

Anomalous Left Coronary Artery Arising from the Right Coronary Sinus Presenting with Acute Coronary Syndrome

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Abstract

Approximately 80% of the coronary artery anomalies are not malignant and asymptomatic. Abnormal origin of the coronary artery has a variable presentation in the young population such as atypical chest pain or unexpected cardiac death. It is necessary to identify these anomalies, as lethal outcomes can be prevented with appropriate treatment and management. Left coronary artery abnormally arising from the right coronary cusp is rare and can be life-threatening if it follows the track between the aorta and pulmonary artery. The second most common cause of sudden cardiac deaths in the young population especially in athletes is coronary artery anomalies following hypertrophic obstructive cardiomyopathy. The case presented here is about abnormally arising left coronary artery following the course between the right ventricle outflow tract and aorta. The patient presented with the non-ST segment elevation-acute coronary syndrome (NSTEMI-ACS) for the first time but after 15 months he again presented with ST segment elevation-acute coronary syndrome (STEMI-ACS) for which he underwent Primary percutaneous coronary intervention of Right coronary artery and for anomalous left coronary artery he was managed conservatively as per patient request.

Keywords: NSTEMI; STEMI; Left coronary artery; Anomalous coronary artery origin; Coronary anomalies

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Introduction

It is estimated that 1-2% of the general population has congenital coronary artery anomalies [1-3]. The most common anomaly seen is abnormally arising right coronary artery from the left coronary sinus of Valsalva [4-7]. Left coronary artery abnormally arising from the right coronary cusp has a prevalence of approximately 0.05% [1]. Sudden cardiac death may occur if it follows the track between the aorta and the pulmonary trunk, as it can cause compression during exertion [1,3,6]. Presenting here a case report of a 35-year gentleman, presented with NSTEMI-ACS (**Figure 1**) for which he underwent left heart catheterization which showed abnormally arising left coronary artery from right coronary cusp. After 15 months he again presented with STEMI-ACS for which he underwent Primary percutaneous coronary intervention of Right coronary artery and for anomalous left coronary artery he was managed conservatively as per patient request.

Case Report

35-year-old gentleman presented to the emergency department of Tabba Heart Institute, Karachi on 20th February, 2019 with complaints of sudden onset of right arm pain while playing badminton for an hour. He had no prior risk factors for coronary artery disease. According to the patient, his father died of myocardial infarction in his 60s. The examination was unremarkable. He presented with NSTEMI-ACS. This gentleman again presented to the emergency department of Tabba Heart Institute, Karachi on 27th May, 2020 with complaints of sudden onset of left arm pain for an hour. The examination was unremarkable. Electrocardiogram was consistent with Inferior wall Myocardial infarction this time (**Figure 2**).

Differential diagnosis

Acute coronary syndrome, Anomalous origin of coronary artery



Figure 1 ECG showing normal sinus rhythm, heart rate of 75/min, Early repolarization changes in lead I, II, V6.



Figure 3 Large Left main coronary artery.

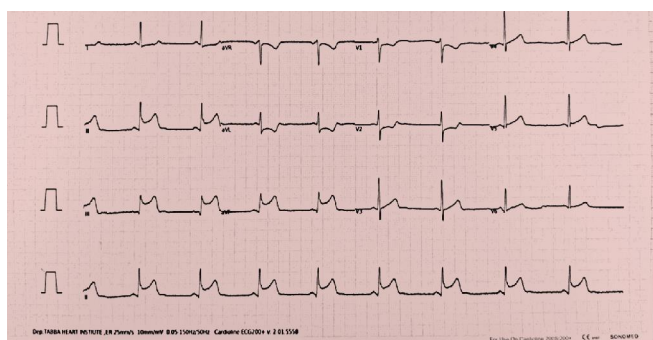


Figure 2 ECG showing normal sinus rhythm, heart rate of 55/min, 3 mm ST segment elevation in lead II, III, AVF.

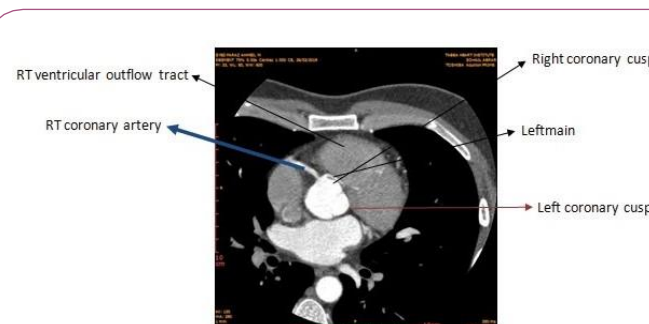


Figure 4 CT coronary angiogram showing origin of left main coronary artery from the right coronary cusp, and travelling in between aorta and right ventricle outflow tract.

Investigations

During the 1st admission baseline investigations including hematological and biochemical laboratory workup were within the normal range. The initial high sensitive Troponin was 1,198.3 g/ml and after 6 hours was 3,412.4 ng/ml (Normal range less than 0.4 ng/ml). Diagnostic coronary angiogram was performed which showed the Dominant right system, Left Main Coronary artery (LMCA) was extraordinarily large (**Figure 3**) and showed the anomalous origin of LMCA from right coronary cusp, 50% obstruction of a mid-left anterior descending artery, 40-50% obstruction of the mid-right coronary artery, Diagonal 2 plaguing. To further delineate the course, a Computed Tomography scan of coronaries was performed that showed the track of LMCA between the aorta and right ventricle outflow tract (**Figure 4**) just below the level of the pulmonary sinus. Patient written consent was taken prior to submission of case report. A detailed discussion with the patient and family was done for surgical correction. The patient refused surgery and opted for conservative management. Risk explained in detail to the patient for physical exertion and exercise-related aggravation of symptoms. The patient was discharged on beta-blockers, nitrates, and anti-platelets. Patient was loss to follow-up.

