

Review

Interesting case of coronary artery fistula and coronary anomaly resulting in coronary steal with literature review

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Coronary artery fistulae are rare and abnormal communications between a coronary artery and a great vessel, which may shunt blood flow away from the myocardial capillary network causing steal phenomenon. We present a 47-year-old man with a unique arteriovenous fistula originating from the proximal portion of the left main coronary artery and entire left anterior descending artery, which then reconstitutes to form a branch of the left pulmonary artery.

Key words: Coronary artery fistula, coronary steal phenomenon, coronary artery anomaly, A-V fistula.

INTRODUCTION

Coronary steal is defined as alteration in blood flow patterns leading to a reduction in the blood directed to the coronary vasculature (Gould, 1989). There are several reasons for this phenomenon and one of the uncommon causes is coronary artery fistula (CAF). Our case is extremely unusual and interesting as it combines CAF and anomalous origin of coronary artery (AOCA). Congenital coronary artery fistula is a rare, isolated anomaly of the coronary artery system that is defined as a direct communication between a coronary artery and another vascular structure. Incidence of CAF in the general population is about 0.002% and the incidence of coronary pulmonary fistulae was similarly reported only to be 0.1% in a study of 11,000 patients undergoing cardiac catheterization (Said and Landman, 1990). Coronary artery anomalies are congenital and the incidence of coronary anomalies varies between 0.6 and 1.5% of patients undergoing invasive cardiovascular imaging (Kardos et al., 1997; Malouf et al., 2004). By definition, these abnormalities are variants of coronary anatomy.

Most of the coronary fistulae and anomalies are found incidentally during angiographic evaluation for coronary vascular disorder.

CASE REPORT

A 47-year-old man with a past medical history of hypertension, dyslipidemia, diabetes mellitus, and a 20 pack years smoking history presented to the emergency room (ER) with a two day history of recurring chest pain. Pain was described as 7/10 pressure like, non-radiating, substernal chest pain which progressively worsened with no significant aggravating or relieving factors. He had occasional shortness of breath and lightheadedness associated with the chest pain. No other symptoms were reported. Negative family history was reported as per the patient. The patient had similar symptoms for the past 5 months, but ignored the symptoms as he thought it was heart burn. Electrocardiogram (ECG) at the time of admission did not reveal any signs of acute ischemia, and cardiac enzymes were negative. Nuclear scan was performed which was equivocal.

Patient underwent cardiac catheterization the following day for unstable angina. AOCA was noted on the right

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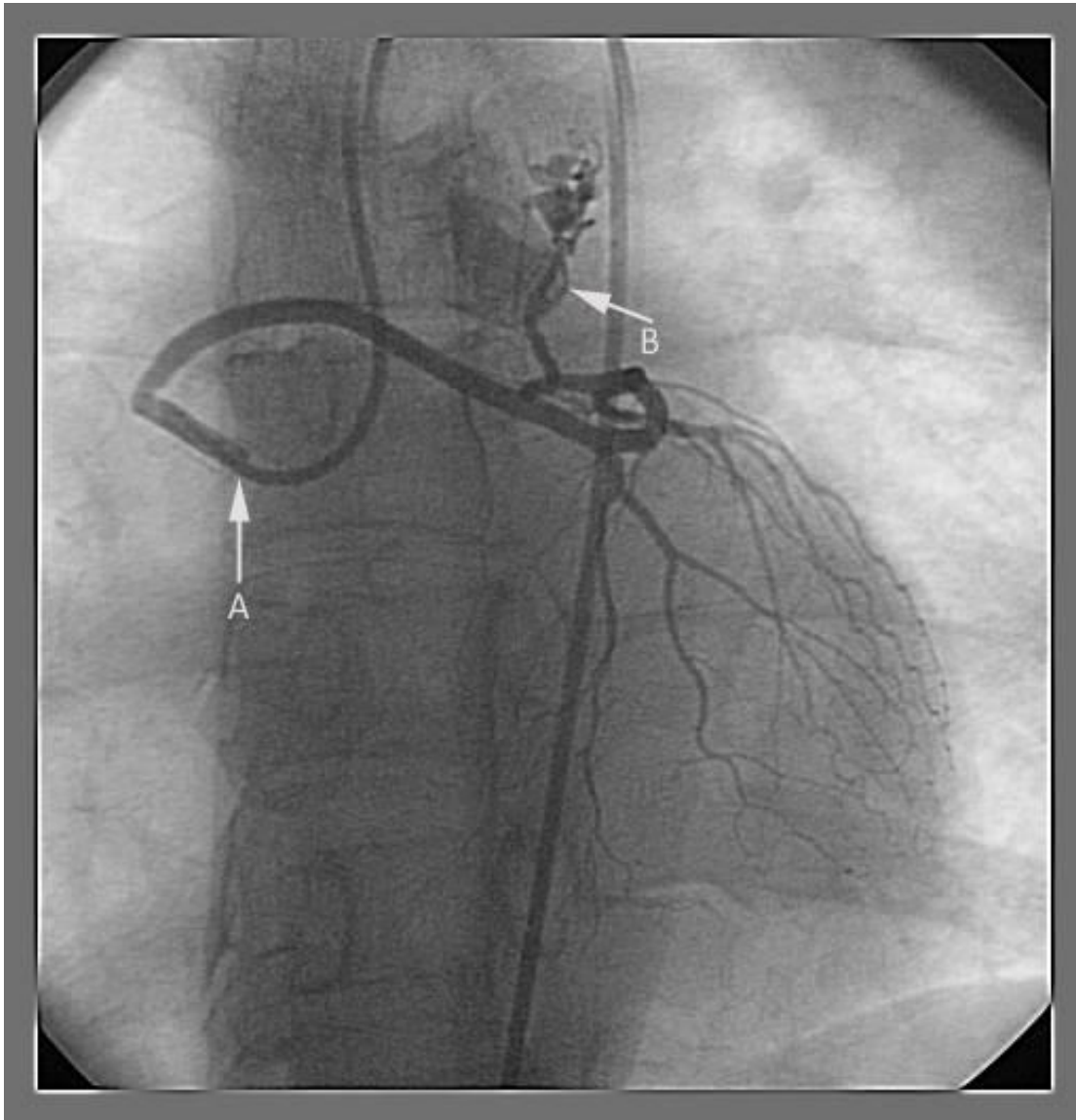


