Case Report

Right Coronary Artery Arising from the Left Main Stem with Dynamic Compression by Great Vessels

Anomalous origin of the right coronary artery from the left main stem is a very rare congenital anomaly. The entrapment of its course between the aorta and pulmonary trunk can cause dynamic blood flow limitation. We report a 68-year-old female presenting with recurrent chest pain and a positive treadmill exercise test. The coronary angiography disclosed anomalous origin of the right coronary artery from the left main stem with dynamic compression between aortic root and pulmonary trunk during the systolic phase. With hesitation about surgical revascularization, the patient was maintained on oral medication uneventfully.

Key Words
- coronary angiography
- coronary artery anomaly
- right coronary artery
- single coronary artery

Sin gle coronary artery is a rare congenital anomaly, and the reported angiographic incidence was 0.044%. Single coronary artery with anomalous origin of the right coronary artery (RCA) from the left main (LM) stem is even rarer and was once thought to be a benign condition. As the anomalous RCA originates from the LM stem, it can pass anterior to the pulmonary trunk, posterior to the aortic root, or between these two great vessels. Nevertheless, as the proximal RCA courses between the aorta and pulmonary trunk, the adjacent great vessels can pinch it, resulting in dynamic blood flow compromise. Severe clinical events including angina pectoris, acute myocardial infarction, syncope, and sudden death have been reported in association with this anomaly. With growing awareness of its pathophysiology, the optimal approach has been switched from conservative medical treatment to aggressive surgical intervention. Herein, we report a 68-year-old female with anomalous origin of the RCA from the LM. Coronary angiography showed the coronary blood flow was dynamically compromised by aortic root and pulmonary trunk in the systolic phase.

CASE REPORT

A 68-year-old female was admitted to our institute due to recurrent episodes of chest pain and a positive
tread mill exercise test. Her risk factors for atherosclerosis included only age and hypertension.

Coronary angiogram revealed a single coronary artery originating from the left sinus of Valsalva (Fig. 1). Moreover, the RCA arose anomalously from the LM coronary artery and passed between the aortic root and pulmonary trunk. Cineangiogram disclosed that the two great vessels dynamically compressed this anomalous RCA during the systolic phase of the cardiac cycle (Fig. 2). The whole coronary tree was free of atherosclerotic lesions.

Due to overt clinical symptoms and compromised RCA blood flow, surgical revascularization was suggested. Nevertheless, the patient hesitated to take this proposal and was medically treated with aspirin, nitrates and calcium channel blocker. Her symptoms have been significantly improved with medical therapy. She was well at last follow-up, 10 months after coronary angiography.

DISCUSSION

Single coronary artery is a rare congenital anomaly.

Among the reported 56 patients with a single coronary ostium, the single coronary arteries arose from the right sinus of Valsalva in 25 cases and the left sinus of Valsalva in 31 cases. As a part of left single coronary artery, the RCA can traverse either anterior to the pulmonary trunk, posterior to the aortic root, or between the
pulmonary trunk and the aorta. For many years, angiographers and pathologists regarded the latter anomaly as a minor variant of no clinical importance.\textsuperscript{1-3} Recently, grave clinical manifestations including angina pectoris, acute myocardial infarction, syncope and sudden death have been described in patients with this anomaly.\textsuperscript{4,5} Therefore, isolated single coronary artery is not always benign and is potentially lethal.

The anatomic and clinical significance of anomalous RCA emerging from the left sinus of Valsalva appears to be related to its course after its origin. Kragel et al.\textsuperscript{8} proposed a practical classification for the various ways in which the aberrant RCA originated from the left sinus of Valsalva and subsequently coursed between the aortic root and the pulmonary artery. In their series of 25 cases with this anomaly, eight cases were fatal. According to their classification, the 2D-type anomaly represents the anomalous RCA originating from the LM and passing between the aorta and the pulmonary artery. In their series of 25 cases with this anomaly, eight cases were fatal. A corresponding to their classification, the 2D-type anomaly represents the anomalous RCA originating from the LM and passing between the aorta and pulmonary artery. Our patient can be classified as a variation of this 2D-type anomaly. In a similar series of 10 necropsy cases with the anomalous RCA from the left sinus of Valsalva and between the aorta and pulmonary artery en route, three had sudden deaths in the absence of coronary atherosclerosis.\textsuperscript{5} Thus, passage of the anomalous RCA between the aorta and pulmonary trunk, either from a separated ostium in the left sinus of Valsalva or from the proximal portion of the LM, has the potential to cause devastating clinical events.

Roberts and his colleagues proposed 2 mechanisms by which the anomalous RCA from the left sinus of Valsalva might cause myocardial ischemia.\textsuperscript{9} First, the anomalous RCA may pass between the aorta and the pulmonary artery and might be dynamically compressed by the 2 great vessels (Fig. 3). Secondly, an upright slit-like orifice of the anomalous RCA from the left coronary sinus may result in decreased ostial blood flow during exercise. In our patient, cineangiogram demonstrated a dynamic change of the RCA blood flow due to great vessel compression during the cardiac cycle. This angiographic finding is consistent with the first mechanism.

Optimal approach of treatment for the anomalous RCA that arises from the left sinus of Valsalva remains controversial.\textsuperscript{10,11} Emerging evidence that the anomaly is potentially lethal has led to aggressive surgical intervention instead of conservative management.\textsuperscript{6,7} The proposed methods of surgical correction in the literature included ostial reconstruction, re-implantation of the RCA into the correct sinus and coronary artery by bypass grafting.\textsuperscript{11,12} Although Rinaldi et al.,\textsuperscript{11} believed that ostial reconstruction did restore the orifice size and correct the

Fig. 3. (A) The anomalous right coronary artery (RCA) originates from the left main (LM) stem and courses between the aorta and pulmonary trunk. (B) During systolic phase, compression of the RCA by distended aorta and pulmonary trunk results in compromised blood flow and subsequent cardiac dysfunction. L = left sinus of Valsalva; R = right sinus of Valsalva; P = posterior sinus of Valsalva; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery.
right angle at takeoff, the im pair ment in blood flow by great vessels compression could not be corrected. The outcome of re-implantation of the RCA into the cor rect sinus is still unde fined. The result of min i mally in va sive di rect cor o nary ar tery by pass (MIDCAB) us ing in ter nal mam mary ar tery as ar te rial con duit has been en cour ag ing.\textsuperscript{13,14} In com par i son with con ven tional cor o nary ar tery by pass graft ing, MIDCAB does not require median sternotomy or the use of cardiopulmonary by pass. These ad va nta ges lessen the pa tients’ suf fer ing and shorten the du ra tion of hos pi tal iza tion.\textsuperscript{14} MIDCAB was re com mended to our pa tient based on the above up dated re view. How ever, the long-term re sult of MIDCAB in this sit u a tion re mains to be de fined because there are still only lim ited case re ports.

In con clu sion, anom a lous or i gin of RCA from the LM stem with sub se quent cours ing be tween the pul mo nary trunk and aor tic root is a very rare con gen i tal anom aly, and its clinical sig nif i cance is po ten tially le thal. Once the clin i cal symp toms are ap par ent and myo car dial is che mia has been doc u mented, ag gres sive sur gi cal in ter ven tion is re com mended. MIDCAB is the pre fer red method for in ter ven tion.

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