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Case study

Multiple coronary artery–left ventricular fistulas causing sudden death in a young woman

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Summary Multiple coronary artery fistulae arising from right and left coronary arteries were found at autopsy in a 22-year-old woman, dying suddenly while playing football. This is the fifth pathologic description of this finding with biventricular involvement. We found microscopic evidence of postischemic scars and foci of myocardial calcifications in the left ventricular wall.

1. Introduction

Coronary artery fistulas (CAFs) are rare, abnormal communications between coronary arterial bed and cardiac chambers or pulmonary circulation. They account for 0.08% to 0.4% of all congenital heart disease [1], and the symptoms depend upon the amount of shunted blood bypassing the myocardial capillary network (so-called steal phenomenon) [1].

CAFs are generally benign, and most patients are asymptomatic due to small shunt size. However, serious complications have been reported, more frequently during adulthood, including pulmonary hypertension, congestive heart failure, bacterial endocarditis, rupture or thrombosis of the fistula, myocardial ischemia, and sudden death (SD).

To our knowledge, only 4 cases of SD for coronary fistulae have been reported, with the age of patients ranging from 21 to 29 years [2,3] for 3 cases and only 1 older patient, a 62-year-old man.

2. Case report

A 22-year-old woman collapsed and died suddenly while playing a football match, showing signs of fatigue a few minutes before death. She had no family history of sudden death or cardiovascular disease. However, she had an "innocent" cardiac murmur not further investigated. At autopsy, the heart weighed 270 g. The whole epicardial coronary tree was tortuous and dilatated; on multiple section, a few coronary branches of a dilated and serpiginous conal artery could be traced down to the endocardial surface of the right ventricle (Fig. 1A and B). A CAF connection existed between the proximal third of the left anterior descending coronary

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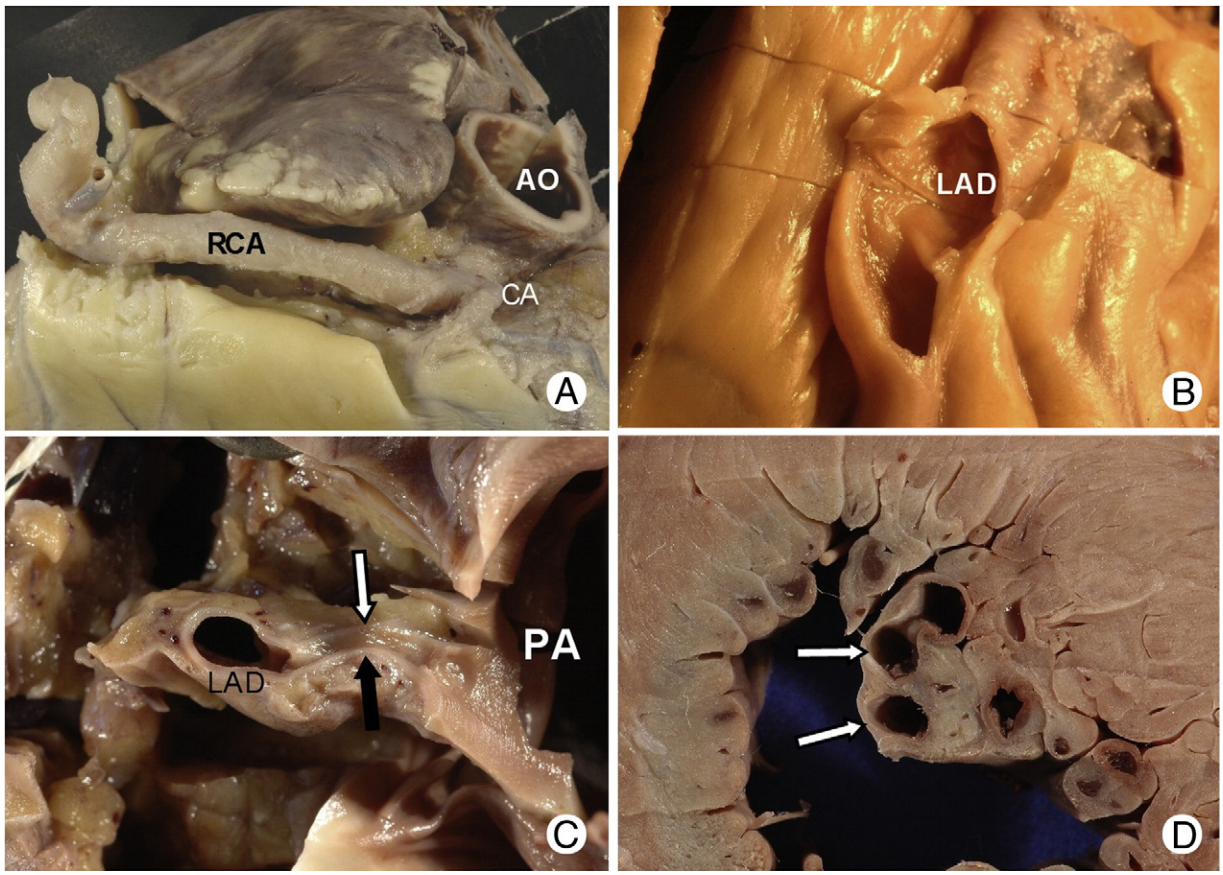

