare case of atherosclerotic coronary artery disease presenting as STEMI with bilateral coronary pulmonary artery fistula.
of vessels originating from LM, proximal LAD and RCA had fistulous communication to main pulmonary artery (Figures 1-4, Videos 1-4). Percutaneous coronary intervention (PCI) was performed to LAD with drug eluting stent (DES) (Figure 5, Video 5) and no intervention for CPAF in view of small and multiple vessels which were hemodynamically insignificant (normal pulmonary artery pressure and Qp/Qs:1.2). Patient was discharged with uneventful recovery. CT CAG was done also revealed bilateral coronary to pulmonary artery fistula (Figures 6-8). On follow-up after 4 months patient remained symptom free and Ecg, Echo were normal. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

**Discussion**

CAFs are usually congenital or rarely acquired connections between the coronary vessels and cardiac chambers or other vascular structures such as vena cava, pulmonary artery or pulmonary veins. The incidence of CAF is 0.1% to 0.2% in all patients undergoing CAG. These fistulas may arise from any of the major coronary arteries however in most cases CAF arises from the RCA. CAFs may affect hemodynamic parameters (3) and can cause various complications including arrhythmias, myocardial ischemia, infective endocarditis and heart failure in adults (5). CAFs were first reported in
1865 (6) by Krause, followed by Haller and Little (7), author described clinical triad of a CAF: a cardiac murmur, a large tortuous coronary artery and atrial or ventricular left-to-right shunt. The incidence of CPAFs has increased as cardiac CT angiography has become widely used, with these fistulas constituting 15–30% of all CAFs (4,8). The most accepted embryologic explanation for a CPAF is the Hackensellner involution-persistence hypothesis. According to this theory among the six branches of the truncus, only two branches from the aortic sinus remain to form the coronary arteries, while rest of the branches involutes. A CPAF is formed due to abnormal persistence of branch of pulmonary sinus and connects to the normal branches from the aortic sinus coronary arteries, pulmonary sinus normally involutes during embryonic life (4). At cardiac CT angiography CPAF shows contrast shunt sign appearance of an abnormal contrast blush, in a relatively less opacified pulmonary trunk or as a well visualized fistulous tract between the pulmonary trunk and coronary artery (9). According to a study by Verdini et al. (9), two types of CPAF occur. The first type is multiple small calibre fistulous connections from the RCA or LAD, which connects to the main pulmonary trunk. Second type is a single prominent fistulous connection between the LAD or RCA and the main pulmonary trunk. A CPAF with multiple connections is less likely to have hemodynamic disturbance related symptoms than single prominent fistulous connection. According to Kim et al. (4), CT angiography shows 59% of CAFs have multiple pulmonary artery origins. This is higher than other studies with conventional coronary angiography which reported 11–16%. The difference may be due to the higher spatial resolution of CT (4). The Vieussens ring and the collateral pathway between the conus branches of the RCA and LAD is often seen along the pre pulmonary trunk in cases of multiple arterial origins in patients with coronary to pulmonary fistulas. In a study by

**Figure 5** Coronary angiography right anterior oblique view reveals the fistula between the proximal LAD artery and the main pulmonary trunk with DES in the proximal LAD artery. LAD, left anterior descending; DES, drug eluting stent.

**Figure 6** Computed tomography thorax shows small multiple leash of vessels from LM coronary artery to pulmonary artery. LM, left main.

**Figure 7** 3D CT angiogram shows fistulous connection between LAD artery to main pulmonary artery with stent in the proximal LAD artery. LAD, left anterior descending.

**Figure 8** 3D CT angiogram shows fistulous connection between LAD artery to main pulmonary artery with stent in the proximal LAD artery. LAD, left anterior descending.
Verdini et al. (9), a total of 103 CPAF patients were evaluated ages ranging from newborn to 88 years. According to this study multiple coronary arteries are rarely involved. Most common involved artery is RCA. This series demonstrates that multiple fistulas are commonly present, with the most common pattern being between the LM/LAD and the main pulmonary trunk. Bilateral fistulas originating from both the coronary systems account for 5% of the total. These types of fistulas terminate more often into the pulmonary artery (56%) than the unilateral fistulas (17%) (10).

In our patient who presented with acute coronary syndrome STEMI with proximal LAD stenosis, initially fibrinolysis followed by PCI (pharmacoinvasive) was done. Incidentally bilateral CAF to pulmonary artery was noticed during PCI and was treated conservatively. On follow-up after 4 months patient remained symptom free. Hemodynamically significant fistula may present with angina due to coronary steal, cardiac failure, arrhythmias. If the patient has hemodynamically significant CPAF following PCI on follow-up if he presents with angina then either instent restenosis or coronary steal phenomenon should be considered. We are reporting a rare case of bilateral CAF to main pulmonary artery with acute coronary syndrome.

Conclusions

We are presenting a case of bilateral CPAF with STEMI which was treated with pharmacoinvasive therapy and conservative management for CPAF. CAF is thought to be present in 0.002% in general population. Multiple fistulas such as in our case being much rarer occurring in 10% to 16% of all CAFs, with involvement of both coronaries is less than 5%. Bilateral coronary pulmonary artery fistula with acute coronary syndrome is very rarely reported in the literature to the best of our knowledge. The main indication for closure of CAF are patient symptoms, heart failure, coronary ischemia, significant shunting. Treatment options include observation, only for hemodynamically significant and symptomatic patients transcatheter embolization or surgical intervention.

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Footnote

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Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

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References


