Research Letter

Anomalous origin of RCA from left coronary sinus presenting as PSVT and recurrent acute coronary syndromes

Keywords:
Anomalous origin of RCA
Supraventricular tachycardia
Acute coronary syndrome
MDCT angiography
Coronary sinus

Abstract

Anomalous origin of the right coronary artery from the left sinus of Valsalva and coursing between the aorta and pulmonary artery is a rare congenital abnormality representing less than 3% of the congenital coronary anomalies. Patients can present with myocardial ischemia, arrhythmias, or sudden cardiac death (SCD). This diagnosis should be suspected in young patients without risk factors for coronary artery disease and the diagnosis can be confirmed easily by 64-slice MDCT angiography. Surgery is generally recommended in symptomatic patients whereas conservative management in asymptomatic patients. Incidence of SCD is rare after the age of 35 years and hence conservative treatment may be advised in asymptomatic patients over the age of 35 years.

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Anomalous origin of the right coronary artery (ARCA) from the left sinus of Valsalva and coursing between the aorta and pulmonary artery is a rare congenital abnormality with prevalence varying from 0.025% to 0.25%. They may be associated with sudden cardiac death (SCD) and it may present with ischemia or arrhythmias or syncope.

A 42-year-old woman presented with angina at rest since 2 days. She had no conventional risk factors for coronary artery disease. She had past history of unstable angina 2 years back when coronary angiography was done and was reported to be apparently normal. She also gave past history of PSVT, which responded to IV adenosine (Fig. 1). Her ECG showed dynamic ST-segment changes and T-wave inversion in leads II, III, aVF, and V2-6 (Fig. 2) at both admissions for unstable angina and cardiac enzyme showed transient rise and fall. Echocardiography was essentially normal.

Her symptoms subsided after the institution of medical treatment and ST-segment and T-wave changes normalized. She was further evaluated by MDCT coronary angiography, which showed anomalous origin of the RCA from left coronary sinus, which was coursing between aorta and pulmonary artery to take its usual distal course (Fig. 3A, B). The RCA showed a slit-like opening and abnormal acute takeoff at the origin. In view of her recurrent symptoms as manifested by ischemia and arrhythmia, she was advised surgical correction of anomalous RCA, but the patient refused surgery. She was started on beta-blocker after which she does not have any recurrence of symptoms on follow-up for 2 years.

Anomalous origin of RCA from the left sinus is a rare congenital anomaly representing less than 3% of the congenital coronary artery anomalies. Anomalous origin of RCA is more common than left coronary artery (LCA) but the incidence of SCD is less compared to anomalous LCA. Majority of the patients are asymptomatic, but they can present with ischemic symptoms or arrhythmias or even SCD.

Pathophysiology of the symptoms are suggested to be due to compression of proximal RCA between aorta and pulmonary artery, slit-like orifice of the origin of RCA, abnormal takeoff of the RCA origin, or propensity of spasm of the proximal segment. Slit-like orifice and acute angle takeoff are seen more commonly in some of the autopsy studies. MDCT is currently the favored imaging modality to diagnose anomalous
right coronary artery (ARCA) due to its higher spatial resolution. As evaluation is often difficult in coronary angiography, the slit-like orifice and abnormal angulation make engagement of ostium difficult and the three-dimensional structure is displayed in two dimensions.

The treatment of ARCA is often controversial. Surgery is recommended in symptomatic patients and conservative management in asymptomatic patients. Reimplantation of the RCA to aorta, unroofing of the ostium, osteoplasty, and bypass graft to distal RCA with proximal ligation are the surgical options. Young patients with symptoms should undergo surgery and older patients without symptoms do not need surgery.

Taylor et al.3 prospectively followed up 52 patients and 25% had SCD during 5-year follow-up. Only variable that was predictive of lower risk of SCD was age more than 30 years. None of the patients above the age of 30 or below the age of 10 years had SCD. Kaku et al.4 studied 56 patients prospectively and followed them on beta-blockers. None of the patients had SCD during the 5-year follow-up period. So patients more than 35 years without symptoms are justified in medical follow-up on beta-blockers and surgery may not be recommended.

We believe that our patient has these attacks of arrhythmias and myocardial ischemia secondary to restriction of blood flow as a result of compression of the coronary artery between the aortic root and the pulmonary artery. CT angiography showed no atherosclerosis; hence, the reason behind her symptoms may be secondary to the RCA anomaly.

To conclude, anomalous origin of RCA from left sinus is a rare congenital coronary anomaly, which can present with ischemia, arrhythmias, syncope, or SCD, especially during physical or emotional stress. MDCT scan is an excellent investigative modality for diagnosis. Surgery is recommended

Fig. 1 – ECG showing narrow QRS regular tachycardia suggestive of AVNRT.

Fig. 2 – ECG during the admission for acute coronary syndrome, showing ST-segment deviation and T-wave changes in inferolateral leads.
in symptomatic patients and asymptomatic young patients. Incidence of SCD is rare after the age of 35 years and hence conservative treatment may be advised in asymptomatic patients over the age of 35 years.

Conflicts of interest

The authors have none to declare.

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