

Congenital ostial atresia of right coronary artery: an exceedingly rare anomaly diagnosed on CT angiography

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DESCRIPTION

A 28-year-old woman, with exertional dyspnoea, underwent transthoracic echocardiography, which revealed a small atrial septal defect. The patient subsequently underwent a cardiac CT angiography (CTA) to look for any other associated cardiac and extracardiac defects. Review of CTA images incidentally detected ostial atresia (yellow arrow) of right coronary artery (RCA) (figure 1). RCA was reformed via a tortuous collateral (*) arising from the proximal left anterior descending (LAD) artery and crossing anterior to the right ventricular outflow tract to reach the proximal right atrioventricular groove, suggestive of 'Vieussens' arterial ring (figure 2A,B).

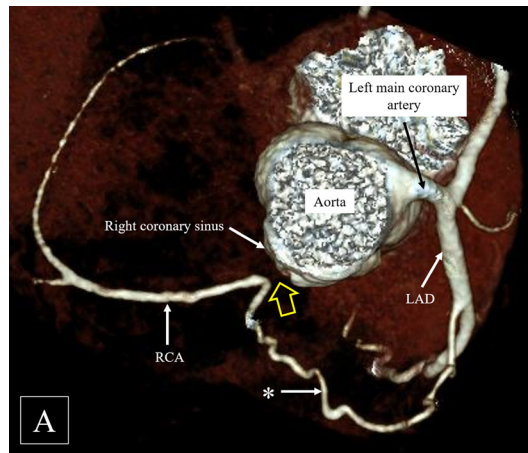


Figure 1 Volume-rendered CT angiography image reveals ostial atresia (yellow arrow) of the right coronary artery (RCA). RCA distal to the atretic segment is reformed by a tortuous vessel (*) arising from the left anterior descending (LAD) artery.

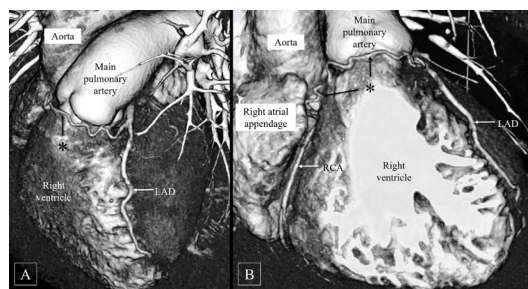


Figure 2 Volume-rendered CT angiography images (A and B) depict the course of the tortuous collateral, traversing anterior to the right ventricular outflow tract and reaching the right atrioventricular groove.

Coronary ostial atresia, especially involving the RCA, is an extremely rare congenital coronary anomaly. Though the defect is congenital; at times, it may not be symptomatic owing to a well-developed collateral circulation from the opposite normal coronary artery. Absence of an ostial stump and presence of only one or two full-calibre connecting collaterals (without evidence of any narrowing at the transition between the supplying and the receiving vessel) is suggestive of its congenital origin, in contrast to the rich network of collateral vessels seen in cases of acquired RCA occlusions.¹ Also in acquired occlusions, CTA can visualise the wall of the occluded segment and thus help rule out coronary ostial atresia.

The major collateral pathways at the conotruncal level providing circulation between the right and left coronary systems include the preconal Vieussens' ring, retroconal anastomotic ring in the interarterial space or the retroaortic Kugel's ring.² 'Vieussens' arterial ring, thought to be a remnant of the embryonic conotruncal circle that gives rise to the coronary ostia, is a crucial intercoronary collateral connection between the conal branches of RCA and LAD artery, providing flow to either arteries in presence of congenital atresia or event of acquired stenosis/occlusion. A surgical bypass procedure is rarely required to treat congenital right coronary ostial atresia as the collateral supply is usually adequate.

Learning points

- ▶ Coronary ostial atresia, especially involving the right coronary artery (RCA), is an extremely rare congenital coronary anomaly.
- ▶ The anomaly may be asymptomatic owing to a well-developed collateral circulation from the opposite normal coronary artery, with the major collateral pathways at the conotruncal level being the preconal Vieussens' ring, retroconal anastomotic ring in the interarterial space or the retroaortic Kugel's ring.
- ▶ 'Vieussens' arterial ring is a crucial intercoronary collateral connection between the conal branches of RCA and LAD artery, providing flow to either arteries in presence of congenital atresia or event of acquired stenosis/occlusion.

Contributors NNP, MS and SK have participated sufficiently in the conception of the idea, development of the intellectual content, design, writing and final approval of the manuscript.



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