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ORIGINAL RESEARCH

ANOMALOUS ORIGIN OF LEFT MAIN CORONARY ARTERY ORIGINATING FROM THE RIGHT SINUS OF VALSALVA: AN ANOMALY WITH FATAL OUTCOMES

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Abstract

Anomalous coronaries are often rare and incidental findings. Patients with coronary anomalies can have a wide array of presentations varying from chest pain to sudden cardiac death. Anomalous coronary artery originating from the opposite sinus (ACAOS) has a low prevalence, but with the advancement of imaging tools, its accidental detection has increased. Here, we describe such a rare case of a middle-aged man with a permanent pacemaker in situ who presented with recurrent syncope. Cardiac catheterization identified an anomalous origin of the left coronary artery from the right coronary sinus, and a computed tomography (CT) angiogram identified a high-risk course of the lesion.

Keywords: Acute coronary syndrome, anomalous coronary artery, computed tomography angiogram, sudden cardiac death, syncope.

Introduction

In adults, coronary anomalies are incidental findings at cardiac catheterization. Although the incidence of coronary anomalies is approximately 5.64%, not all are associated with poor outcomes [1]. ACAOS remains the most feared with 0.92% comprising of anomalous origin of RCA from the left sinus and 0.15% incidence of anomalous origin of left coronary artery (LCA) from the right sinus [2]. Since it is rare, the data on clinical presentation and management protocol is limited. Majority of these cases are asymptomatic but certain rare presentations of sudden cardiac death, myocardial ischemia, arrhythmias and syncope are reported. CT angiogram remains the gold standard to identify and confirm the diagnosis of ACAOS.

We present such a rare case of an adult male presenting with a history of syncope which on evaluation indicated second-degree AV block for which a permanent pacemaker was implanted. CT angiogram confirmed the diagnosis of ACAOS.

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Case Report

A 42-year-old man was brought to the emergency department with a history of syncopethat occurred 8 hours ago. This episode was sudden in onset and was associated with light-headedness, diaphoresis and followed by loss of consciousness for around 2-3 minutes.On evaluation, an electrocardiogram was suggestive of second-degree AV block (2:1) **[Fig. 1a]** and magnetic resonance imaging (MRI) brain was normal. The patient was hemodynamically stable with negative cardiac markers. His troponins remained in the normal range. The decision to implant a permanent pacemaker was made, Dual-Chamber Demand Pacing Device was implanted on the next day **[Fig. 1b]** -p/r wave was 2.0/9.4 mv, threshold (RA/RV) was 1.25/0.5 volts, impedance (RA/RV) was 570/710 Ω **[Fig. 2].**

After 1 week patient again had episodes of recurrent syncope followed by loss of consciousness. On admission, his cardiac markers turned out to be negative, electrocardiogram showed a right bundle branch block **[Fig 3].** On fluoroscopy there was no change in lead position and on further evaluation the pacemaker was working normally. The patient underwent coronary angiography via radial approach. Multiple attempts to engage the left system were unsuccessful followed by successful engagement of the right coronary artery (RCA). A careful review of the angiograms suggested an absent left system and an anomalous origin of the left coronary artery from the opposite ostia (ACAOS) **[Fig. 4].** The coronaries had no angiographically definable stenosis. A CT coronary angiogram was planned further to identify the course which confirmed the absence of the left coronary artery and anomalous origin of the left main coronary artery from opposite ostia **[Fig. 5].** The left anterior descending artery (LAD) has an intramural course **[Fig. 6a]** and the left circumflex (LCx) was coursing posterior to the aortic valve and anterior to the left atrium (retro-aortic course) **[Fig. 6b].**

