Congestive Heart Failure in a Patient with Giant Aneurysm-like Right Coronary AV Fistula

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Coronary arteriovenous fistula is a rare congenital cardiac anomaly. Symptoms may be mild or absent in young patients, and some are associated with other anatomic anomalies. Angiography remains the gold standard for diagnosis and a guide to therapy, although other noninvasive modalities are increasingly useful. Both surgery and coil embolism are options for correcting this anomaly. The final choice, however, depends on the precise anatomic distribution of the fistula. Here we report a patient who had a giant aneurysm-like right coronary artery resulting from a fistula terminating in the coronary sinus and another fistula arising from the circumflex artery. Surgical ligation was chosen over coil embolism to prevent unintended distal embolism. The patient underwent a successful operation and no longer experienced her previous symptoms.

Key Words: Coronary arteriovenous fistula • Aneurysm-like

CASE REPORT

A 73-year-old Asian woman presented with an 8-year history of fatigue and progressive exertional dyspnea. More recently, she had developed exertional angina. She had a history of hypertension, treated with diuretics and an angiotensin-converting-enzymes inhibitor.

On admission, her blood pressure was 129/67 mmHg, pulse 90/min, and respiratory rate 24/min. Auscultation revealed a loud, continuous grade V/VI murmur at the left midsternal border without radiation. No other obvious abnormalities were demonstrated on physical examination. Electrocardiography revealed left ventricular hypertrophy, and chest x-ray demonstrated cardiomegaly and no increased lung markings observed.
continuous murmur was no longer heard. The patient re-
covered well after surgery and continued to do well
without chest discomfort or dyspnea up to 2 months of
follow-up.

**Figure 1.** Parasternal long axis view (left upper panel) revealed a huge conduit originating from right coronary artery orifice. Color Doppler revealed abnormal flow leaving away from the proximal orifice (right upper panel) with zoom-in detail (left lower panel). Another parasternal short axis view again revealed a huge right coronary orifice with abnormal flow pattern.

**Figure 2.** A huge and tortuous right coronary artery measured up to 30 mm from right anterior oblique view was disclosed by contrast enhancement by a 6 French right Judkin catheter. Distal drainage into the coronary sinus and then filling into the right atrium (white arrow) in a short time was also demonstrated.

**Figure 3.** Another left anterior oblique view still revealed quite a tortuous and dilated right coronary artery by a 6 French right Judkin catheter. Distal drainage into coronary sinus (white arrow) was also demonstrated.
This report describes a patient whose huge coronary arteriovenous fistula (CAVF) had resulted in exaggerated dilatation of the right coronary artery as well as related angina and congestive heart failure. CAVF was first described by Krause\(^1\) in 1865. It comprises about 0.2% to 0.4% of congenital heart disease. CAVF is defined as an abnormal connection between a coronary artery and any of the great cardiac veins entering the heart. CAVF may occur as an isolated anomaly or may be associated with other congenital cardiac anomalies, including patent ductus arteriosus, ventricular or atrial septal defect, and tetralogy of Fallot.\(^2\)

CAVFs arise most commonly from the right coronary artery (60%) and the rest from the left coronary artery (40%) except for a very small percentage arising from both coronary arteries.\(^4\) In addition, most (about 90%) terminate in the right side of the heart, especially in the right ventricle (45%).\(^4\) Other endpoints include the right atrium (25%), pulmonary trunk (15%), coronary sinus (7%), left atrium (5%), and left ventricle (3%). Our patient thus had a rare anomaly on 2 counts, in that there were fistulae involving both right and left coronary arteries and they both terminated in the coronary sinus.

The hemodynamic consequences of CAVFs depend both on their size and the communicating anastomosis. Eighty-percent of asymptomatic patients are younger than 20 years old, while over half of patients older than 20 are symptomatic. Reported symptoms include fatigue, dyspnea, angina, palpitations and arrhythmias of all kinds, including atrial tachycardia, premature ventricular contractions, ventricular tachycardia and conduction disturbances. The symptom of angina in our patient was attributed to a steal phenomenon,\(^3\) since her coronary arteries were otherwise normal under angiogram. Complications and the risk of death secondary to CAVFs increase with age, with some patients eventually developing aneurysms in the abnormal vessels.\(^3\)

Treatment is necessary in all symptomatic patients, and in asymptomatic patients with abnormal findings (e.g. ventricular hypertrophy, cardiomyopathy or abnormalities on the chest x-ray) not attributable to other disorders. Surgery has been shown to be safe and effective for treating CAVF under such circumstances, but controversy remains as to whether it is indicated in asymptomatic individuals.
in whom the anomaly is an incidental finding. Coil embolization has recently been found to be useful in elderly patients and those with proximal fistulas or fistulas that terminate elsewhere than in the normal coronary artery distribution. In our patient, a close proximity between the orifice and the ending of a relative wide fistula had made proper coil placement quite difficult. For one, coil dislodgment or part of the coil protruding into coronary artery might lead to subsequent myocardial infarction. Secondly, since the huge right coronary arteriovenous fistula itself terminated directly into the coronary sinus, a coil might easily become dislodged and thus result in pulmonary embolism. Therefore, surgical ligation was chosen over coil embolism for our patient.

REFERENCES