

Case Report:

A life-threatening association of coronary anomalies

Tom Devasia,¹ Mohan V. Sumedha Maturu,² Hashir Kareem,¹ M. Sudhakar Rao¹

Departments of ¹Cardiology, ²Medicine, KMC, Manipal

ABSTRACT

Anomalous origin of the left coronary artery from the right sinus of Valsalva, even though rare, has been documented well in literature. However the association of this anomaly with coronary fistulae has been rarely reported so far.

We report the case of a 76-year-old female who presented to us with exertional dyspnoea. General physical and cardiovascular examination revealed no significant abnormalities. All laboratory investigations were normal. Chest radiograph was normal. Electrocardiogram showed left bundle branch block. Echocardiogram revealed a globally hypokinetic left ventricle with reduced ejection fraction. Coronary angiogram showed anomalous origin of left coronary artery from right coronary sinus along with a small coronary-cameral fistula connecting obtuse marginal artery to left ventricle, there was no significant stenosis of epicardial coronaries. This case report, documents the rare association of an anomalous coronary origin of left coronary artery with coronary fistula.

Key words : *Coronary vessel anomalies, Fistula*

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INTRODUCTION

Anomalous origin of the left coronary artery (LCA) from the right sinus of Valsalva has been documented to cause myocardial ischaemia, particularly when the left main artery courses between the aorta and pulmonary trunk. This condition is seen in otherwise asymptomatic young adults who present with syncope, angina or sudden death during or immediately after exertion. Coronary-cameral fistulae are another set of rare congenital anomalies which have been described with respect to termination of coronaries. Here we report the case of a patient in whom anomalous origin of LCA was associated with coronary-cameral fistula. Coronary steal phenomenon caused by coronary-cameral fistula can further aggravate the ischaemia caused due to anomalous coronary arteries and thus this association may be highly fatal. To the best of our knowledge, such an association has rarely been reported and thus prompted us to report this case.

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Corresponding author: Dr M. Hashir Kareem, Assistant Professor, Department of Cardiology, KMC, Manipal, India.
e-mail: hashirkareem@gmail.com

CASE REPORT

In October 2014, a 76-year-old lady presented to our cardiology out-patient service complaining of effort intolerance and dyspnoea on exertion which was increasing for the past 3 months. She reported no other relevant cardiovascular symptoms like chest pain, syncope, orthopnoea, paroxysmal nocturnal dyspnoea, and pedal oedema. She was receiving treatment for hypertension. Patient was active individual previously without any limitation of daily activity. Her vital parameters were stable and general physical examination was unremarkable. Cardiovascular examination also revealed no significant abnormalities. Rest of the physical examination was unremarkable.

Laboratory investigations including complete blood count, serum biochemistry including renal and hepatic function testing were normal. Electrocardiogram (ECG) showed left bundle branch block. Previous ECG was not available



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and so we were unable to assess if it was a new onset bundle branch block. Echocardiography revealed global hypokinesia with a left ventricular ejection fraction of 44%. Suspecting ischaemic heart disease she was taken up for coronary angiography.

Coronary angiogram in left anterior oblique (LAO) cranial view with catheter engaging and injection into right sinus of Valsalva showed origin of both left main coronary artery and right coronary artery (RCA) from the right sinus of Valsalva. Ostia of both left and right coronary arteries were simultaneously seen on injecting dye into right sinus of Valsalva (Figure 1). Faint filling of right coronary artery was also evident (Figure 2).

Coronary angiogram in right anterior oblique (RAO) caudal view with catheter engaging and injection into left coronary ostium showed long course of LCA which divided into a type I left anterior descending artery (LAD) and a large left circumflex (LCx) artery. A large obtuse

