Superdominant Right Coronary Artery with Absent Left Coronary Artery and Left Circumflex Artery with Anomalous Left Anterior Descending Artery

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Introduction

The occurrence of super-dominant “single coronary artery” is an extremely rare and seldom reported phenomenon. The heart is dependent on a single vessel which makes its occlusion, if present, catastrophic. Here, the authors present an extremely rare combination of superdominant right coronary artery coexisting with absent left coronary artery and left circumflex artery with abnormal origin of left anterior descending artery from right coronary sinus. Precise morphological and physiological knowledge and evaluation of these anomalies is a must for opting the best available treatment.

Case History

We report a 50-year-old nondiabetic non-hypertensive female who presented with complaints of palpitations and episodic dyspnea for 1 year. Her vitals were stable. ECG showed Qs waves in lead II, III, and aVF and ST elevation in lead III and aVF (inferior wall ischemia). Cardiac biomarkers were normal. 2D-echocardiography showed regional wall motion abnormality and ejection fraction: 57%. Routine blood tests were normal. Computed tomography (CT) and invasive coronary angiography were done subsequently with suspicion of CAD. CT coronary angiography showed the prominent and dilated right coronary artery (RCA) originating from the right coronary sinus and giving off a prominent marginal artery (►Fig. 1). After coursing through posterior right atrioventricular (AV) groove and posterior cardiac crux, it was entering posterior left AV groove, running parallel to coronary sinus (the usual course of LCX) (►Fig. 2), then continuing in anterior left AV groove ending at anterior cardiac crux. LCA and LCX were absent (►Fig. 3). LAD originated from the right coronary sinus with separate ostium. It appeared attenuated and revealed focal kink followed by a tiny saccular aneurysm in front of the right ventricular outflow tract (RVOT) and thereafter coursing...
along the anterior surface of the right ventricle (demonstrating the anterior free wall course) (►Figs. 1 and 4). Invasive coronary angiogram showed similar findings (►Fig. 5).

Discussion

Coronary artery anomaly (CAA) is defined as any morphological feature of coronary arterial system with prevalence of <1%. The incidence of CAAs varies from 0.3 to 5.6%.

Single coronary artery (SCA) is one such rare anomaly with an incidence of 0.05%. In SCA, only one coronary artery arises from aorta by a single ostium, supplying the whole heart. A detailed classification was proposed by Lipton et al, later modified by Yamanaka and Hobbs. According to this, Group I coronary arteries are super-dominant which follows the course of either RCA or LCA.

Superdominant RCA (R-I variant of Lipton classification) is the seldom reported anomaly. It is almost always associated with the congenital absence of LCX as a part of anatomical compensation to meet the demands of usual territory of LCX.

Fig. 1 3D volume reconstruction image showing superdominant RCA arising from right coronary sinus (yellow arrow). Note the tortuosity of vessel. LAD is also arising from the right coronary sinus (black arrow). 3D, three-dimensional; LAD, left descending artery; RCA, right coronary artery.

Fig. 2 3D volume reconstruction image showing superdominant RCA (yellow arrow) running in posterior right and left AV groove, entering LCX territory. Note prominent PDA branch (black arrow). 3D, three-dimensional; LCX, left circumflex artery; PDA, posterior descending artery.

Fig. 3 3D volume reconstruction image showing superdominant RCA in LCX territory (red arrow). Note absent LCA from left coronary sinus (blue arrow). 3D, three-dimensional; LCX, left circumflex artery; RCA, right coronary artery.

Fig. 4 Oblique sagittal reconstruction MIP image of CT coronary angiography showing origin of superdominant RCA (blue arrow) and anomalous LAD (orange arrow) from the right coronary sinus. LAD, left descending artery; MIP, maximum intensity projection; RCA, right coronary artery.
such that approximately 90.4% of patients with congenitally absent LCX have super-dominant RCA. The congenitally absent LCX itself has reported incidence of 0.067%. The origin of LAD from the right coronary sinus is another rare anomaly. It is usually seen associated with various congenital heart diseases like tetralogy of Fallot or double outlet left ventricle as LAD is influenced by pulmonary conus development. The presence of this anomaly in normal heart is rarely reported with incidence of 0.03%. The LAD after its origin may follow any of following pathways: prepulmonic (anterior to RVOT)—seen in our case, retroaortic, interarterial, transeptal/subpulmonic, and retrocardiac. The explanation of clinical symptoms in these anomalies is largely unknown. Various causes are attributed—spasm of coronaries due to endothelial injury, myocardial squeezing, acute angles of take-off, slit-like orifices, and vessel hypoplasia. Other explanation is “steal phenomenon,” where an increased blood supply to a territory causes transient ischemia in other territories leading to symptoms. In our case, ECG suggested inferior wall ischemia, which may be caused by insufficiency of super-dominant RCA to adequately supply the LCX territory. Literature suggests no definite correlation between CAA and atherosclerosis. In our case, no atherosclerotic change was noted.

According to a literature review by Fugar et al, super dominant RCA with absent LCA and LCX is reported with other concomitant anomalies like atretic mid LAD originating from the sinus of Valsalva, dual LAD, and non-existing left subclavian artery. However, there are very few reported cases of superdominant RCA with absent LCA and LCX along with LAD arising from the right coronary sinus. In one case, LAD was arising from the same ostium as superdominant RCA; while in other cases, LAD originated from different ostia (here, LAD had septal course rather than anterior free wall course). To our knowledge and best of literature search, our “combination of superdominant RCA with absent LCA and LCX with LAD originating from right coronary sinus with different ostia and having anterior free wall course” has been reported only once before by Enezate et al in 2017. Radiologists and Cardiologists should be acquainted with these anomalies as occlusion of a single coronary vessel may have deadly consequences. Knowledge of these anomalies would help in accurate revascularization in the presence of coronary artery disease. Precise anatomical and physiological evaluation of these anomalies is a must for opting for the best available therapeutic modality and better prognosis.

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Conflict of Interest
None declared.

References