During the 2nd admission when the gentleman presented with Inferior myocardial infarction, the baseline investigations including hematological and biochemical laboratory workup were

within the normal range. The initial high sensitive Troponin was 390 ng/ml and after 6 hours was 2983 ng/ml (Normal range less than 0.4 ng/ml). Diagnostic coronary angiogram was performed emergently which showed the Dominant right system, LMCA was extraordinarily large and showed the anomalous origin of LMCA from right coronary cusp, 60-70% obstruction of mid left anterior descending artery, 80% obstruction of the mid right coronary artery, Diagonal 2 plaguing. Primary Percutaneous angioplasty of right coronary artery was performed. Next day fractional flow reserve of Left Anterior Descending artery (LAD) was done which came out to be positive with a value of 0.76. So it was planned that Coronary artery bypass grafting of LAD will be performed after 6 months of dual anti-platelets. Echocardiography showed normal ejection fraction with normal valves. A detailed discussion with the patient and family was done during this admission as well for surgical correction. Risk explained in detail to the patient for physical exertion and exercise-related aggravation of symptoms. The patient was still reluctant for surgery and was discharged on optimized medical treatment.

Discussion

The abnormally arising left coronary artery from right coronary sinus has four anatomical courses [2] that are main stem runs in

front of Right ventricle outflow tract (RVOT) or inter-arterial course or intra-myocardial septal continuation or runs posteriorly behind the aorta. The inter-arterial course has the worst prognosis. The most common clinical presentation is that with angina, syncope, dyspnea, palpitations, dizziness [8] that occurs mostly with strenuous exercise. In our patient, there were no prior symptoms until the time of presentation. But they can also present with an unexpected death in the younger population especially athletes. The commonest cause of death (13%) is the abnormally arising left coronary artery from the right coronary cusp as it was seen in a registry of 286 athletes. The pathophysiology is thought to be the compression of the abnormally arising coronary artery between the aorta and the pulmonary artery [9]. There is an obstruction to blood flow [3] of the affected artery [5]. Another explanation could be, during strenuous exercise, there is an increase in the aortic root and pulmonary trunk diameter that causes an increase in coronary artery angulation and thus reduces the lumen diameter in the proximal portion of the coronary artery [3,6]. Some studies showed that the myocardial ischemia has been caused by the acute angulations [5] of the anomalous coronary vessel. Also, the exercise-induced increase in diameter of the aortic root and the pulmonary artery compresses, resulting in further ischemia [7]. Sudden death in such cases can be explained by the possibility that frequent episodes of myocardial ischemia lead to life-threatening dysrhythmia. In our patient, prior ischemia was excluded as he had a normal left ventricular ejection fraction. Diagnosis is often difficult, as sudden cardiac death is often the first presentation. Others can be investigated for their symptoms like syncope or chest pain especially in the young population. Workup for diagnosis includes electrocardiography, Holter monitoring. Cardiac computed tomography or cardiac magnetic resonance imaging can be done as the next imaging modality. If the anomaly is confirmed, one should undergo nuclear stress testing to rule out exercise-induced ischemia. Treatment is variable and depends on the anatomy of the anomaly. Surgery is the mainstay of treatment, although medical treatment including beta-blockers and calcium channel blockers can be used to decrease ischemic symptoms. Symptomatic patients should be treated with beta-blockers [2] and should be advised to avoid strenuous exercise [8]. There is currently no defined treatment strategy and varies for individual cases. Coronary artery Bypass

Grafting by using the internal mammary artery as a conduit, re-implantation of the anomalous coronary artery to the relevant sinus, and finally unroofing is probably the treatment of choice especially in the inter-arterial course to prevent unexpected cardiac death. The unroofing technique is increasingly being used these days, as it is more physiological. In this report, we present a case with classical angiographic findings of abnormally arising left coronary artery from the right coronary cusp. He presented as a case of NSTEMI-ACS and later on with STEMI-ACS. The detail anatomy and the course was delineated in CT coronary angiogram for which he was advised to undergo surgery. A case report with abnormally arising left coronary artery from the right coronary cusp with NSTEMI-ACS has been reported [10].

Follow-up

The patient is doing well, walks daily for 3 km and is asymptomatic till the date 26th march 2021 and is still reluctant for surgery.

Learning Objectives

Abnormal origin of the coronary arteries may present with unexpected cardiac death, although some patients may remain symptoms-free. Abnormally arising left coronary artery from right coronary sinus with the inter-arterial path is a rare but important diagnosis. Surgical correction is always recommended for patients with an interatrial course of abnormally arising left coronary artery from right coronary sinus as these patients are at risk of life-threatening outcomes.

Conclusion

Abnormally arising coronary artery could be incidental findings. The clinicians should be aware of this condition. Since it is mostly asymptomatic and can lead to sudden cardiac death, especially when it takes an inter-arterial course, an aggressive treatment strategy should be planned. The individuals of having coronary artery anomalies should be referred to as cardiologist's timely appropriate management and successful prevention of sudden cardiac death can be done. Our patient was diagnosed at the age of 35 years with a sudden onset of symptoms. He remains at risk of the future life-threatening outcome if his anomalous coronary artery is not treated surgically as medical treatment is not treatment of choice.

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