Letter to the Editor

Adult-onset right main coronary artery atresia: a case description and literature analysis

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Introduction

Coronary artery atresia (CAA) is a rare congenital anomaly of the coronary arteries, of which there have been only approximately 100 reported cases to date (1). CAA predominantly affects the left main coronary artery (LMCA), and involvement of the right coronary artery (RCA) is extremely rare. To our knowledge, fewer than 5 reported cases of right main coronary artery atresia (RMCAA) have been identified by either digital subtraction angiography (DSA) or coronary computed tomography angiography (CCTA).

Case presentation

A 58-year-old woman presented with complaints of recurrent discomfort and pain in the precordial area for over 1 month. The pain was intermittent and lasted for several days during each episode. The patient had a history of hyperthyroidism for over 10 years, which was managed with medication. She had independently discontinued her medication 1 year ago, and thyroid function test results showed no abnormalities. CCTA was performed, and it revealed that the left coronary sinus gave rise to the LMCA, which subsequently branched into the left anterior descending (LAD) artery and the left circumflex (LCx) artery. The LAD artery traversed the anterior interventricular groove, whereas the LCx artery travelled along the left atrioventricular groove, crossing the posterior interventricular groove along the right atrioventricular groove and retrogradely ascending along the right atrioventricular groove (*Figure 1A-1C*). The RCA was not clearly visualized; a slender blood vessel of about 2 cm length, that appeared as a thread-like shadow and originated from the right coronary sinus was observed, indicating distal occlusion. There was no communication between the distal LCx transition and this thread-like shadow (*Figure 2*). Moreover, there was no significant plaque formation in the coronary arteries. The imaging diagnosis was 'RMCAA'. With the clinical consideration of angina, oral metoprolol tartrate tablets (dose of 25 mg) were prescribed twice daily for 3 weeks. The symptoms disappeared at the end of the treatment course. A follow-up phone call 6 months later revealed that the patient was not experiencing significant discomfort.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this article and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

In embryology, CAA is caused by the regression of the left/right main trunk during development, resulting in the absence of the proximal main coronary artery or the presence of a blind-ended thread-like structure, with blood supply coming from 1 or more collateral arteries from the opposite side (2,3). CAA can be classified into 2 categories according to the age at the first onset, namely infant/childhood-onset CAA (symptoms appear early in life) and

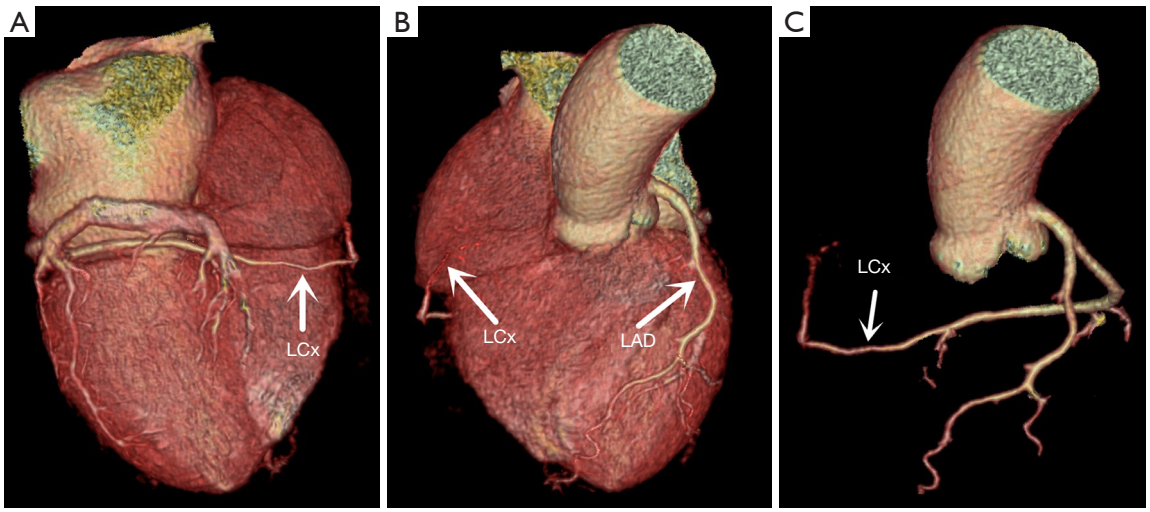

Figure 1 CCTA in a 58-year-old female with RMCAA. (A-C) The LCx artery crosses the atrioventricular groove, retrogradely ascending along the right atrioventricular groove (white arrow), and the right coronary sinus does not show the emergence of the RCA. LCx, left circumflex; LAD, left anterior descending; CCTA, coronary computed tomography angiography; RMCAA, right main coronary artery atresia; RCA, right coronary artery.