Fig. 1: (a) ECG image on admission (b) post pacemaker implantation

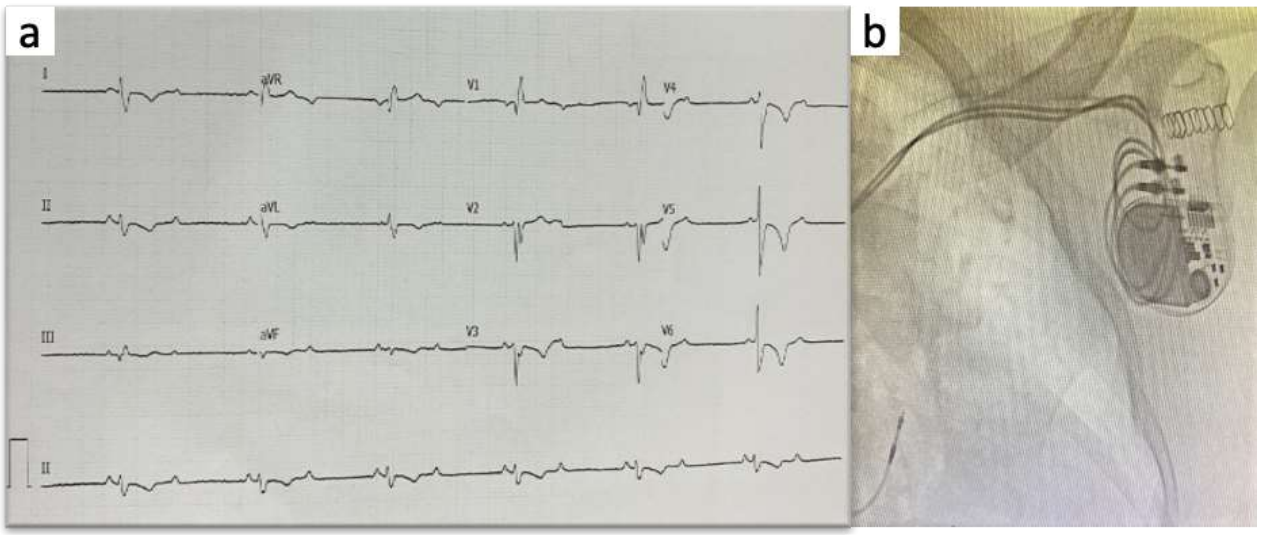

Fig. 2: ECG after pacemaker implantation

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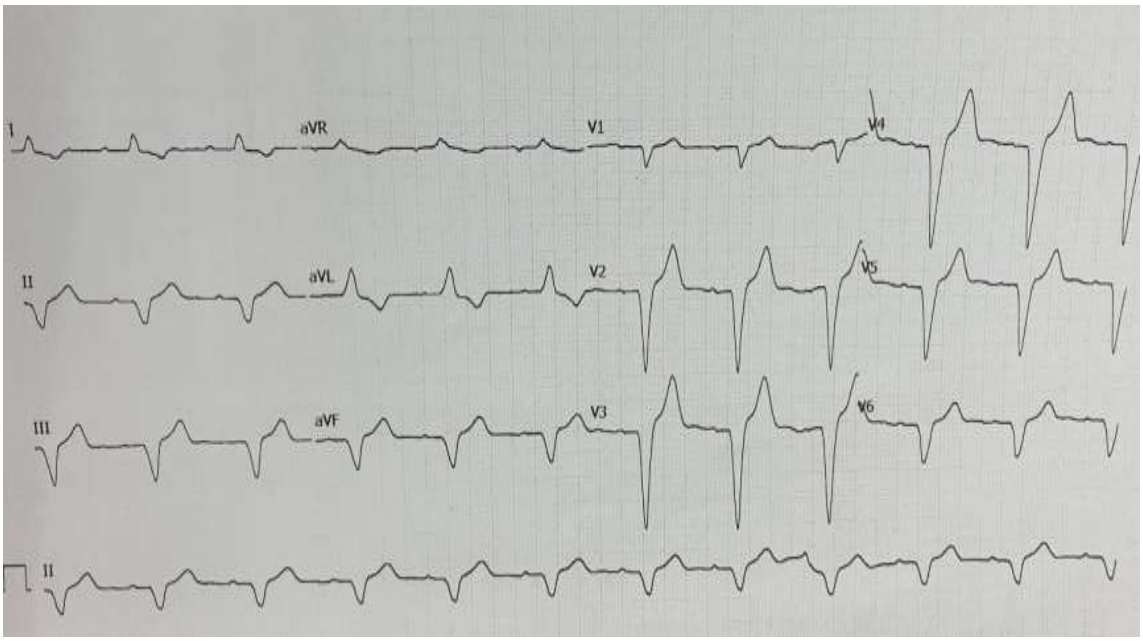

