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Clinical and Pathological Findings of an Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery

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Abstract

An anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital coronary artery malposition. We describe its surgical re-implantation in a 49-year-old man with marked dilatation of both coronary arteries. We decided to perform surgery based on intraoperative esophageal echocardiographic findings, 15 min after clamping the right coronary artery at its origin. Re-implantation from the main pulmonary artery to the ascending aorta was performed successfully. The dilated left coronary artery had diminished in size. Pathological findings of the excised proximal arterial wall of the ARCAPA showed much amount of elastic fibers in the arterial wall as an elastic artery thought to have originated from the pulmonary trunk.

Keywords: Pulmonary artery; Palpitation; Cardiopulmonary bypass; Angiography

Abbreviations: ARCAPA: Right Coronary Artery from The Pulmonary Artery; Ao: Aorta, PA: Pulmonary Artery, RCA: Right Coronary Artery, LCA: Left Coronary Artery; IEL: Internal Elastic Lamina

Introduction

An anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital coronary artery malposition [1]. There was no report of pathological findings of ARCAPA. We present the case with ARCAPA and pathological findings, which was incidentally diagnosed with the condition after coronary 3DCT performed for palpitation.

Case Study

A 49-year-old man was admitted to the hospital with a 20-year history of palpitation. Coronary 3DCT revealed an ARCAPA with marked dilatation of both coronary arteries. Cardiac catheterization revealed a Qp/Qs ratio of 1.52 and a left to right shunt ratio of 34% (Figure 1). After full median sternotomy, cardiopulmonary bypass was established by ascending aortic cannulation and right atrium drainage. We first clamped the RCA near the pulmonary trunk for 15 min and trans esophageal echocardiography revealed that his septal wall motion was decreased from ischemia. Thus, the RCA was ligated and divided near the pulmonary trunk and then anastomosed to the ascending aorta with continuous sutures using 6-0 polypropylene.

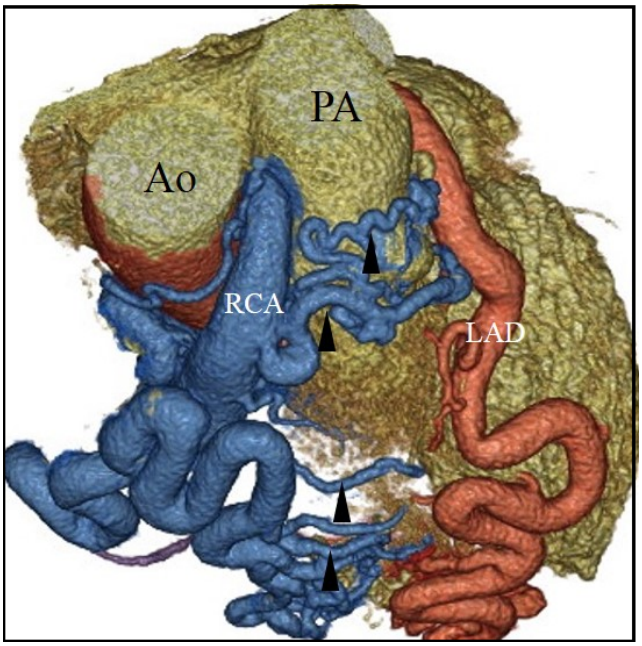

Figure 1: Tree-dimensional computed tomography revealed that the right coronary artery was originating from the pulmonary trunk. (): Collateral arteries between RCA and LCA.

Postoperative 3DCT revealed disappearance of collateral arteries between the RCA and LCA, whose size were decreased in size (Figure 2a). Histological examination of the excisional proximal wall of ARCAPA, stained by Masson Trichrome stain showed that the wall was consisted of intima, internal elastic lamina, media and well thickened adventitia with much amount of elastic fibers (Figure 2b).

