Case report - Coronary
Right coronary artery to superior vena cava fistula presenting with ‘steal’ phenomenon
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Received 19 April 2004; accepted 7 June 2004

Abstract
A coronary arteriovenous fistula is a rare occurrence. We report a case of a right coronary artery to superior vena cava fistula presenting with myocardial ischemia.

Keywords: Coronary arteriovenous communications; Surgery

1. Case report
Coronary arteriovenous fistulae (CAVF) are rare, with a reported incidence of 0.1 to 0.2% [1]. The right coronary artery (RCA) is the most common site of origin and the right ventricle (approximately 40%) and right atrium are the most common draining chambers. We report a case of successful surgical management of an extracardiac RCA to superior vena cava (SVC) fistulous connection.

A 42-year-old man was referred for evaluation of typical angina symptoms. He consulted the emergency room with an episode of substernal chest pain with left arm radiation triggered by exertion and a new onset of atrial fibrillation with spontaneous resolution. He referred to a one-month history of fatigue and dyspnea with moderate exertion. Physical examination was unremarkable and the chest radiography and electrocardiogram were negative. A Cardiolyte nuclear exam showed equivocal reversible antero-septal myocardial ischemia. A cardiac catheterization followed, and showed the unusual finding of a large, tortuous communication between the RCA draining presumably in the SVC. The RCA had a large and high take off from the aorta and the fistula originated 1 cm from the common RCA trunk posteriorly and ran lateral to the aorta for approximately 10-cm (Fig. 1).

The patient underwent surgery through a limited (5 cm) upper partial sternotomy. A 0.8 cm diameter tortuous extracardial fistula was identified (Fig. 2). The fistulous conduit lay against the medial aspect of the right atrium, with multiple bends covered by thin serosa. There were several areas of calcification and fibrous thickening. A thrill was palpable. The proximal end was dissected 1 cm distal to the RCA, avoiding a small accessory conal branch, and was occluded to confirm the diagnosis and then ligated. The distal end was dissected and ligated close to the SVC end. The procedure was completed and the patient returned to the intensive care unit. The postoperative course was uneventful and the patient discharged home on postoperative day 4.

2. Comment
Coronary arteriovenous fistulae are rare. Most commonly they involve the right coronary artery (RCA) and drain with variable frequency to the right ventricle (40.3%), right atrium, pulmonary artery, left atrium and occasionally to...
the coronary sinus, bronchial veins or superior vena cava [1]. Associated congenital anomalies occur in 40% of patients. Patients after the third decade usually present with symptoms. Exertional angina, in the absence of coronary artery disease, is a common finding in older patients and presumably occurs due to coronary flow ‘steal’. In addition, CAVFs draining into right heart chambers may produce significant right to left shunt and develop in congestive heart failure. Endocarditis, aneurismal dilatation and rupture have been reported as well [2,3].

The management of the asymptomatic fistula is controversial; however most authors agree that the presence of a significant shunt or aneurysmal dilatation justifies the closure of it [1]. Symptomatic CAVFs should be closed or resected. Surgical closure, with or without cardiopulmonary bypass (CPB), has been the gold standard [4]. The reported success rate is high and the operative morbidity is very low. The fistulae that are intracameral, short, close to a critical coronary supply or associated with an aneurysm are frequently closed with the use of CPB. On the contrary, CAVFs that are extracameral and anatomically accessible are usually controlled with ligature without the use of CPB. The use of percutaneous transcatheter closure devices has been successfully described and has been increasingly used, especially in the pediatric population [5].

The rarity of this case was the presence of a long tortuous extracameral fistula voiding into the superior venous cava that was readily accessible through a less invasive localized exposure. The presence of calcification, near common take off with the RCA and the unclear site of drainage decided the surgical approach.

References