Congenitally Absent Left Circumflex Artery

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Abstract
Congenital coronary anomalies are uncommon with rarest being absent left circumflex artery (LCX) having prevalence of 0.003%. We report a case of a 68 year old male having acute coronary syndrome and left ventricular dysfunction whose coronary angiogram showed an absent LCX with super dominant right coronary artery (RCA). Precise morphological evaluation is needed for best suited management strategy.

Introduction
Congenital coronary anomalies have an uncommon occurrence with the incidence of approximately 0.1–2%. Majority of coronary anomalies (80%) are asymptomatic and clinically silent, which are detected incidentally, while approximately 20% of them have been reported to have a clinical presentation. One of the rarest vascular anomaly is congenitally absent left circumflex artery (LCX) with a prevalence of 0.003% where LCX artery is not seen to develop in the left atrioventricular groove. While commonly being detected incidentally on various imaging studies, congenital absence of LCX artery has been seen to be associated with myocardial infarction, vasospastic angina and systolic click syndrome.

We hereby describe a case of 68 year old male who presented with recent worsening of breathlessness with a dilated left ventricle and poor left ventricular ejection fraction on echocardiography. Coronary angiography showed a long segment of disease involving the left anterior descending (LAD) artery with absent LCX artery and a large super dominant right coronary artery (RCA). Aortic root injection did not show any anomalous origin of LCX artery. MDCT angiography further confirmed the absence of LCX artery. Acute coronary syndrome, left ventricular dysfunction and coronary artery disease in association with congenital absence of LCX artery is rarely described.

Case Report
A 68 year old gentleman with diabetes presented with history of breathlessness for 2 months which worsened over one week before presentation. He had no history of palpitations or chest pain in the past. Physical examination revealed an average built male with a pulse rate of 110 beats per minute and a blood pressure of 160/90 mm Hg. Peripheral pulses were palpable and normal. Routine investigations were within normal limits. Systemic examination revealed bilateral basal crepitations while on cardiac auscultation S3 was audible in addition to the normal heart sounds. Chest radiograph showed borderline cardiomegaly with mild pulmonary oedema. A 12 lead electrocardiogram showed normal sinus rhythm with T wave inversions in anterior leads suggestive of anterior wall ischemia. Echocardiography was suggestive of dilated left ventricle (LVIDD = 6.0 cm; LVIDS = 5.2 cm) with mild concentric hypertrophy of the left ventricle. There was global hypokinesia with a left ventricular ejection fraction of 35%. He was stabilised with cardioprotective medication and subsequently underwent a coronary angiography.

Coronary angiography was performed via the femoral approach. On left coronary injection, a long segment of disease involving the LAD was seen with near about 70% stenosis in the mid LAD segment and 90% stenosis in the distal LAD was seen. LCX was not visualised (Figure 1). RCA was a large super dominant ectatic vessel (Figure 2). Prominent posterolateral and posterior descending artery were seen supplying the posterolateral wall of left ventricle. Aortic root injection further confirmed the absence of any anomalous origin of left circumflex artery.

On the following day, 128 slice MDCT angiography was performed which showed the absence of LCX. No anomalous LCX artery was visualised. LAD showed a long segment of stenosis while a super dominant RCA was seen with no discrete stenosis (Figures 3 and 4).

Discussion
Though varying from mild to life threatening presentation, anomalous coronary arteries are a rare occurrence. Most commonly seen coronary anomaly
is LAD and LCX arising separately from left sinus of valsalva. Failure of development of LCX in the left atrioventricular groove leading to an absent LCX is an extremely rare anomaly with a reported incidence of 0.003%.³

There are many schools of thoughts for the congenital absence of LCX artery. Some believe that this condition is not a real congenital anomaly, and that it is the anomalous origin of the LCX artery from distal RCA.⁴ Recently the new acronym, congenital ostial stenosis or atresia (COSA) has been coined. Most of the times, this anomaly is a benign condition. However, it can cause chest pain, particularly on exertion. Our patient also complained of occasional chest pain, but his main complaints were that of breathlessness.

Ueyama et al. have previously reported an anomalous origin of LCX where the vessel was seen arising from the right sinus of valsalva or from the RCA.⁵ When following a normal origin and course, LCX runs around the atrioventricular groove along with RCA and forms a circle. RCA thereby compensates for the absence of LCX by perfusing its territories. This compensatory mechanism leads to a benign outcome in majority of cases.

Mievis et al. have reported myocardial infarction in a 31 year old male where on coronary angiography no significant stenosis could be seen involving the coronaries, but it showed an absent LCX.⁶ As seen by Mohsen et al.⁷ in our case too, a super dominant RCA perfused a large area extending to the left ventricle.

Though conventional angiography is well known for its ability to detect coronary anomalies but MDCT being a non invasive modality with a three dimensional imaging and reconstruction is well suited for defining coronary arterial anatomy and their course. At times, there may be difficulty in differentiating between congenital absence of an artery or an ostial stenosis. A detailed angiographic evaluation is must to understand the course of each coronary artery and its relationship with various clinical presentations so that the best treatment modality can be selected.

References