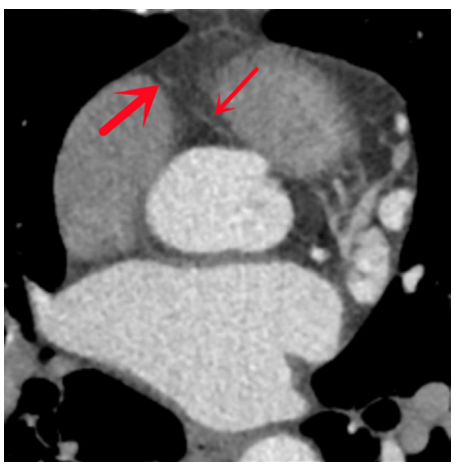

Figure 2 The right coronary sinus gives rise to a slender thread-like structure (thin arrow), which is occluded at the distal end and does not communicate with the distal part of the LCx artery (thick arrow). LCx, left circumflex.

adult-onset CAA (asymptomatic or symptoms appear only in old age) (1). In infants, the most common symptom is congestive heart failure, whereas children over 2 years of age mainly present with fainting and chest pain. In over half of adult patients, the predominant symptom is angina pectoris (1), and most symptoms result from myocardial ischemia or inadequate compensation by collateral circulation. A small percentage of pediatric patients may have concomitant cardiac abnormalities, such as mitral valve prolapse and aortic valve stenosis.

Tomanek *et al.* defined an incomplete connection between the main trunk and the coronary sinus as 'atresia', which was observed as a 'dead end' at the distal end of the main trunk during angiography (4). LMCA atresia is primarily characterized by the absence or thread-like appearance of the left main coronary opening, with the normal connection of the LAD and LCx arteries, and blood supply to the RCA is established through collateral circulation from the LAD and/or LCx (3). RMCAA is extremely rare, with only 4 reported cases identified in the literature (5-8) (*Table 1*). Of them, 2 cases presented as complete occlusion of the main trunk of RCA (with 1 case being slender blind ended), the LCx artery retrogradely ascended along the right atrioventricular groove, supplying the right atrium and right ventricle (5,6). The other 2 cases presented as occlusion at the RCA opening, with blood supply provided by a large side branch from the LAD forming a "Vieussens' ring" to establish anastomotic connections with the RCA; the mid-distal lumen of the RCA appeared normal in the presence of the "Vieussens' ring" (7,8). Moreover, the congenital origin of this defect is further supported by the presence of 1–2 side branches (9). The RMCAA case in this report falls into the former category. Notably, there is a short, slender blood vessel in this instance rather than

Table 1 Summary of 4 reported cases of RMCAA in the literature

Reference, year	Gender	Age (y)	Presentation	Diagnostic methods	DSA/CCTA findings	Other examinations	Treatment	Post-treatment
Mandurino-Mirizzi A, *et al.* (5), 2022	Male	65	Sudden cardiac arrest	DSA + CCTA	Circumflex running into the atrioventricular groove toward the RCA and ending with hypoplastic small branches close to the right coronary cusp	ECG, echocardiogram, cardiac magnetic resonance, and myocardial ^{18}F-FDG PET/CT: (–)	No coronary surgery	Follow-up
Gupta MD, *et al.* (6), 2015	Male	62	Atypical chest pain	DSA + CCTA	RMCAA, and circumflex running into the atrioventricular groove toward the RCA	ECG and echocardiogram: (–)	N/A	N/A
Saremi F, *et al.* (7), 2011	Male	46	Palpitation, dizziness	DSA + CCTA	Atresia of the right coronary ostium, and an enlarged right conus artery joined the proximal LAD to form a large "Vieussens' ring"	ECG: ventricular tachycardia	No coronary surgery	Follow-up
McMahon CJ, *et al.* (8), 2000	Male	2	Heart murmur	DSA + CCTA	Atresia of the right coronary ostium, a large communicating feeding vessel from the LAD to the mid portion of the RCA	ECG and echocardiogram: (–)	No coronary surgery	Follow-up

RMCAA, right main coronary artery atresia; y, years; DSA, digital subtraction angiography; CCTA, coronary computed tomography angiography; RCA, right coronary artery; ECG, electrocardiogram; ^{18}F-FDG PET/CT, ^{18}fluoro-desoxyglucose-positon emission tomography/computed tomography; N/A, not applicable; LAD, left anterior descending.

the RCA being totally absent. This implies that the patient's RCA malformation is as a result of congenital developmental abnormalities, a condition recognized by Tomanek, Mandurino-Mirizzi, and others (4,5) as a CCA manifestation.

