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A rare case of double coronary artery fistula from left circumflex artery draining to the left atrium in a rheumatic heart disease patient

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Introduction: Coronary artery fistula (CAF) is a connection between one or more of the coronary arteries and a cardiac chamber or great vessel. This is a rare defect and occurs in only 0.2% of the general population. Most of the cases are congenital but acquired causes of the defect were also reported. A CAF may occur as an isolated cardiac defect or may occur with other cardiac diseases such as rheumatic heart disease (RHD). However, only a few cases of co-existing CAF and RHD have been reported in literature. Local data only reported that congenital anomalies involving the coronary arteries only comprise 0.69% of congenital malformations of the heart. Moreover, only 61 patients among all patients who underwent coronary arteriography in 34 years were reported to have a coronary artery fistula. In this case report, we account the second case of coronary artery fistula admitted in our institution.

Case: A 46-year old female with rheumatic heart disease with severe mitral stenosis came in due to progressive dyspnea. The coronary angiogram revealed a two fistulous tracts originating from the left circumflex artery draining into the left atrium. She underwent mitral valve replacement surgery with closure of the drainage of the fistula at the left atrium and left atrial plication. The postoperative course was uneventful and she was discharged. She was compliant to her regular follow-up.

Discussion: A CAF is often asymptomatic until the second decade of life. However, if not treated early, this may progress and cause ischemic and heart failure signs and symptoms. The presence of mitral stenosis caused elevated LA pressure which might have prevented increase in the volume of blood draining from the left circumflex artery to the LA through the fistulas. Hence, mitral stenosis might have prevented the dilatation of the two fistulas. Surgical correction is indicated in the fistula of this patient since if only the mitral valve replacement was done in this patient, resolution of the mitral stenosis will decrease the LA pressure which might result to dilatation of the fistulas and may later on complications.