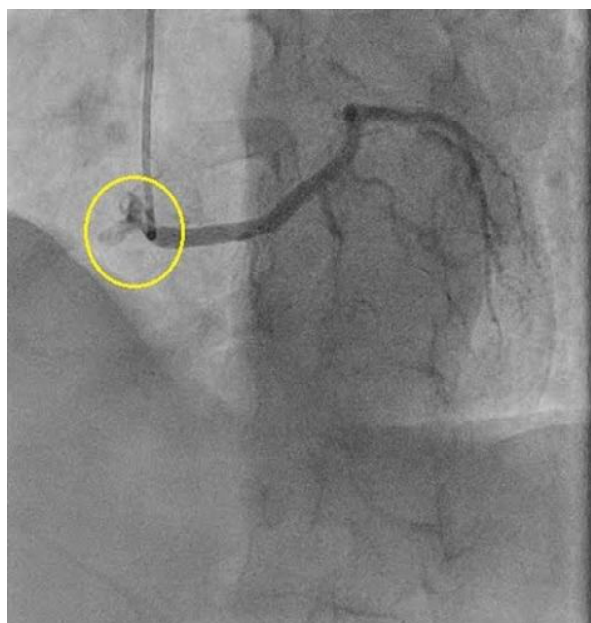


Figure 1: Coronary angiogram in LAO cranial view with catheter engaging and injection into right sinus of Valsalva. Yellow circle in picture shows ostia of both left coronary artery and right coronary artery in the same injection

LAO = left anterior oblique

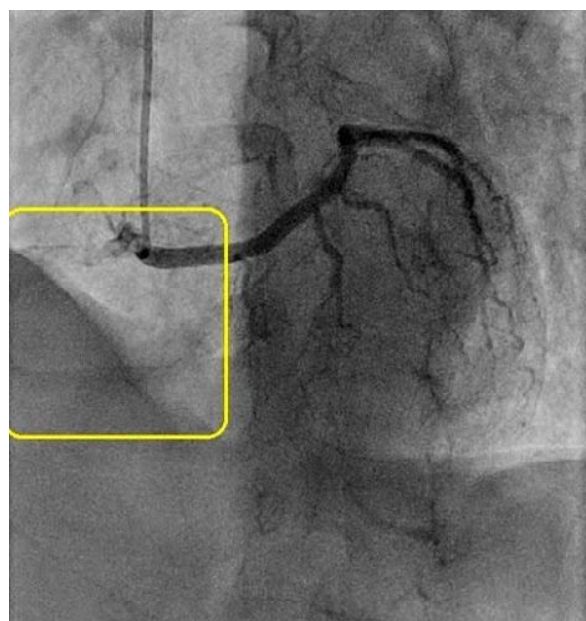


Figure 2: Coronary angiogram in LAO cranial view with catheter engaging and injection into right sinus of Valsalva. Yellow box in picture shows faint filling of right coronary artery simultaneously in the same injection

LAO = left anterior oblique

marginal (OM) branch was also seen arising from the LCx artery and a coronary-cameral fistula seen between OM branch and Left Ventricle (Figure 3) All the visualised coronary arteries were normal in calibre and flow, with no evidence of occlusion.

As she never had episodes of syncope or suffered angina and as the fistula was small with all the epicardial coronaries having normal flow, she was advised conservative management and was advised to avoid strenuous activity. As her ejection fraction was low, she was discharged with vasodilators and was advised general heart failure measures. Her symptoms improved with conservative management. After 3 month follow-up, the patient never had recurrence of symptoms.

DISCUSSION

Incidence of coronary artery anomalies in healthy individuals ranges from 0.3%-1%.¹ Patients usually present with angina or syncope, especially with exercise. Unfortunately, the presenting clinical symptom may be sudden

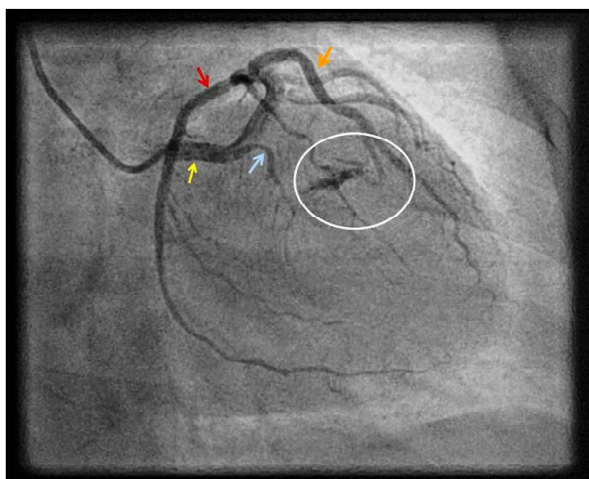


Figure 3: Coronary angiogram in RAO caudal view with catheter engaging and injection into left coronary ostium. Yellow arrow shows left main coronary artery which gest divided into type I left anterior descending artery (light blue arrow) and a large left circumflex artery (red arrow). A large obtuse marginal branch (orange arrow) is seen arising from left circumflex and coronary cameral fistula (white circle) is seen between OM branch and left ventricle