Figure 1. Patient cardiac catheterization: (A) AOCA was found on the right side where there was left anterior descending arising from the right coronary sinus and (B) significant fistulous formation arising from the left anterior descending artery which then reconstitutes to form a branch of the left pulmonary artery.

side, where there was left anterior descending arising from the right coronary sinus (Figure 1A). The catheterization also revealed a significant fistulous formation arising from the left anterior descending artery which then reconstitutes to form a branch of the left pulmonary artery (Figure 1B). Surgery was consulted and a ligation procedure along with correction of the anomaly was planned. Patient underwent corrective surgery for both anomalies and follows at a different center.

PATHOPHYSIOLOGY

Fistulae, most commonly drain into the right side of the

heart and seldom seen drain into the left atrium or the left ventricle. Occasionally, congestive cardiac failure occurs due to volume overload from redirected blood flow, and myocardial ischemia rarely occur in adults (Fernandes et al., 1992). The mechanisms that lead to myocardial ischemia in the patient who has AOCA from the opposite sinus that course between the pulmonary and aortic roots are unclear, but it was hypothesized that the inter arterial course places the anomalous coronary at risk of compression between the great arteries.

DISCUSSION

The first case of coronary artery fistula (CAF) was

described by Klause (1865). CAF is defined as any abnormal communication through which coronary artery blood is shunted into a cardiac chamber, great vessel, or other vascular structure without passing through the myocardial capillary bed (Sapin et al., 1990). Bjork and Crafoord (1947) reported the first successfully corrected coronary artery fistula. The first successful transcatheter closure of a coronary artery fistula was reported by Reidy et al. (1983). Fistulae more frequently involve the right coronary artery and usually drain into one of the right heart chambers (Lowe et al., 1981). Symptoms and signs are dependent on the size of the fistulous connection; rarely, large fistulae can have a significant left-to-right shunt with resultant congestive heart failure and cardiomegaly in infancy (Hsieh et al., 2002). In a review of all patients who underwent coronary arteriography from 1971 to 1981 at the Cleveland Clinic Foundation, a total of 122 fistulae were identified. Of these, 17% was drained into the left ventricle and 6% was drained into the left atrium (Hobbs et al., 1982).

Looking into the embryological development, coronary artery to pulmonary artery fistulae are remnants of a vascular communication that was probably normal in the mediastinal mesoderm (Rittenhous et al., 1975). These structures represent an anomalous origin of a supernumerary coronary artery arising from a coronary bud in the posterior pulmonary portion of the truncus arteriosus (Demirkilic et al., 2004). In the fetus, pulmonary artery pressure exceeds aortic pressure, and hence, perfusion through such connections is antegrade. The smaller structures usually disappear when the pulmonary artery pressure falls below the systemic pressure at birth, when flow reverses. Persistence of arterial and venous embryologic communications into adulthood results in a fistula. Maintenance of such connections in the adult requires the presence of a large hemodynamic gradient and limited distal resistance. Rarely do these connections attain sufficient size to require surgical ligation or intervention in adults (Lloyd and Klein, 2008).

CAF was present in different ways clinically. In a review of 174 patients, fistula-related complications such as congestive heart failure (12%), myocardial infarction (4%), bacterial endocarditis (3%), and death (6%) occurred, with an overall complication incidence of 21% (Liberthson et al., 1979). Other complications including giant aneurysmal dilatation of fistula (Kato et al., 1999), fistula dissection and rupture with cardiac tamponade (Bauer et al., 1996), embolization of mural thrombi to the distal coronary bed with subsequent myocardial infarction (Shirai et al., 1994), and sustained ventricular tachycardia (Moro-Serrano et al., 1992) were also reported. These complications may lead to the development of premature atherosclerosis or become life threatening.