Fig. 1 A and B, Dilated right coronary artery (RCA) and branching of the conal artery (CA) (A) and left anterior descending coronary artery (LAD) (B); a close up of the fistula (arrows) connecting the pulmonary artery (PA) to the LAD (C); large sinusoidal cavities in the subendocardial myocardium of the left ventricle (arrows) (D). AO indicates aortic root.

artery (LAD) and the pulmonary trunk, behind the left pulmonary valve cusp (Fig. 1C). Just beneath the subendocardial surface of the left ventricle, there were many large blood-filled, thin-walled vascular spaces (Fig. 1D). Many perforating branches of the epicardial coronaries showed markedly enlarged caliber with a through-and-through course.

The histolopathologic appearance of the CAFs was as reported by Gittenberger-de Groot et al [4]. Many coronary intramyocardial branches, bypassing the capillary network in both ventricles, opened directly into the ventricular lumen (Fig. 2B) resembling ventriculocoronary arterial communications (VCAC). They showed diffuse dysplastic changes (Fig. 2A and B), with irregular shape of the lumen in transverse section. Medial smooth muscle cells were disorganized [4]. In addition, a great number of anomalous intramural arterioles opened up in vascular spaces without a defined tunica media, located in the subendocardial myocardium, showing a thin sinusoidal wall. The endothelial lined lumen, partly obstructed by large and thick intimal fibrous cushions (Fig. 2C), did not show thrombotic layering. Multifocal fibrous scars were seen in the myocardium of the left ventricle, close to the larger sinusoidal spaces. Focal "mummified," calcified (postnecrotic) myocytes were found within large fibrous scars located in the left ventricle wall (Fig. 2D). No acute necrotic (ie, coagulative necrosis) changes were shown in the myocardium.

3. Discussion

Most CAFs arise from the right coronary artery and terminate in the right side heart chambers or in the pulmonary artery, but in few reports, as in the present case, the LAD is the predominant vessel involved with multiple fistulous connection to the left ventricular cavity [2,3]. The proportion of symptomatic patients increases with age, and when present, clinical signs depend on the number and size of the CAFs as well as their coronary origin and site of drainage [1].

In the 4 previously reported cases of SD and in our patient, CAFs were shown between the left epicardial coronary branches and the left ventricular cavity. In the present case, right and left coronary arteries were dilated, suggesting that both arteries were involved by large CAFs.

CAFs are commonly considered a failure of obliteration of primordial communications between the fetal coronary circulation and the heart cavities.

Several factors are involved in coronary arteriogenesis including sprouting, deprogramming, and reprogramming

