Case report - Congenital

Saccular aneurysm of congenital coronary arteriovenous fistula

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Abstract

A rare aneurysm of a congenital coronary arteriovenous fistula is reported. Echocardiography showed a dilated left coronary and left circumflex artery with normal left anterior descending artery. A dilated structure near the crux was confirmed to be a large aneurysm of circumflex coronary artery on selective angiography. It drained into the posterior right atrial wall. Under cardiopulmonary bypass, the drainage site was identified and directly closed from within the right atrium. Fistula was ligated at origin. The diagnostic and different treatment modalities are reviewed.

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1. Introduction

Aneurysm of a congenital coronary artery fistula (CAVF) is very rare. A case of aneurysm of CAVF of left circumflex coronary artery draining to the right atrium is reported.

2. Case report

2.1. Patient

A 30-year-old man had a right parasternal continuous murmur. Physical examination and ECG were normal. Transthoracic echocardiography showed a dilated left coronary artery. The left circumflex artery was dilated with the left anterior descending artery of normal caliber. A dilated structure near the crux and a turbulent color flow in the right atrium were seen. Selective arteriography showed a dilated left coronary artery, bifurcating into a left anterior descending artery and a dilated circumflex artery. The circumflex artery coursed along the left atrioventricular groove and formed a huge aneurysm near the crux. This was seen in the left lateral (Fig. 1) and frontal (Fig. 2) views. The saccular aneurysm was \(4.2 \times 5.3\) cm in size. This immediately drained into the posterior right atrial wall. Right coronary artery was dominant and normal. Right-sided pressures were normal. Left-to-right shunt was 2:1. Workup for connective tissue disease or arteritis was negative.

2.2. Treatment

During surgery, a large left coronary artery was seen bifurcating into left anterior and circumflex arteries. The circumflex branch passed posterior into the atrioventricular groove and gave rise to the aneurysm. The aneurysm opened into the posterolateral wall of right atrium. The course of left anterior descending artery was normal. Under cardiopulmonary bypass, the drainage site was identified and directly closed from within the right atrium. Ligation of CAVF at the origin was also done. The patient was free of symptoms at follow-up.

3. Discussion

3.1. Coronary artery fistula

This is a rare congenital anomaly and occurs in 0.2–0.4\% of congenital heart diseases [1]. The coronary artery communicates with a right-sided chamber, systemic...
vein or pulmonary artery. Commonest site of origin is right coronary artery and of drainage is right ventricle [2]. Ninety percent of them drain into the right side of heart. Twenty-five percent drain into the right atrium. About 40% of all CAVF arise from left coronary artery. Aneurysm occurs in 19% of CAVF [3]; but achievement of the size, which our patient did, is very rare.

3.2. Diagnosis

Selective coronary angiography is the best method to image coronary artery aneurysm and fistula [1]. Transesophageal echocardiography (TEE) is complementary in the diagnosis. With its better acoustic windows, TEE allows superior imaging; only TEE could identify the drainage site in some cases. TEE could guide surgical management of a patient with failed previous surgery, identifying the precise location of drainage site [4].

3.3. Complications

These generally occur in large fistulae. They are left-to-right shunt and pulmonary hypertension or congestive heart failure and myocardial ischemia. Ischemia may be due to coronary steal [1,2] or spasm. Steal occurs as blood is diverted from the high resistance myocardial capillary bed to the low-resistance fistula [2]. Rupture and infective endocarditis have been reported rarely.

3.4. Indications for closure

Symptomatic patients have angina or volume overload due to left-to-right shunt. One-fifth of patients <20 years and two-thirds of those >20 years are symptomatic [1]. CAVF tend to grow with age. Hence it is recommended to close them early in symptomatic patients or in asymptomatic patients with continuous murmurs or systolic murmurs with early diastolic component [1]. Symptoms in small children may be misleading and clinical, electrocardiographic and radiological abnormalities have been suggested to guide therapy [5]. Asymptomatic patients with no findings have no unanimity with regard to treatment strategy [5]. Some authors offer surgery to asymptomatic patients because of the risk of complications [6].

3.5. Surgery

Surgical closure of aneurysm is done both at the origin and drainage site; cardiopulmonary bypass is required for inaccessible lesions or for those patients with co-existing cardiac lesions that need tackling [3]. But bypass would not facilitate exposure to more anterior drainage sites and TEE guided closure without bypass is possible [7]. Intraoperative TEE is useful for: (a) localization of drainage site, (b) identify feeding vessels, (c) document abolition of flow (all without need for cardiopulmonary bypass) and (d) continuous monitoring of ventricular function to detect ischemic effects of ligation [7].

Since a persistent tract can lead to a larger run-off with time, ligation should be complete. Further, it should abolish only the drainage tract and not interfere with distal coronary flow. If drainage is to a distal aspect of a vascular arcade, then coronary flow is not much compromised. If it is proximal, ischemia or incomplete abolition is possible [7]. Because of the large size of the fistula in our patient, surgical closure was done. Ligation was done at the distal low-pressure end to reduce chances of compromised flow in the feeding artery. Surgery requires median sternotomy and cardiopulmonary bypass to open the chambers to define the drainage and ligate the point of entry [8]. Mortality is low,
but the risks are not negligible [8]. Ischemia, thrombosis of parent coronary artery, rupture of fistula and ventricular fibrillation have been reported [8].

3.6. Percutaneous closure

Coil occlusion is an alternate option [9]. It is not always successful and embolization of coil and transient arrhythmia are reported [9]. Factors favoring coil closure are: (a) ability to cannulate the feeding branch adequately and safely, (b) absence of large branches that may be compromised, (c) presence of single narrow, restrictive drainage site and (d) absence of multiple communications. In a retrospective angiographic analysis, only 6 of 16 (37%) successfully operated patients were thought to be amenable to coil occlusion [5]. This figure is similar to the success result in larger coil occlusion series [3]. The reasons were adjacent vessels at risk, multiple communications, small age that limited safe access to the feeding vessel and large size of the fistula. The advent of Amplatzer occluder may widen this net [10].

Echocardiographic follow-up is required, although CAVF have not recurred after surgical or coil closure.

The case is reported to highlight the rare occurrence of an aneurysm of CAVF and discuss the different modes of treatment.

References