Various cardiac imaging modalities are utilized for diagnosis and for planning before surgical or percutaneous interventions if closure of the coronary fistula is indicated.

A significantly enlarged coronary artery can usually be detected by two-dimensional echocardiography. Continuous turbulent systolic and diastolic flow pattern characterizes the shunt entry site (Lin et al., 1995; Cox et al., 1996). Use of contrast microbubbles to enhance the color Doppler signal assists to define the location and extent of coronary artery fistulae (Goswami and Zabalgoitia, 2002). When a coronary artery fistula is present, a dilated feeder vessel with an abnormal flow pattern can be readily identified. Multiplane transesophageal echocardiography can accurately define and provide a high quality panoramic view of the origin, course, and drainage site of coronary artery fistulae.

Coronary catheterization remains the gold standard for the evaluation of coronary artery fistula. It can be used to reliably identify the size and anatomical features of the fistulous tract (Frommelt et al., 2001), but the relation of coronary artery fistula to other structures, their origin, and course may not be apparent and it may be difficult to measure abnormal tortuous blood vessels in one section. Magnetic resonance imaging (MRI) and multidetector computed tomography (CT) have also become alternative methods to evaluate the anatomy, flow, and function of CAF.

AOCA from the opposite sinus of Valsalva was associated with myocardial ischemia, ventricular arrhythmias, and sudden death, particularly, when the anomalous coronary courses are between the great arteries (Frommelt et al., 2001; Liberthson et al., 1979; Roberts, 1986; Frescura et al., 1998; Taylor et al., 1992). Although, AOCA from the noncoronary or posterior sinus of Valsalva has been described; it is rare and is not associated with myocardial ischemia or sudden death (Frommelt and Frommelt, 2004). Similarly, AOCA can occur from the opposite sinus of Valsalva (either the right coronary artery arising from the left sinus or the left coronary arising from the right sinus of Valsalva), but is not associated with myocardial ischemia unless the anomalous coronary courses are between the great arteries (Frommelt and Frommelt, 2004; Roberts and Shirani, 1992).

The ischemia from interarterial course is more likely due to the deformation of the anomalous coronary within the aortic wall during the periods of systemic hypertension, particularly, in patients who have an intramural course (Berdooff et al., 1986).

The aorta will exhibit greater wall tension than the intramural coronary within the aortic wall which results in deformation of the coronary and diminished cross-sectional area, because wall tension is determined by the radius of a vessel. As aortic wall tension increases with increasing aortic pressure during exercise, the anomalous coronary becomes flattened and coronary reserve is reduced to a point where myocardial oxygen requirements are not met. Coronary catheterization also remains the gold standard for evaluation of coronary anomalies.

Management

The management is controversial and recommendations are based on anecdotal cases or small retrospective series due to variable natural history of coronary artery fistulae (Umana et al., 2002). The main indications for closure are clinical symptoms especially of heart failure and myocardial ischemia and in asymptomatic patients with high-flow shunting, to prevent the occurrence of symptoms or complications, especially in pediatric population (Balanesu et al., 2001). Surgery and direct epicardial or endocardial ligations were traditionally viewed as the main therapeutic method for the closure of coronary artery fistulae (Balanesu et al., 2001). Surgical correction is safe and effective, with good results (Balanesu et al., 2001; Wang et al., 2001). Catheter-based closure of the fistulous connection is the non-surgical treatment option for closure of coronary fistulae, with good success reported (Qureshi and Tynan, 2001; Alekyan et al., 2002; Armsby et al., 2002). Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, controlled-release coils, controlled-release patent ductus arteriosus coils, patent ductus arteriosus plug, regular and covered stents, and various chemicals (Mullasari et al., 2002; Sreedharan et al., 2004; Pettersen et al., 2001). Repair of AOCA is primarily by surgery and techniques of the repair are beyond the scope of this paper.

Treatment of adult asymptomatic patients with non-significant shunting is still a matter of debate. Results from the transcatheter and surgical literature show that both approaches have similar early effectiveness, morbidity, and mortality (Armsby et al., 2002). The safe and effective results of both approaches support the option for elective closure of clinically significant coronary artery fistulae in childhood (Armsby et al., 2002). Antiplatelet therapy is recommended, especially in patients with distal coronary artery fistulae and abnormally dilated coronary arteries (Umana et al., 2002). Prophylactic measures for subacute bacterial endocarditis are recommended, as bacterial endocarditis is a known complication.

CONCLUSION

Congenital coronary artery abnormalities are rare isolated anomalies that are important to recognize symptomatic patients. Usually, isolated coronary anomalies are asymptomatic; however, certain forms are associated with myocardial ischemia, congestive heart failure, and sudden cardiac death. Recognition of signs and symptoms that may indicate a congenital coronary artery anomaly should lead to additional testing, and a thorough evaluation of coronary artery anatomy using cardiac CT, MRI, and other imaging modalities are necessary.

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