Fig. 3: ECG on the second admission

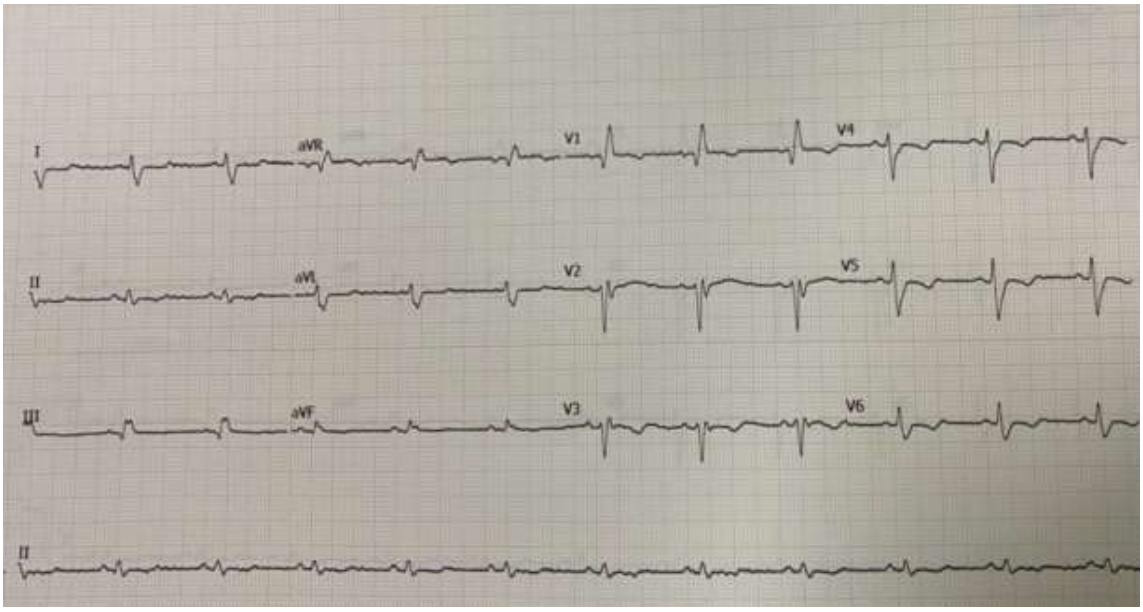

Fig. 4: a) Coronary angiogram shows the common origin of RCA, LAD, and LCx arising from right sinus in LAO cranial view b) LAO caudal view

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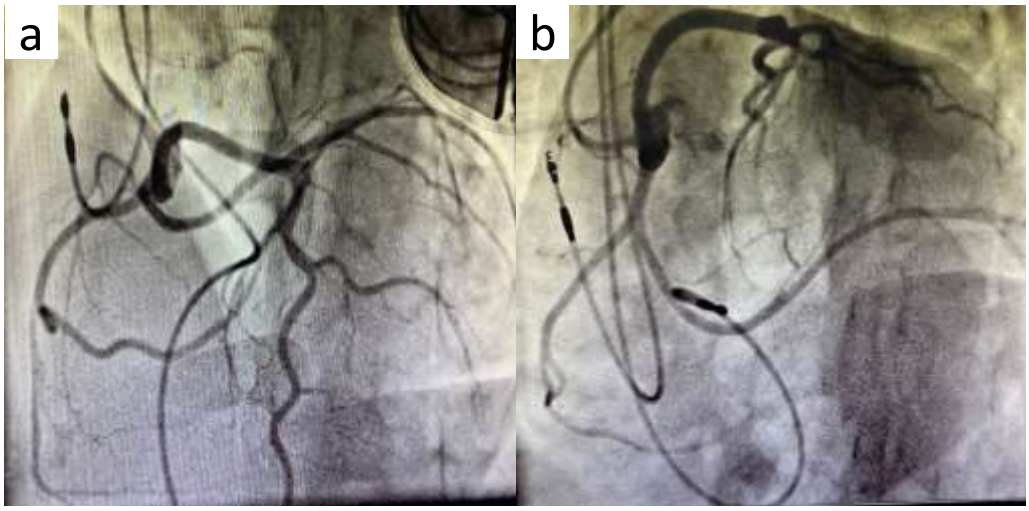

Fig. 5: CT coronary angiogram showing absence of the LCA and anomalous origin of LCA from opposite ostia