Revascularization surgery can significantly reduce mortality in both children and adults with CAA (3,10). Early surgical intervention, especially in children, has been shown to improve prognosis, and surgical intervention can even be considered in infants diagnosed with CAA (11). The main surgical approaches include coronary artery bypass grafting and coronary artery reconstruction osteoplasty. In recent years, coronary artery reconstruction osteoplasty has been preferred in the surgical setting because it can restore the physiological blood flow pattern in the coronary arteries and provide more blood flow reserve to the myocardium. However, coronary artery osteoplasty is not only technically challenging but also requires the presence of a well-developed main coronary artery. Additionally, percutaneous coronary intervention (PCI) has recently been successfully used to treat LMCA atresia. However, the implementation of PCI requires a regressive main trunk, and it is difficult to perform PCI in cases without a regressive main trunk (12). In this case, the patient had been asymptomatic for 58 years and recently developed intermittent precordial pain, and the presentation was consistent with angina pectoris induced by myocardial ischemia (inadequate compensation by collateral circulation). Moreover, the patient responded well to conservative therapy. Therefore, instead of a surgical intervention at this time, the patient has been recommended to follow up with our department at regular intervals.

In conclusion, RMCAA is an uncommon condition, and CCTA is important for its diagnosis. The choice between surgical intervention and conservative management should be based on the patient's vascular anatomy and clinical symptoms.

Acknowledgments

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-23-1808/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Declaration of Helsinki (as revised in 2013). Written informed consent was obtained from the patient for publication of this article and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

References

1. Alsalehi M, Jeewa A, Wan A, Contreras J, Yoo SJ, Laks JA. A case series of left main coronary artery ostial atresia and a review of the literature. Congenit Heart Dis 2019;14:901-23.
2. Unzué L, García E, Parra FJ, Palomo J, Friera LF, Solís J. Congenital atresia of the left main coronary artery in an adult: A rare anomaly with an unfavorable prognosis. Review of the literature. Cardiovasc Revasc Med 2015;16:498-502.
3. Musiani A, Cernigliaro C, Sansa M, Maselli D, De Gasperis C. Left main coronary artery atresia: literature review and therapeutical considerations. Eur J Cardiothorac Surg 1997;11:505-14.
4. Tomanek R, Angelini P. Embryology of coronary arteries and anatomy/pathophysiology of coronary anomalies. A comprehensive update. Int J Cardiol 2019;281:28-34.
5. Mandurino-Mirizzi A, Arzuffi L, Negrello E, Crimi G, Ferlini M, Marinoni B, Repetto A, Mauri V, Marconi S, Ferrario M, Oltrona Visconti L. Right coronary artery atresia in an athlete presenting with cardiac arrest: a case report. Coron Artery Dis 2022;31:64-5.
6. Gupta MD, Girish MP, Vignesh V, Narang P, Trehan V, Tyagi S. Absent right coronary artery: A case of single coronary artery or congenital ostial atresia? Indian Heart J 2015;67 Suppl 3:S11-3.
7. Saremi F, Goodman G, Wilcox A, Salibian R, Vorobiof G. Coronary artery ostial atresia: diagnosis of conotruncal anastomotic collateral rings using CT angiography. JACC Cardiovasc Imaging 2011;4:1320-3.
8. McMahon CJ, El Said HG, Clapp SK. Atresia of the proximal right coronary artery in association with coronary artery fistula. Catheter Cardiovasc Interv 2000;50:337-9.
9. Pandey NN, Sinha M, Sharma A, Rajagopal R, Bhambri K, Kumar S. Anomalies of coronary artery origin: Evaluation on multidetector CT angiography. Clin Imaging 2019;57:87-98.
10. Tanawuttiwat T, O'Neill BP, Schob AH, Alfonso CE. Left main coronary atresia. J Card Surg 2013;28:37-46.
11. Amaral F, Tanamati C, Granzotti JA, Haddad JL, Leite JR, Barbero-Marcial M. Congenital atresia of the ostium of the left coronary artery. Diagnostic difficulty and successful surgical revascularization in two patients. Arq Bras Cardiol 2000;74:339-42.
12. Yin T, Zhong X, Tao L, Hu T. Revascularization by percutaneous coronary intervention in a case of left main coronary artery atresia. Catheter Cardiovasc Interv 2021;98:1343-8.

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