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Case Report

Anomalous right coronary artery from the main pulmonary artery (ARCAPA): Incidental finding in an asymptomatic septuagenarian ☆

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ABSTRACT

Anomalous right coronary artery (RCA) from the main pulmonary artery (ARCAPA) is a rare finding. Clinical presentations range from asymptomatic to sudden death. We present the case of ARCAPA in a septuagenarian initially suspected on a screening chest computed tomography (CT) and later confirmed on cardiac CT. A summary of important points related to this entity is also discussed.

Introduction

Anomalous right coronary artery (RCA) from the main pulmonary artery (ARCAPA) is found in <0.002% of the population according to angiographic data [1]. With the growing use of computed tomography (CT) for coronary calcium scoring, lung cancer screening as well as frontline testing for patients without known coronary artery disease who present with chest pain as recommended by the recent new chest pain guidelines [2], the use of chest and cardiac CT is expected to increase in the coming years. We present an incidental finding of ARCAPA in an asymptomatic elderly gentleman which was first suspected on screening chest CT. Given the expected rise in CT studies in the coming years, incidental findings such as the one presented in this report are likely to increase.

Case presentation

The patient is a 74-year-old man with a history of hypertension and hyperlipemia who worked as a firefighter for 30 years. He was never a smoker. A chest CT ordered for lung cancer screening showed multiple serpiginous vascular structures along the anterior pericardium concerning a coronary vascular malformation (Fig. 1). He was referred for cardiac CT coro-

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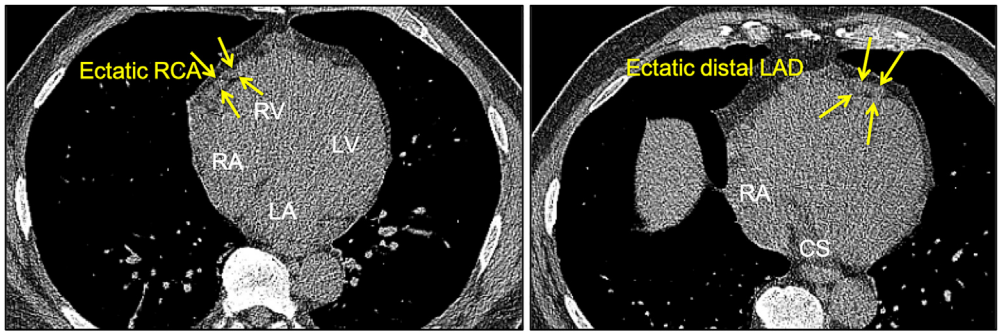

Fig. 1 – Noncontrast chest CT ordered for lung cancer screening showed multiple serpiginous vascular structures along the right-ventricular free-wall corresponding to an ectatic right coronary artery (left panel, yellow arrows) and along the apex of the heart corresponding to ectatic distal left coronary artery (right panel, yellow arrows) as well as area of fistualization. RCA, right coronary artery; LAD left coronary artery; RA, right atrium, LA, left atrium; RV, right ventricle; LV, left ventricle; CS, coronary sinus.