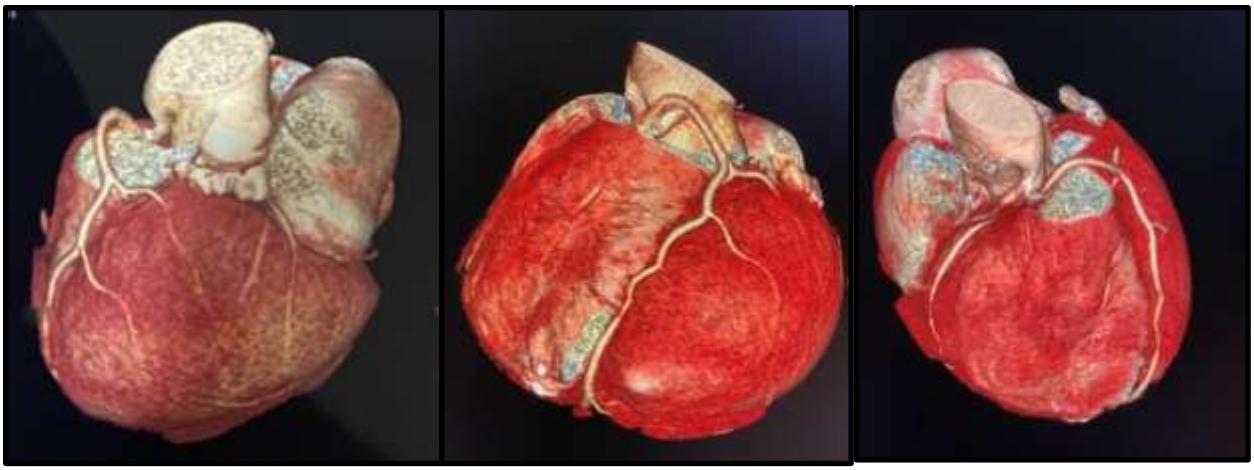

Fig. 6: a) LAD having intramural course b) LCX is coursing posterior to the aortic valve and anterior to the left atrium (retro-aortic course)

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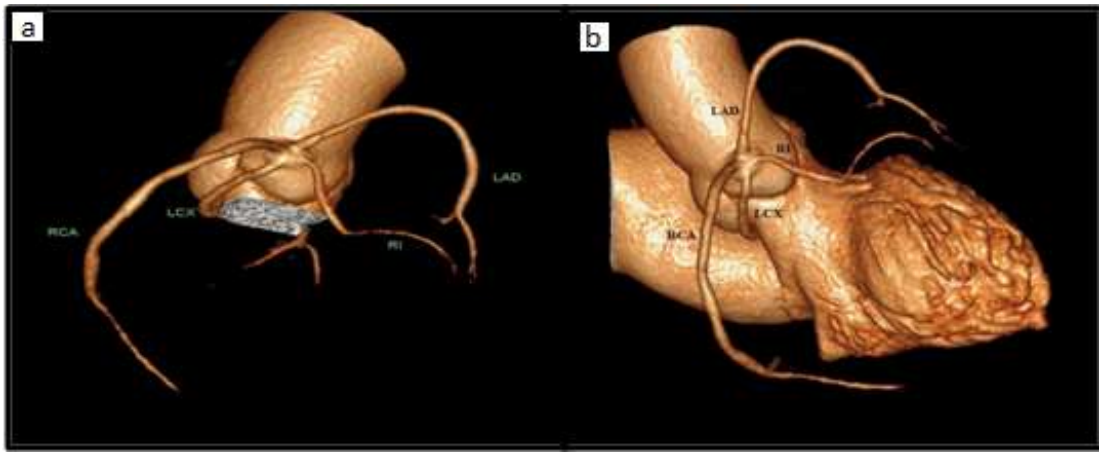

Discussion

Coronary anomalies remain the second most common cause of sudden cardiac death (SCD) after hypertrophic cardiomyopathy in children and young athletes with most diagnoses made at autopsy [2]. After AAOCA is diagnosed, whether through transesophageal echocardiogram (TEE), computed tomography angiogram (CTA), or coronary angiogram, the subsequent stage involves a more detailed examination of the vessel's anatomy. Non-invasive modern imaging techniques like multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI), enable a comprehensive examination of the origin and route of the anomalous vessels with a three-dimensional reconstruction. This information assists surgeons and interventional cardiologists in making informed decisions regarding the most suitable treatment approach.

