Case Report

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Left Main Atresia: Supply from the right coronary circulation - a rare coronary artery anomaly

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Abstract

Coronary artery anomalies are rare incidental findings, about 20% with clinical significance of possible increased risk of related adverse cardiac events including sudden death. Multi detector computed tomography has become the first-line imaging modality to visualize the origin and course of anomalous blood vessels, assisting the further categorization and treatment recommendations. We present a case of 52 years old woman with left main atresia and right coronary artery supplying the entire cardiac muscles through the connection of posterior lateral artery to the distal segment of the left circumflex artery.

Keywords: Coronary Artery; Left Main Atresia.

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Background

Coronary artery anomalies occur in approximately 1% of the population, majority representing incidental findings without any clinical relevance.[1] However, few anomalies are associated with potential devastating consequences including angina pectoris, myocardial infarction, congestive heart failure and sudden cardiac death. This distinction is important, as hemodynamic significant coronary artery anomalies are the major contributor of sudden cardiac death in 33% of young adults.[2] Recently, multidetector computed tomography (MDCT) has emerged as a superior modality for identifying the origin and course of coronary artery anomalies compared to catheter angiography due to its visualization of non-cardiac anatomy and precise delineation of the vessel course.

Case report

Our case is a 52 years old, obese (BMI: 28.15kg/m2), female, with the history of anxiety disorder, hypothyroidism, fibromuscular dysplasia of the bilateral carotid arteries, which was diagnosed on cerebral angiogram recently. She is nonsmoker with no history of substance or alcohol abuse. She presented to emergency with chest pain for two weeks. Pain was dull sensation of something being stuck around the xiphoid and epigastric area, also occurring in the bilateral jaw. Symptoms were not exacerbated with activity, eating, or lying supine. She denied associated shortness of breath or palpitations. Family history of CAD was present in both parents, who underwent bypass surgery in their 50s, as well as a brother who suffered an MI at the age of 52. Electrocardiogram showed no significant ST-T changes. Echocardiogram showed normal LV systolic function with no regional wall motion abnormalities. Her physical findings were unremarkable and blood investigations were within the normal limits, including her troponin I. Given her family history and age, she was advised for coronary computed tomography (CT) to rule out presence of coronary artery disease.

Coronary CT angiography revealed atretic left main, which was visible in axial image as a thin strand measuring 0.9 mm in diameter and was continuous from the left coronary sinus to the bifurcation of the left anterior descending artery and left circumflex artery (Figure 1 and 2). The circulation to the left coronary arteries was provided by the right coronary artery through the communication between the posterior lateral branch of the right coronary artery and the distal end of the left circumflex artery (Figure 3). All the native arteries were slightly larger in size except the left main artery, which was atretic, and there were no atherosclerotic changes in the entire visible coronary branches. The arteries followed the normal course to their respective territory. Since her symptoms were unlikely to be caused by the anomalous coronary anatomy, she was advised for the regular follow up and counseled about the possible symptoms and consequences.

Figure 1: Axial image showing thin atretic left main artery.

Figure 2: 3D Volume rendered image showing the deficient normal anatomy of the left main artery.

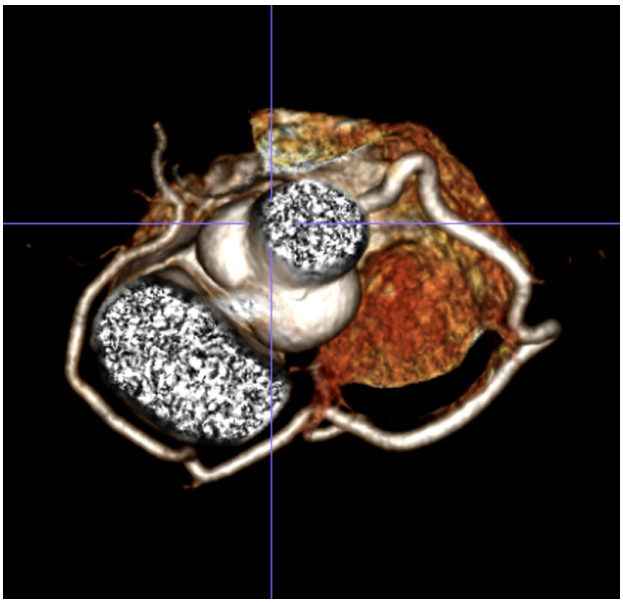

Figure 3: 3D volume rendered image showing the collateral circulation from right coronary artery to the left coronary arteries.

Discussion

Left main atresia is a rare entity that is accompanied by collateral circulation between left and right coronary arteries. The patients may present with coronary occlusive disease in childhood as the demand of left ventricle cannot be maintained. Patients can remain asymptomatic if the collateral circulation is providing the necessary blood to the heart. Left main atresia is different from single coronary artery that contains the single coronary ostium, but the distal portion of the artery is found in its normal anatomic location and communicates via a connecting branch with the normal artery arising from the sinus. The functional implication of these anomalies depends on the course of the connecting branch. There have been few case reports with presence of collateral circulations between right coronary artery and left anterior descending artery/ left circumflex artery. Our case has only one collateral connection between the left circumflex artery and right coronary artery. The patient had no prior ischemic symptoms signifying the normal supply of the blood to the entire cardiac muscles.

Invasive coronary angiography (ICA) has been the technique of choice for the diagnosis of the coronary anomalies, however, it has limited number of 2D projection images with absence of soft tissue details which limits its ability to identify the course of the arteries and the relation of the vessel to the other structures of the thorax. MDCT is now considered gold standard for the diagnosis of the coronary anatomy anomalies due to its accurate depiction of the origin and course of the anomalous vessels.[3] Diagnosing the anomalous origin as well as the course of the vessel is imperative to avoid future sudden cardiac deaths, potential lethal arrhythmias, and accidental coronary artery trauma during surgery. Proper diagnosis and timely intervention can prevent these risks. Percutaneous intervention as well as surgery has been performed in the patients with coronary anomalies with success.[4]

Coronary artery anomalies are rare incidental findings, with approximately 80% without any clinical significance and are considered benign. Potentially malignant variants include anomalous interarterial course, ostial stenosis, acute take-off angle, fistulous connections and anomalous left coronary artery from the pulmonary artery (ALCAPA). Our patient though had an atretic left main artery, but her coronary circulation was maintained by collateral circulation from right coronary artery to the entire left territory via connection between posterior lateral branch and left circumflex artery. However,

there is a risk of myocardial ischemia and sudden death due to sudden blockage of the right coronary artery, which will terminate the circulation to the entire heart. This entity should be kept in mind and patient should be educated about the possible risk and the probable ischemic symptoms if sudden occlusion occurs. The patients should also be aware of coronary artery disease risk factors and need of cardiovascular risk reduction for prevention of future events.

Conclusion

Left main atresia is a rare coronary artery anomaly that is often diagnosed incidentally. These anomalies though benign may have potential risk to the patient. MDCT is a powerful diagnosing modality that can help in diagnosing and potentially preventing from sudden outcome after timely correction.

Reference

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