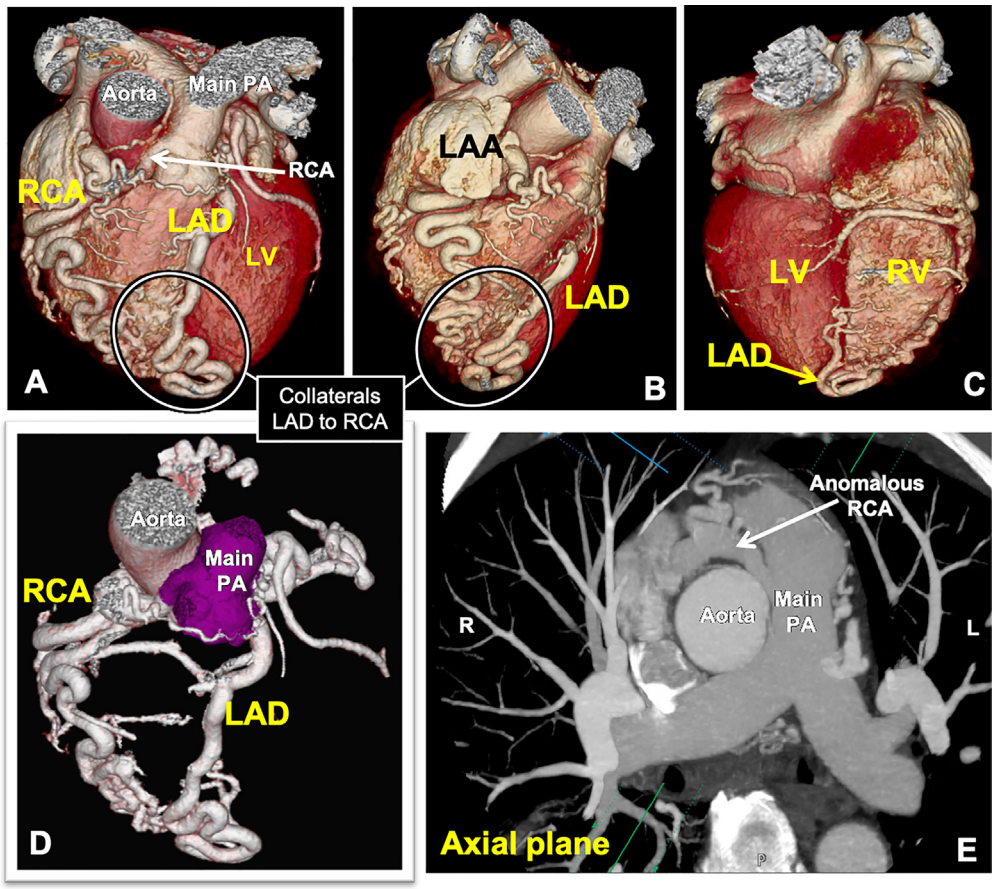

Fig. 2 – 3D reconstruction of the patient's heart on cardiac CT showing anomalous RCA (panel A, arrow) and ectatic coronary vessels. There is collateral formation between the LAD and RCA (circle, panel A and panel B). An ectatic RCA is noted in panel B and the posterior heart is visualized in panel C illustrating the dominance of the RCA with the LAD extending to supply the LV apex. Panel D shows the coronary tree with extensive collateralization between the LAD and RCA. Panel E shows the upper mediastinum in the axial plane. Here the ascending aorta and main pulmonary artery are seen. The anomalous RCA is also visualized as it comes off the main PA. RCA, right coronary artery; LAD, left coronary artery; LV, left ventricle, PA, pulmonary artery; LAA, left atrial appendage; RV, right ventricle.

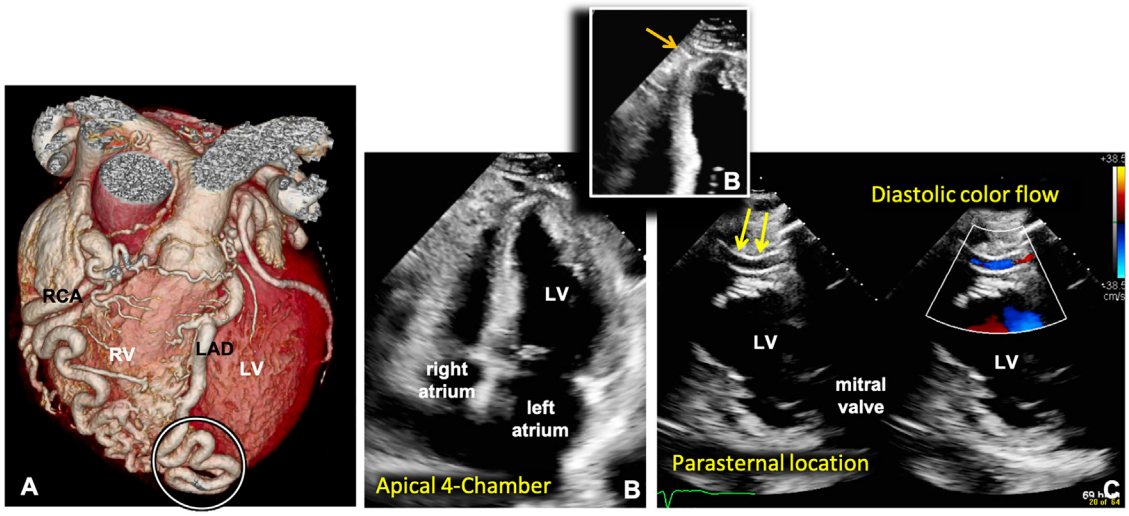

Fig. 3 – In this figure, the echocardiogram performed at the time of cardiac CT is shown alongside the 3D reconstruction (panel A). Evidence of ectatic LAD and RCA can be seen in the apical 4-chamber view (panel B, orange arrows). The vessel at the LV apex is likely the LAD while the vessel at the RV free wall is likely the RCA. Diastolic color flow is noted (see EKG tracing in green) in the coronary vessels which appear to be best interrogated in the parasternal window (panel C). RCA, right coronary artery; LAD, left coronary artery; LV, left ventricle; RV, right ventricle.