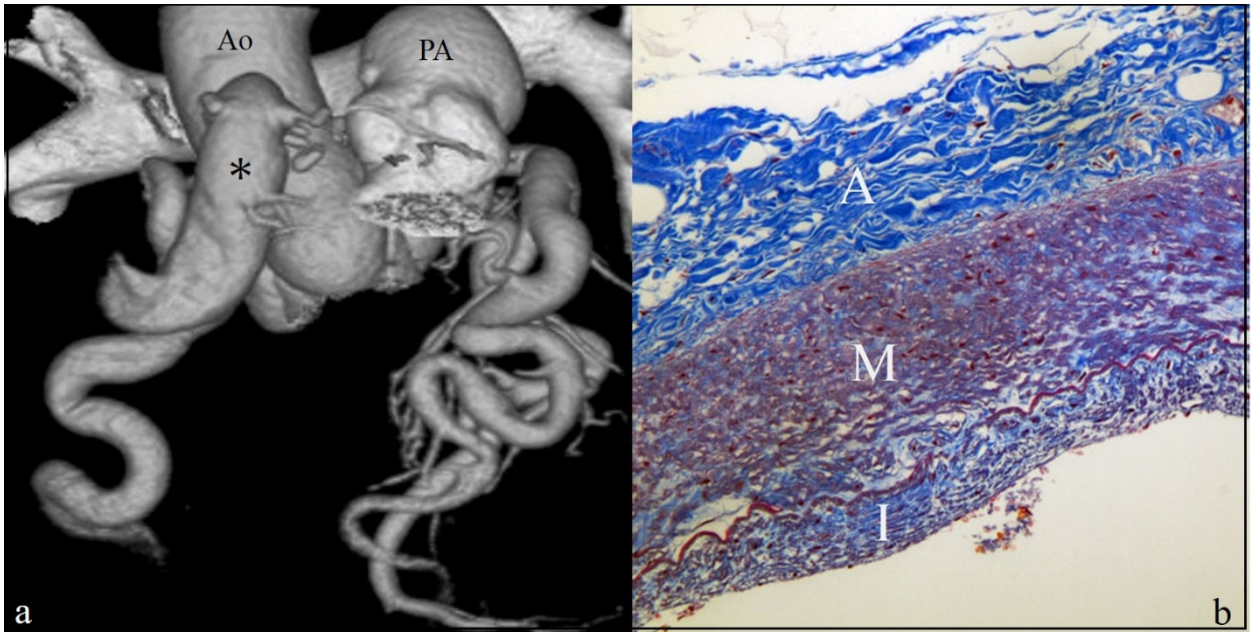

Figure 2a and 2b: 2a-Postoperative tree-dimensional computed tomography showed disappeared collateral arteries between reconstructed RCA (*) and LCA. 2b-In the excisional proximal wall of ARCAPA containing intima (I), internal elastic lamina (IEL), media (M), and well thickened adventitia (A) with much amount of elastic fibers (blue stain).

Discussion

An ARCAPA is an extremely rare congenital heart disease. Yamanaka and Hobbs detected ARCAPA in 0.002% of the population who underwent angiography [1]. Most patients with ARCAPA are asymptomatic until old age. ARCAPA could show different clinical signs of heart murmur, dyspnea, mitral regurgitation, angina, congestive heart failure, and cardiac arrest [2,3]. Recently, coronary computed tomography angiography (CCTA) has become quite useful for diagnosing and confirming the anatomical images for ARCAPA [4].

With respect to treatment, most patients reported to have ARCAPA underwent re-implantation of the ARCAPA from the pulmonary trunk to the aortic root, and others underwent simple ligation. Regression of the diameter of the dilated coronary artery was expected after re-implantation [2]. However, there has been a report of a patient who experienced coronary rupture after re-implantation who had fragile vessels with low pulmonary arterial pressure for a long period that may have been unable to withstand systemic blood pressure [5]. Thus, careful consideration should be given in older patients with a dilated coronary artery. The lower part of the septal wall and inferior wall of the left ventricle are at considerable risk for myocardial ischemia or necrosis in patients with ARCAPA [4], so we performed re-implantation. After that operation, his coronary artery decreased in size without rupture and the collateral arteries between LCA and RCA disappeared. There was much amount of elastic fibers in resected proximal RCA, suggesting more like elastic artery than muscular artery and it was different from the normal coronary artery of muscular artery. This finding indicates that the proximal ARCAPA had arisen from the pulmonary sinus and that it was connected to the distal RCA from the myocardium [3,5].

Conclusion

We report a very rare case of an adult ARCAPA and successfully underwent re-implantation to diminish the coronary artery and cause the collateral arteries between the RCA and LCA to disappear. The pathological findings of the ARCAPA indicated an elastic artery thought to have originated from the pulmonary trunk.

Conflicts of Interest

There are no conflicts of interest for the present study.

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