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Case Report

Congenital coronary artery fistula presenting later in life

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Summary A 53-year-old male presented to our tertiary medical center with complaints of dyspnea and exertional chest pain with mild left ventricular dysfunction and right ventricular enlargement on echocardiography. Cardiac catheterization showed a congenital right coronary artery fistula communicating with the right sided chambers. Using contrast enhanced multi-detector computed tomography scan, the fistula was clearly draining into the coronary sinus. We describe briefly the etiology of coronary artery fistula, its clinical presentation, and the common tests used to confirm diagnosis. We further discuss the types of treatment modalities that are currently available.

Introduction

Congenital coronary artery fistulae are rarely seen in our daily practice. They comprise less than 1% of all congenital heart defects [2]. Symptoms occur in childhood or early adulthood depending on the type of fistula, size, and shunt severity [1]. Congenital coronary artery fistulae are usually found incidentally on coronary angiograms. Coronary computed tomography angiogram is a helpful non-invasive method to diagnose coronary artery fistula as it can delineate the nature of the fistula, number, origin and insertion sites. Patients can be managed conservatively or may require fistula closure with increased shunt severity (Qp/Qs >1.5) [1]. This is done surgically or using percutaneous transcatheter closure devices based on type and nature of the fistula [5]. We describe below a patient who presented in late adulthood with a right coronary artery fistula to the coronary sinus and significant left to right shunting. We review briefly the etiology of coronary artery fistulae, clinical presentation, diagnostic tests used to establish diagnosis and current treatment modalities.

Case report

A 53-year-old male with chronic persistent atrial fibrillation and hypertension presented with several year history of

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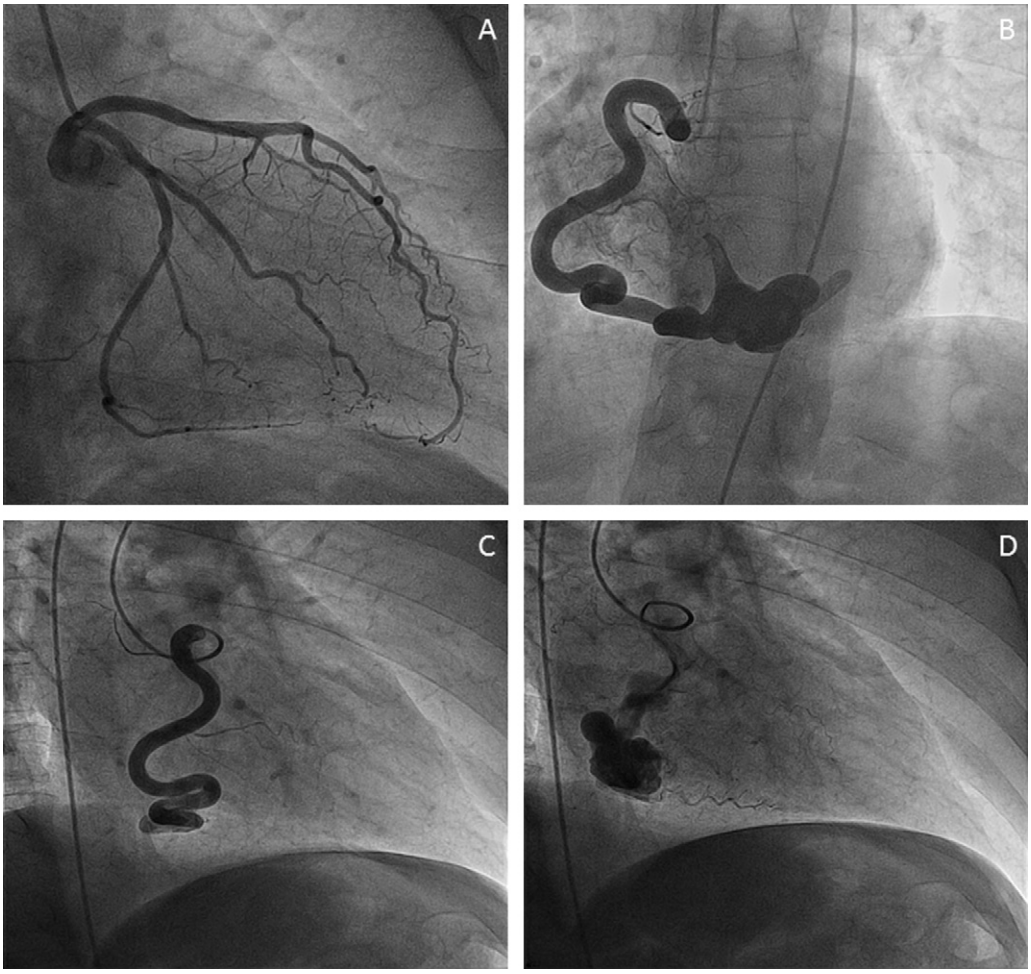

Figure 1 Coronary angiogram showing: (A) RAO caudal view of the left coronary artery with normal LAD and LCX arteries. (B) AP view of the RCA with a markedly dilated and tortuous RCA with apparent fistula draining into right-sided chambers. (C) RAO view showing a dilated RCA with increased tortuosity distally. (D) High-density contrast is seen in the right atrium consistent with a RCA fistula communicating with the right sided chambers. AP, anteroposterior; CAF, coronary artery fistula; LAD, left anterior descending; LCX, left circumflex; RAO, right anterior oblique; RCA, right coronary artery.

worsening dyspnea, fatigue, and exertional chest pain. Physical exam, chest X-ray, and serial cardiac enzymes were all unremarkable. Electrocardiography (ECG) showed atrial fibrillation rate controlled. Transthoracic echocardiography showed mildly reduced left ventricular function with basal inferoposterior wall hypokinesis, mild biatrial and right ventricular enlargement. The patient underwent conventional coronary angiography that showed normal left coronaries and a massively dilated right coronary artery (RCA) communicating with the right atrium (RA) (Fig. 1). Right cardiac catheterization confirmed the above findings with step up in oxygen saturation from 61% (RA) to 76% (right ventricle and pulmonary artery). A left to right shunt (Qp/Qs 1.4) was present; mean pulmonary artery pressure and wedge pressure were 20 mmHg and 15 mmHg respectively. As distal drainage site was not well identified, a dedicated contrast-enhanced ECG-gated multidetector computed tomogram (GE 64 multidetector CT scanner; GE Healthcare, Milwaukee, WI, USA) was performed that clearly demonstrated a tortuous and markedly dilated RCA, giving rise to a large fistula that drained into the coronary sinus (Fig. 2). Due to extreme vessel tortuosity and difficulty in cannulating the distal fistula, the patient was not a candidate for percutaneous transcatheter closure and was referred for surgical ligation of the fistula by cardiothoracic surgery. The patient requested postponing his surgery for at least a year and to be managed conservatively.

Discussion

This patient's coronary artery fistula (CAF) is a rare congenital form of coronary artery anomalies previously described in the literature [1] with an incidence of 0.2—0.47% of all congenital cardiac defects [2]. CAF can be also acquired secondary to trauma that is either iatrogenic or accidental [3]. Clinical presentation varies according to etiology, age, and duration of shunt. In congenital CAF, around 20% of patients are symptomatic in the first two decades that later increases to 60% after the age of twenty years. Neonates and infants may present with heart failure with left to right shunting or cyanotic heart disease depending on shunt type.