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A giant right coronary artery related to a small fistula to the coronary sinus ☆

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ABSTRACT

Coronary artery fistulas to the coronary sinus are very rare coronary anomalies most often resulting in extreme dilatation of the donor coronary artery and the receiving vessel. However, despite common impressive appearance, their clinical and functional impact may be extremely various from asymptomatic and benign cases to disabling and life threatening situations. To adequately stratify the inherent risks and to plan the most appropriate therapeutic strategy, an overall investigation is necessary. We herein report the case of a 56 year-old woman with a giant right coronary artery related to a small and restrictive fistula to the coronary sinus that was extensively investigated by multi-imaging strategy before decision of a therapeutic abstention and long-term follow-up.

1. Introduction

Coronary artery fistulas are uncommon coronary artery anomalies, being reported in around 0.1–0.8% of coronary angiograms and computed tomography (CT) scanners [1–4]. Coronary artery fistulas to the coronary sinus (CS) are an even rarer features with less than 30 cases described in the literature worldwide.

We herein report the case of a 56 year-old woman with a giant right coronary artery (RCA) related to a small and restrictive fistula to the CS that was extensively investigated by multi-imaging strategy before decision of a therapeutic abstention and long-term follow-up.

2. Case report

A 56 year-old woman was admitted to our hospital for atypical chest pain at rest. Of note, she had been diagnosed with a systolic murmur and possible congenital triatrial heart on the base of a sole transthoracic echocardiography (TTE) 15 years earlier.

At admission, electrocardiogram (ECG) was strictly normal, but a slight increase in troponin was noted. Non-ST-elevation acute coronary syndrome (ACS) was subsequently suspected, and the patient underwent a coronary angiogram that revealed no atherosclerotic lesion and a giant RCA that was extremely tortuous and dilated with drainage into an enlarged chamber resulting to be the CS (Fig. 1).

She underwent TTE powered by 3D real time acquisitions that showed normal systolic and diastolic bi-ventricular functions with no sign of right heart overload. Ultrasound imaging and Color-Doppler/PW indicated a normal and non-stenotic drainage of the enlarged CS to right atrium (RA) by a 7 mm-diameter orifice but did not show precisely the fistula between RCA and CS. Any significant compression of cardiac chambers by the vascular formation was excluded (Fig. 2).

A CT scanner with prospective ECG gating was then performed aiming a more detailed analysis of anatomic characteristics of the arterio-venous formations. RCA was found having an internal diameter between 8 and 23 mm, but its communication to the CS was restricted to a 5 mm-diameter channel. The CS was massively dilated with maximal diameter of 52 mm. No thrombotic apposition was present inside vascular formations. CS to RA passage was identified normal and measured to only 5 mm of diameter as well (Figs. 1–4).

Functional impact of the fistula was evaluated by a 6 minute walking test that was sensibly normal with a perimeter of 390 m, normal heart rate, normal oxygen saturation and Borg scale rating of perceived dyspnea pointed at 0 by the patient, and exercise performance estimated at more than 4 metabolic equivalents (METs). ECG-Holter monitoring revealed only rare atrial ectopic activity. Finally, she underwent right heart catheterization that measured a cardiac output at 5.58 L/min and cardiac index at 3.65 L/min/m^2. There were no significant shunt (QP/QS = 1.1), normal pulmonary artery pressure at 28/9 mmHg (mean 17 mmHg), normal pulmonary resistances at 113 dyn.s/cm^5 and no increase of oxygen content in RA (SaO2 75% in superior vena cava, 78% in RA, and 78.7% in pulmonary artery).

Subsequently to the lack of any functional impact of such an impressive congenital abnormality, no invasive treatment was performed, and the patient was dismissed with indication to clinical and imaging follow-up. At 1 year, the patient is doing perfectly well

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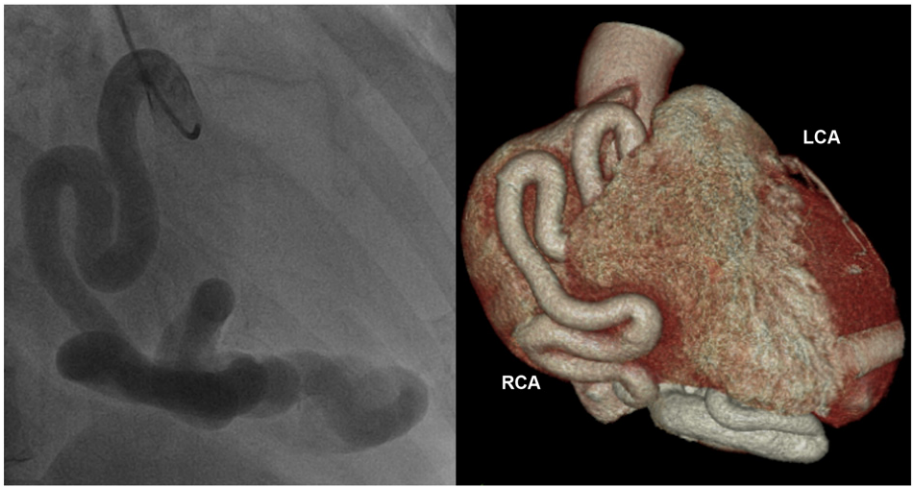

Fig. 1. Coronary angiogram and volume rendered multi detector computed tomography showing giant right coronary artery (RCA) to dilated coronary sinus (CS) fistula.

with no recurrent chest pain and no significant modification in TTE and CT scanner images.