nary angiography (CCTA) for further evaluation. CCTA showed an anomalous right coronary artery (RCA) originating from the main pulmonary artery (Figs. 2A–E) with ectatic, tortuous vasculature (Figs. 2B–D) without the significant atherosclerotic disease. The RCA was dominant and noted to receive collaterals from the left coronary artery (LCA) (Figs. 2A and B). A transthoracic echocardiogram showed an upper normal left ventricular (LV) end-diastolic diameter (57 mm) with a normal LV ejection fraction (65% using Simpson biplane analysis). The right ventricle was normal in size and function. Ectatic coronary vessels were noted at the LV apex (Figs. 3A and B) and on parasternal short-axis views (Fig. 3C) with evidence of diastolic intra-coronary flow on color Doppler (color scale velocity = 38.5 cm/s).

Discussion

ARCAPA can be associated with hypoxia and ischemia in the myocardial tissue supplied by the anomalous artery. This is variably counteracted by LCA-RCA shunting or fistulization leading to arterial remodeling and ectasia. Surgery in symptomatic patients with ventricular dysfunction or myocardial ischemia [3] was afforded a class IIa recommendation by the 2018 American College of Cardiology guidelines. Our patient played golf several times a week and was able to climb 2 flights of stairs without symptoms, and was consequently deemed to have an excellent performance status. It was therefore decided not to intervene or perform further testing at this time. A systemic review of 223 cases of ARCAPA found that symptomatic patients have a bimodal age distribution at presentation, the first near birth and the second around 50 years [4]. Interestingly, 38% of patients were asymptomatic at diagnosis and only identified after a continuous murmur was heard on physical examination [4]. Echocardiographic findings typically included LV dilation, coronary artery dilation, and collaterals between the LCA and RCA. When patients with ARCAPA are referred for surgery the operation typically involves either re-implantation of the RCA onto the aorta with the re-creation of a dual coronary system or ligation of the RCA with coronary bypass grafting. Operative mortality of at least 2.5% has been reported which is not insignificant [4].

The cardiac CT in this patient demonstrated severely dilated and ectatic right and left coronary systems, a finding which has been described in prior cases. Anomalous coronary arteries arising from the pulmonary trunk can be associated with hypoxia in the myocardial tissue supplied by the territory of the anomalous artery and consequent ischemia. This is variably counteracted by LCA-RCA shunting or fistulization due to the presence of a pressure gradient between the coronary and pulmonary circulations thus leading to arterial remodeling and ectasia via the production of hypoxia-inducible factor (HIF)-1 and vascular endothelial growth factors (VEGF) [5]. Myocardial ischemia related to ARCAPA is generally not as profound as that occurring secondary to anomalous left coronary artery originating from the pulmonary artery (ie, ALCAPA) due to the lower oxygen consumption of the right ventricle compared with the left ventricle. Right-dominant circulations, however, are less tolerant of this disorder than left-dominant systems [1].

To our knowledge, this is the first case of incidental ARCAPA in an asymptomatic elderly patient with right dominant coronary circulation and no adverse cardiac remodeling. Given the growing use of CCTA many more such cases may be uncovered in the future. Indeed, the above-quoted incidence of ARCAPA might be an underestimation [1].

Patient consent

This is to confirm that consent to publish the case report was obtained from the patient.

REFERENCES

[1] Vergara-Uzcategui CE, das Neves B, Salinas P, Fernandez-Ortiz A, Nunez-Gil IJ. Anomalous origin of coronary arteries from pulmonary artery in adults: a case series. Eur Heart J Case Rep 2020;4(2):1–5.
[2] Gulati M, Levy PD, Mukherjee D, Amsterdam E, Bhatt DL, Birtcher KK, et al. 2021 AHA/ACC/ASE/CHEST/SAEM/SCCT/SCMR guideline for the evaluation and diagnosis of chest pain: a report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. J Am Coll Cardiol. 2021;78(22):e187–285.
[3] Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: executive summary: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation 2019;139(14):e637–97.
[4] Guenther TM, Sherazee EA, Wisneski AD, Gustafson JD, Wozniak CJ, Raff GW. Anomalous origin of the right coronary artery from the pulmonary artery: a systematic review. Ann Thorac Surg 2020;110(3):1063–71.
[5] Mantha Y, Harada R, Hieda M, Assaf A, Tatiraju U, Feldman M, et al. Masson Tumor in the Left Atrial Appendage Presenting as Cardioembolic Cerebral Infarction. JACC Case Rep. 2020;2(12):1969–73.