CORONARY ARTERIOVENOUS FISTULA PRESENTATION IN ADULT LIFE: CASE REPORT AND REVIEW OF LITERATURE

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CORONARY ARTERIOVENOUS FISTULA results from the persistence of embryonal sinusoids in the myocardium. Coronary arteriovenous fistula is a rare congenital cardiac defect. It is usually discovered incidentally during coronary angiography but occasionally it presents in adult life with severe symptoms.1,4,5

Case Report

A 45-year-old male Yemeni patient had complained of exertional dyspnea and typical angina for seven years. Six years previously he had been evaluated by a cardiologist who discovered the presence of a machinery murmur on the pulmonary area, and the presence of bilateral pulmonary plethora on a chest roentgenogram. Echocardiographic and Doppler study revealed the presence of moderate enlargement of cardiac chambers, aneurysmal dilatation of the left main coronary artery, the presence of a single large left coronary-right-atrial fistula, with shunt flow from left to right, Qp/Qs on 5:1, estimated pulmonary artery systolic pressure of 45 mmHg, and a left ventricular ejection fraction of 61% (Figure 1).

On admission, the patient presented with an attack of severe chest pain, associated with shortness of breath and palpitation. On examination, collapsing pulse, blood pressure of 130/40, elevated jugular venous pressure, and a continuous murmur on the pulmonary were found. Electrocardiography revealed the presence of P-mitral, left axis deviation, non-specific ST-T wave changes, and left ventricular hypertrophy by voltage criteria. Chest roentgenogram showed cardiomegaly (mainly right ventricular), prominent pulmonary artery, and increased pulmonary vascular markings (Figure 2). Cardiac enzymes were normal and repeat echo-doppler study confirmed the previous findings and additionally revealed that the coronary sinus is hugely dilated. The mitral valve was thickened, and there was mild-moderate mitral regurgite.

Cardiac catheterization demonstrated the presence of an aneurysmal fistula between the left coronary artery near the origin of left anterior descending coronary artery. The track of the fistula was tortuous and ending into the coronary sinus, the right coronary artery (RC) was dominant, the septal collaterals of RC fill retrograde the left anterior descending coronary artery, and oxygen step-up of 8 at mid-right-atrium. During his hospital stay, the patient received antianginal medications in the form of intravenous nitroglycerine,

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Figure 1. Two-dimensional echocardiogram showing an aneurysmally dilated left main coronary stem (LCA).
digoxin, and diuretics. He developed intermittent bouts of atrial fibrillation that responded well to digitalization, then sinus rhythm was resumed.

Operation was performed through a median sternotomy, under cardiopulmonary bypass, and bicaval cannulation. The heart was examined before starting the bypass. An aneurysm of 5 cm wide was found adjacent to the pulmonary artery that was extending behind the heart reaching close to the junction between the right atrium and the inferior vena cava. A thrill was palpable at the beginning of the aneurysm near the left coronary artery. A coronary vessel (left anterior descending artery) was found running over the aneurysm sac near its neck. It was possible to pass a ligature around the neck of aneurysm that was snared temporarily for a total period of 30 minutes without any electrocardiographic changes and the thrill disappeared completely: An intraoperative TEE showed no wall motion abnormalities, therefore, the ligature was tied permanently. The vessel that was found running over the fistula sac was also ligated, and bypass was discontinued without difficulty. Postoperative echo-Doppler showed a normal left ventricular dimensions and function, mild hypokinesia of interventricular septum, smaller coronary sinus than the preoperative size, no evidence of shunts, and mild/moderate mitral regurge. The patient was discharged home on the tenth postoperative day.

Ten days later, the patient presented with recurrent chest pain. Electrocardiography showed the presence of anterolateral subendocardial injury. Stress thallium study revealed reversible ischemia in the left anterior descending artery territory. He was started on heparin infusion, and a repeat angiogram eight days later revealed that remnants of the fistula and the LAD were not filling antegradely, but the septal collaterals of the RC were filling it retrogradely. It was decided to perform a redo operation to graft the LAD. Operation was performed the same day, where a saphenous vein graft was used, as the left mammary artery was of poor quality. The patient then had a smooth postoperative period. Postoperative echocardiography and stress thallium were found normal, and he was totally free of symptoms before discharge.

Discussion

Although there is a common belief that coronary artery fistulae are asymptomatic, 55% of these cases are symptomatic at presentation. As the underlying
An innocent murmur is the usual presentation in asymptomatic patients. It is usually a continuous murmur over the site of the fistula communication, a murmur that may be quite similar to that of a patent ductus, a VSD with aortic incompetence or aortopulmonary window.

In our case the left main coronary artery was the feeding artery, however, the most common feeding artery in these cases is usually the right coronary artery. The draining chambers in our case was the coronary sinus; in contrast it is the right ventricle which is the most common site as a draining chamber, much less frequently, the pulmonary artery and the left side of the heart are the draining chambers.2.4.10

Angiography seems to be the key tool in preoperative evaluation of this pathology. Not only does it identify the feeding artery and the draining chamber but also the presence of other associated anomalies and excludes a VSD, and sinus of val salva fistulae. In large fistulae as in our case, an aortic root injection will identify the anatomy clearly but in small ones selective arteriography is needed.

There is general consensus that all large fistulae and those responsible for symptoms, irrespective of their size, should be surgically treated because their natural history is doomed with fatal complications.6.8 The fistulae are best approached by ligating them at their entrance into the draining chamber, however, if the communication is multiple, then the chamber is opened under cardiopulmonary bypass and all connections obliterated. In our case, because the whole sinus was involved in the pathology and because of the difficulty in identifying the exact anastomosis connections, we decided to ligate the fistula at its origin and that, unfortunately, had to include the orifial of the left anterior descending artery. Temporary occlusion was done initially for 20 minutes, no ECG or hemodynamics changes were observed, so a double ligature was tied around the fistula. Unfortunately the patient presented again after discharge with angina and a strongly positive stress thallium was indicative of ischemia in the LAD region, and the LAD was grafted later on as an independent procedure.

The overall survival of these patients seems to be excellent. A 99.1% survival rate was reported in the Texas Heart Institute Series in 1983 to indicate that the overall prognosis was excellent which was updated in 1992 by Fernandes et al.11

In the Duke University operative series, the results were similar with no operative mortality and no report of recurrence 10 years on.10

References