THINKING OUT OF THE BOX IN CARDIAC STEAL SYNDROME:

LATE PRESENTATION OF A RARE CORONARY ARTERY FISTULOUS ANOMALY

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BACKGROUND

A coronary artery fistulous (CAF) anomaly is the abnormal communication between the coronary artery and one of the four chambers of the heart (coronary-cameral fistulae) / a great vessel (coronary artery arteriovenous malformations).

The approximate prevalence of CAFs is 0.002% of the general population and found in 0.2%-0.25% of individuals undergoing cardiac catheterization.

Most of the cases stem from the right coronary artery (50-60%), followed by the left anterior descending artery (25-42%).

These communications most often are due to deviations from normal embryological development or the persistence of sinusoids during the development of the heart.

CASE

A 51-year-old African male with a past medical history significant for hyperlipidemia and diabetes mellitus type 2 presented to the clinic with a chief complaint of dizziness for 15 mins. He had a sudden onset of dizziness, profuse sweating, fatigue, and shortness of breath. He has had similar shorter episodes since then.

On arrival, the first set of vitals were normal. A physical examination was unremarkable. An electrocardiogram (EKG) showed normal sinus rhythm with standard ST and T waves. An echocardiogram showed an EF of 65-70%, with ectatic coronary arteries. He underwent a nuclear stress test which was nonischemic.

CT angiogram during the same time visualized a congenital coronary artery to the left ventricle fistula. There was a left-to-right shunt. The right coronary artery was dilated from its origin and along its entire course up to approximately 8 mm. It courses in the right atrioventricular groove and extends inferiorly and posteriorly around the heart in the distribution of the posterior lateral branch. At the level of the inferior aspect of the left atrium, the dilated vessel tapers where it enters the posterior portion of the left ventricular myocardium. The vessel measures approximately 2 mm as it traverses the left ventricular myocardium and drains into the left ventricle. This finding of a rare coronary artery anomaly may account for the patient's symptoms.

DECISION-MAKING

Our patient was referred to a cardiothoracic surgeon for a symptomatic coronary artery anomaly. However, the patient decided not to pursue further management.

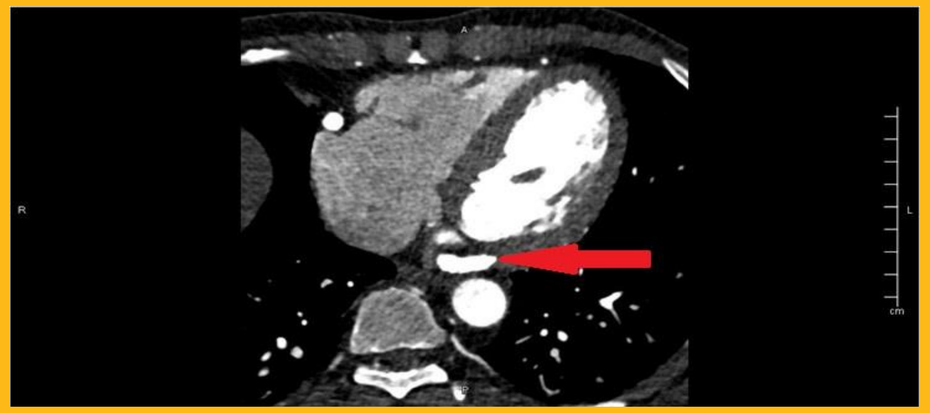

Figure 1: **CORONARY ARTERY FISTULOUS ANOMALY**

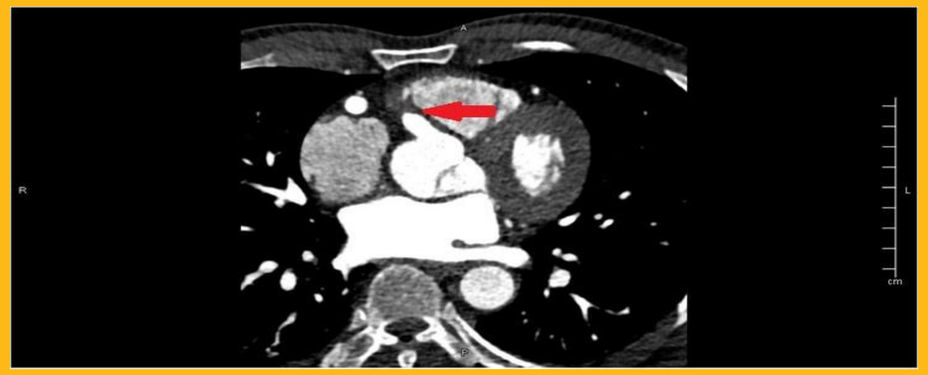

Figure 2: **ORIGIN OF DIALTED RIGHT CORONARY ARTERY**

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LEARNING POINTS

- Most cases of CAF are asymptomatic; however, they can also manifest as an acute coronary syndrome (ACS) or chronic myocardial ischemia, arrhythmias, sudden cardiac arrest, heart failure, cardiomyopathy, pulmonary hypertension, endocarditis, thrombosis, and rupture of the fistula.
- The resultant physiologic derangement is determined by the site of origin and termination of the abnormal connection, and the size of the connection.
- Coronary angiography is the gold standard for diagnosis.
- Transesophageal echocardiography is sensitive for detecting the entrance and termination site of the shunt, characterized by a continuous turbulent systolic and diastolic flow pattern.
- Its management depends on site of origin of the fistula, size of the fistula, patient's symptoms, presence of any complication caused by the fistula, age of the patient, the anatomy of the fistula, and co-existence of other indications to undergo an invasive procedure.
- Definitive treatment options are surgery or catheter closure.
- Appropriate informed consent/counseling of the affected individual with the known risks is necessary. Vigilance for cardiac events is recommended. Individuals with untreated abnormalities should be restrained from competitive sports.

CONCLUSION

The fistulous connection results in blood bypassing the myocardial capillary network hence a coronary steal phenomenon.

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