In such anomalous origin of the coronary artery from opposite sinus, the vessel after arising from the opposite sinus takes one of the 4 paths described below. The precise path taken by the artery is clinically important.

a. Interarterial: In this case, a coronary artery (such as the RCA, LCA, LAD or LCX) arises from the opposite sinus and courses between the aortic root and pulmonary artery/right ventricular outflow tract. This anomaly has been linked with sudden cardiac death. Several pathologic processes have been implicated such as a narrow slit-like orifice, an acute angle of the ostium with a tangential proximal course of the ectopic coronary artery, and an intramural course where the coronary artery exits the aortic lumen and courses into the aortic wall before emerging on the surface.
b. Transseptal (subpulmonic): This describes the situation where the left main artery or LAD follows a path beneath the pulmonary artery. It travels in an anterior and downward direction through the interventricular septum and follows a course within the heart muscle, branching off septal arteries before reemerging at its usual position on the outer surface of the heart. It is considered a relatively benign anomaly, though in some cases of sudden and unexpected cardiac death it has been found to be the only reported abnormality.
c. Retro-aortic: This is the most common coronary artery anomaly, seen in 0.9% of the population. The ectopic coronary artery (more commonly the LCX) runs posteriorly between the aortic root and the left atrium.

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d. Pre-pulmonic: The ectopic coronary artery runs anterior to the pulmonary artery or right ventricular outflow tract.

Only 20% of patients have clinical manifestation of angina, dyspnea or syncope [3]. In young athletes, sudden cardiac death can be its common presentation. This could be due to occlusion caused by compression between the aorta and pulmonary artery during exertion [4,5]. ACAOS is accidentally detected during imaging to identify coronary artery disease. ACAOS might present with symptoms of CAD as they are prone for atherosclerosis which might be due to long and anamolous course of the vessel and compression between bigger vessels causing injury to the endothelium precipitating atherosclerotic changes [6].

Presence of ACAOS always doesn't require surgical correction. Adequate anatomic and hemodynamic evaluation with invasive and noninvasive imaging, guides in deciding whether ACAOS is responsible for the clinical presentation or increase of risk of cardiac death. Data from few studies have proposed that the length of the intramural segment and the extent of the elliptic proximal vessel shape (defined as height/width ratio of >1.3) is responsible for the increased risk for ischemia in these patients [7].

The development of myocardial ischemia in patients with ACAOS depends on the severity of dynamic and fixed components. Hence management is decided on the basis of this pathophysiology. After assessing whether the ACA is of benign or malignant origin, a decision can be made regarding the course of action. This decision may involve choosing between surgical intervention or a conservative approach, which encompasses medical therapy or percutaneous coronary intervention (PCI) in the event of obstructive coronary artery disease (CAD). Patients who present with ischemia-like symptoms from obstructive coronary disease and favorable anatomy of the anomalous LAD arising from a separate ostium with no interarterial or intramural course may be managed non-surgically with a PCI. Lipton et al. classified single coronary artery (SCA) depending on the site of origin and its anatomical distribution into right and left type **[Table 1]** with further subdivisions as mentioned in **Table 2**.

Table 1: Lipton's classification for SCA

Ostia location	**Anatomic distribution of subtypes**
Right sinus of valsalva	RI- SCA follows the course of normal RCA. RII-SCA from the right sinus gives off an anomalous transverse branch that crosses the base to reach the contralateral side. RIII-SCA from the right sinus, with LAD and LCx arising from separate coronary trunks instead of single trunk.
Left sinus of valsalva	LI-SCA follows the course of a normal LMCA.

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	LII-SCA from the left sinus gives off an anomalous transverse branch that crosses the base to reach to the contralateral side.
LAD, left anterior descending artery; LMCA, left main coronary artery; RCA, right coronary artery; SCA, single coronary artery.	

Table 2: Subtypes of type II single coronary artery based on the course of the aberrant vessel

Anatomic distribution	**SCA aubtypes based on the course of the aberrant vessel**
RII or LII	Type A-courses anterior to the pulmonary trunk Type B-courses between pulmonary artery and aorta Type P-courses posterior to the aorta Type S-septal type courses above the interventricular septum Type C-combined
SCA, single coronary artery.	

The above-presented case is a rare incidental finding of ACAOS originating from the right sinus of Valsalva with Lipton type RIII pattern. Since the patient had a pacemaker placed at the initial visit, it rescued the patient from developing any life-threatening events of sudden cardiac death.

Conclusion

We report a case of an incidentally identified SCA with Lipton type RIII pattern. Although the majority of the patients are asymptomatic further evaluation with coronary CTA is warranted to rule out potential malignant pathology.The risk of SCD is highest with the interarterial course of the aberrant vessel.The outward expansion of the aortic root and pulmonary trunk during exertion can lead to external compression of the left maincoronary artery which can result in acute myocardial infarction or sudden cardiac death.Stress testing is often not reliable in assessing the functional status of the patient. The optimal approach involves risk stratification, multidisciplinary management, and surgical intervention in appropriate patients.

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