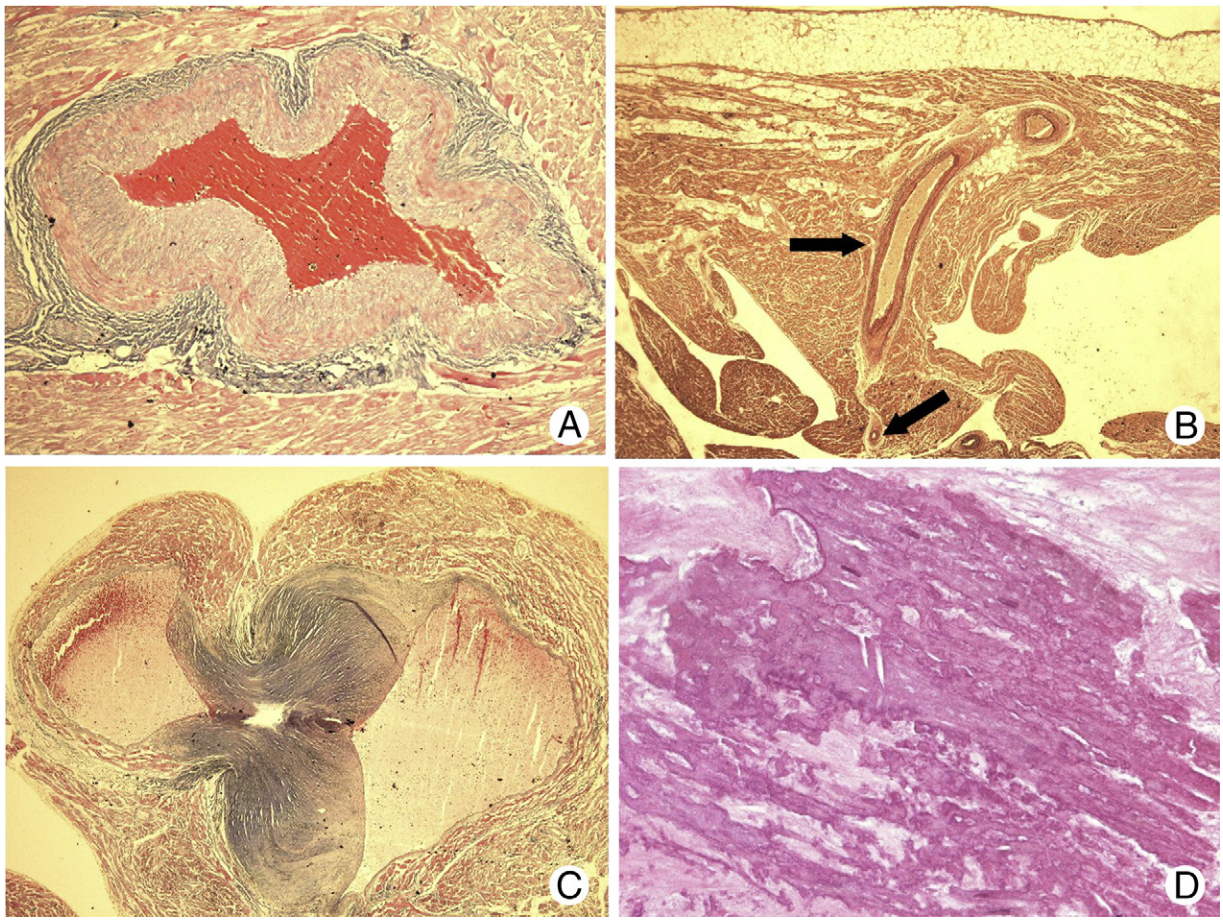

Fig. 2 A, Irregular luminal shape of a dysplastic intramyocardial arterial-sinusoidal vessel (Masson trichrome stain, original magnification ×200). B, Low-power view of a dilated coronary dysplastic coronary branch, through the whole thickness of the right ventricle wall (arrows) (Verhoeff-van Gieson stain). C, Severe luminal narrowing of a subendocardial sinusoid (Masson trichrome stain original magnification ×100). D, Postnecrotic calcified myocardial fibers, embedded in a fibrous scar (hematoxylin-eosin stain, original magnification ×150).

of venous progenitors [5]. Thus, a fistulous communication may demonstrate a failure of this complex developmental process with severe modifications of coronary microcirculation.

Our study disclosed an anomalous persistence of 2 embryonal structures; the first ones were constituted by direct communications between the coronary subepicardial arteries and the ventricle lumen (VCAC); the latter ones were sinusoid vascular spaces, otherwise described "myocardial sinusoids" (MS) and reminiscent of embryonal dilated trabecular sinusoids [4].

The VCAC and sinusoid vascular spaces that we found in our study are common in pulmonary atresia with intact ventricular septum; nevertheless, concerning to the former, experimental studies support the separate origin of VCAC and pulmonary atresia. Moreover, it is still debated whether there is a gradual transition between VCAC and MS [6]. On the other hand, since 1933 [7], a detailed description of available pathways in the normal coronary microcirculation identified morphological entities as "arterio-luminal" vessels and myocardial sinusoids, respectively, similar to VCAC and MS seen in our case.

The most effective treatment of CAFs is not clear, although timely diagnosis may be life saving. Percutaneous intervention could obliterate, at least partially, abnormal or fistulous communications. Avoiding overexertion may also be preventive. In this case, were the diagnosis known, this young girl could be prevented from participating in strenuous sports. In retrospect, further investigation of her heart murmur may have saved her life. This case supports the argument for improved screening of young athletes with heart murmurs for SD-related anomalies such as by coronary computed tomography angiography [8]. Unfortunately, this "innocent murmur" was not so innocent.

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