3. Discussion

Congenital coronary arterio-venous fistulas are rare anomalies resulting from persistence during embryonic development of primordial epicardial vessel junctions with intra myocardial sinusoidal circulation [1,5]. Iatrogenic or post-traumatic fistulas have also been exceptionally described [6–8]. Fistulous connection to the CS is generally characterized by marked dilatation of the donor coronary artery and CS. RCA is the most frequently coronary artery involved [9], but fistulas involving left main, left circumflex or left anterior descending coronary arteries are also reported [2,10]. Anatomic and functional impact as well as clinical presentation are extremely variable, from totally asymptomatic state to progressive cardiac failure [1,5,6,8,11]. Sudden complications as thrombosis, infective endocarditis and rupture with cardiac tamponade are also described [1,12,13]. However benign evolution of such congenital abnormalities is rather the rule than the exception despite frightening appearance of diagnostic imaging and coronary artery fistulas are often discovered late in life.

It is important to note that diagnosis is often challenging, and many patients receive wrong interpretations of their anomalies before being correctly diagnosed by multi-imaging analysis, as it was the case for our patient. She had already undergone TTE 15 years before, and the anechoic space next to the RA and probably the wrong setting of Color-Doppler scale suggested a diagnosis of congenital triatrial heart. The patient was perfectly asymptomatic at that time, and no other investigation was proposed.

The treatment of such congenital abnormalities is not well defined. Percutaneous or surgical corrections are feasible but carry a significant risk of peri-operative myocardial infarction. Rupture of the dilated coronary artery after fistula ligation is also reported [9,14]. Therefore functional impact has to be well established before proposing any invasive treatment. Surgical correction may be proposed in presence of significant left-to-right shunt with pulmonary and right chambers overload, left atrium compression and mitral annulus deformation, risk of intra-pericardial rupture of dilated cavities.

The originality of the present case is the important discrepancy between the extreme dilatation of both RCA and CS and the lack of any significant clinical and functional consequences. To state this, we decided to investigate the vascular anomaly by multi-imaging strategy in order to carefully analyze all necessary information before deciding the most appropriate therapeutic option. In our case, coronary angiogram was the first exam performed because the patient was initially referred for suspected acute coronary syndrome. Even if it was the exam that initially leads to the right diagnosis, it is however not the exam that should be performed in first intention to explore congenital heart malformations. Coronary angiogram carried in the present case limited information with impossibility to exactly determine the size of the CS itself on one side and of RCA to CS and CS

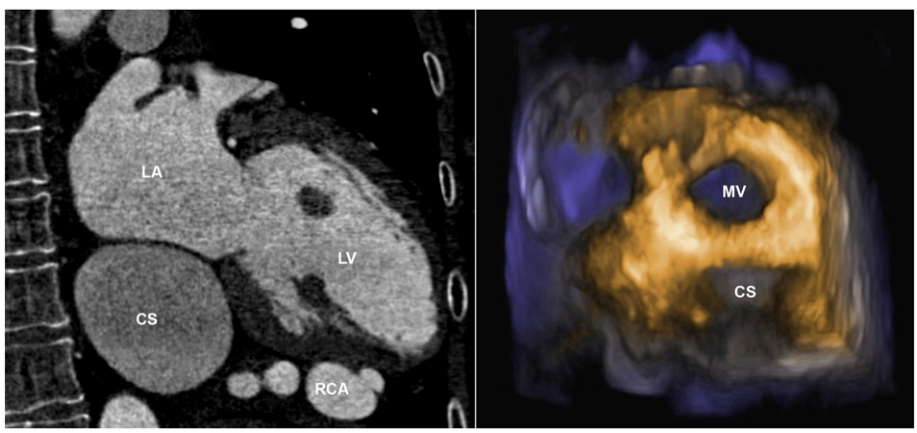

Fig. 2. Anatomic relationship between dilated coronary sinus (CS) and heart chambers depicted by multi detector computed tomography scanner (left) and 3D transthoracic echocardiography (right). LA: left atrium, CS: coronary sinus, RCA: right coronary artery, MV: mitral valve, LV: left ventricle.