RAO = right anterior oblique; OM = obtuse marginal

death, particularly in young athletes, especially during conditions where myocardial demand increases as in during exercise.²⁻⁴ Among the anomalous coronary origins which are also referred as abnormal aortic origin of coronary arteries (AAOCA), the anomalous origin of LCA from right sinus of valsalva is the rarest and clinically most significant as it presents with sudden death. The mechanism for ischaemia can either be due to compression of vessel during its subsequent course between the aorta and pulmonary artery to the left ventricle or it may be secondary to acute angle of origin of anomalous coronary artery that may lead to obstruction of blood flow.⁵

Coronary fistulae are another set of anomalies which are rarely observed. A coronary fistula is communication between a coronary artery and either a chamber of the heart (coronary-cameral fistula) or any segment of the systemic or pulmonary circulation (coronary arterio-venous fistula). Major sites of origin of fistula are the right coronary artery (55%), left anterior

descending (35%), both coronary arteries (5%), and others being rare. Major termination sites are the right ventricle (40%), right atrium (26%), pulmonary arteries (17%) and less frequently the superior vena cava or coronary sinus and least often the left atrium and left ventricle.⁶ The mechanism by which coronary artery fistula cause ischaemia is myocardial stealing or reduction in myocardial blood flow distal to the site of the coronary artery fistula which is related to the diastolic pressure gradient and runoff from the coronary vasculature to a low-pressure receiving cavity.⁷ Haemodynamically significant fistulae may result in clinical symptoms or sequelae which include chronic myocardial ischaemia, myocardial infarction, heart failure, pulmonary hypertension, endocarditis, rhythm abnormalities. Thrombosis of the fistula or an associated aneurysm, rupture of the fistula can also occur but are rare.

Importance of this case report is that the patient presented at older age than average population. She never had episodes of syncope or angina. She was found to have anomalous origin of left main coronary artery which is rarest among the anomalous origins and in addition, she also had coronary cameral fistula which was arising from obtuse marginal branch terminating into left ventricle which has rarely been reported in literature so far. Both conditions can individually cause myocardial ischaemia and together the probability of myocardial infarction is much higher. Yet, she did not suffer myocardial infarction as flow in all coronaries is normal and fistula is small.

Treatment for AAOCA is usually surgical which is indicated for symptomatic patients, particularly with origin of the left coronary artery from the right sinus, such as those with serious ventricular tachyarrhythmia or documented myocardial ischaemia. There are no controlled studies that have evaluated the

outcome of intervention in asymptomatic individuals. Documented interventions include coronary artery bypass grafting (CABG) or unroofing (marsupialization) of the coronary artery to prevent compression from a change in the angulation.⁸ Intracoronary stents can also be used as an alternative to CABG in symptomatic patients with ischaemia.

Treatment of coronary fistulae includes closure of fistulae. Fistulae which are clinically silent and not associated with other abnormal findings may not require further treatment. Large, haemodynamically significant fistulae should be closed by ligation, either surgical or via percutaneous catheter techniques. Smaller coronary fistulae tend to get larger with age. As a result, it is usually recommended that elective closure be performed early in patients who have symptoms or who are asymptomatic but have a continuous murmur or a systolic murmur with an early diastolic component.⁹

Patient who presented to us had neither suffered symptoms of myocardial ischaemia in the past nor had any abnormal cardiovascular findings on clinical examination. Despite her coronaries being anomalous in origin, calibre and flow within coronaries were normal and coronary cameral fistula was also small. Keeping in view of her previous asymptomatic period, she was not taken up for coronary stenting or ligation of fistula. She was advised life style modifications and general heart failure measures. Our patient had significant reduction in symptoms with medical management itself and so we also emphasize that despite the presence of fatal complications, it is the

patient's symptoms which guide our treatment and unnecessary interventions need to be avoided so that iatrogenic complications are reduced.

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