Bilateral coronary - pulmonary fistulae, diagnosed by transoesophageal echocardiogram

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Abstract

Bilateral coronary artery fistulae to pulmonary artery is a rare condition. We diagnosed this condition by transoesophageal echocardiogram and successfully treated with surgery.

Introduction

A coronary artery fistula (CAF) is a communication between a coronary artery and a chamber of the heart or any segment of the systemic or pulmonary circulation bypassing the myocardial capillary bed. We present an interesting case of bilateral CAFs to pulmonary circulation at rest (shunt fraction calculated by transthoracic echo Qp/Qs = 1:1). There was no increased flow in the pulmonary circulation at rest (shunt fraction calculated by transesophageal echocardiogram to delineate the cause for continuous murmur, which showed a dilated left main coronary artery, draining into the left pulmonary artery (Figure 1). Her right coronary was also dilated and drained into the left pulmonary artery (Figure 2). She had a CT coronary angiogram and a coronary angiogram which confirmed the findings. There was no increased flow in the pulmonary circulation at rest (shunt fraction calculated by transthoracic echocardiogram was normal. We further investigated her with a transesophageal echocardiogram to delineate the cause for continuous murmur, which showed a dilated left main coronary artery, draining into the left pulmonary artery (Figure 1). Her right coronary was also dilated and drained into the left pulmonary artery (Figure 2). She had a CT coronary angiogram and a coronary angiogram which confirmed the findings. There was no increased flow in the pulmonary circulation at rest (shunt fraction calculated by transesophageal echocardiogram was normal.

Case Report

A forty six year old lady was referred to us for evaluation of exertional angina with a positive exercise stress test. Her only cardiac risk factor was her 30 pack years of smoking.

Clinical examination revealed a pulse rate of 70 beats/minute in sinus rhythm. Her BP was 130/70. Jugular venous pressure was not elevated. Her heart sounds were dual and normal. She had a continuous murmur in the 2nd and 3rd left intercostal space. Other systemic examination was unremarkable.

The ECG was in sinus rhythm with a partial right bundle branch block pattern. Her chest x-ray was within normal limits. To evaluate the continuous murmur, she had a transthoracic echocardiogram which was normal. We further investigated her with a transesophageal echocardiogram to delineate the cause for continuous murmur, which showed a dilated left main coronary artery, draining into the left pulmonary artery (Figure 1). Her right coronary was also dilated and drained into the left pulmonary artery (Figure 2). She had a CT coronary angiogram and a coronary angiogram which confirmed the findings. There was no increased flow in the pulmonary circulation at rest (shunt fraction calculated by transesophageal echo Qp/Qs = 1:1). Patient had exertional angina due to coronary steal which was demonstrated by stress test. Stress ECG showed lateral ST depression at 4 minutes with chest pain. Patient was considered for surgical management due to her symptoms and complexity of the fistulae. She underwent surgical ligation of her fistulae using cardiopulmonary bypass which resolved her symptoms.

Discussion

Coronary arteries arise from the root of the aorta and taper progressively as they branch to supply the cardiac parenchyma. A fistula exists if a substantive communication arises that bypasses the myocardial capillary phase and communicates with a low-pressure cardiac cavity (atria or ventricle) or a branch of the systemic or pulmonary systems. Most fistulae arise from the right coronary artery and terminate in the right side of the heart. The most frequent sites of termination, in descending order, are the right ventricle, right atrium, coronary sinus, and pulmonary vasculature. Bilateral coronary fistulae to pulmonary artery are a very rare entity.

Coronary artery fistulae (CAF) causes myocardial stealing or reduction in myocardial blood flow distal to the site of the CAF connection. The coronary vessel attempts to compensate by progressive enlargement of the ostia and feeding artery. Eventually, myocardium beyond the site of the fistula’s origin is at risk of ischemia, which is most frequently evident in association with increased myocardial oxygen demand during exercise or activity. The factors that determine the hemodynamic significance of the fistulous connection include the size of the communication, the resistance of the recipient chamber, and the potential for development of myocardial ischemia.

CAF is suspected following detection of a continuous murmur on routine examination. Site of the CAF murmur is heard lower on the sternal border than usual; thus, the location often is atypical for other causes of continuous murmurs. In addition, the murmur may have an unusual diastolic accentuation, and the continuous murmur of a CAF often peaks in mid-to-late diastole. The differential diagnosis is other causes of continuous murmur such as patent ductus arteriosus (PDA), pulmonary A-V fistulae, ruptured sinus of Valsalva aneurysm, ventricular septal defect with aortic insufficiency, Venous hums or intrathoracic systemic fistulae.

Most children with coronary fistulae are asymptomatic. Fistulae progressively enlarge over time, and complications, such as congestive heart failure (CHF), myocardial infarction, arrhythmias, infectious endocarditis, aneurysm formation, rupture, and death can occur. Spontaneous closure has been rarely reported. Fistula-related symptoms or complications are present in 19% of patients younger than 20 years and in 63% of patients older than 20 years.

Transthoracic or transesophageal echocardiograms may reveal left atrial and left ventricular enlargement as a consequence of significant shunt flow. The feeding coronary artery
often appears enlarged, ectatic, and tortuous with high-volume flow in color-flow imaging at the origin or along the length of the vessel. Multidetector row computed tomography (MDCT) cardiac imaging has provided excellent distal coronary artery and side branch imaging. MRI has been a good alternative for imaging proximal coronary abnormalities, and provided improved anatomic imaging as well as indices of coronary flow and function. Cardiac catheterization remains the modality of choice for imaging the coronary artery and fistulae. Closure of the fistulae can be undertaken by either transcatheter embolization or surgical repair.

Coronary – pulmonary artery fistulas can be ligated without cardiopulmonary bypass. But when the fistula is large or multiple, beside ligation, it can be directly closed from the inside of the pulmonary artery by using cardiopulmonary bypass to prevent recurrence of the fistulae. In our case because of the multiple fistulae we used cardiopulmonary bypass.

Here we presented a patient with angina and examination revealing a continuous murmur due to the rare bilateral coronary artery fistulae draining to pulmonary circulation, successfully diagnosed by transesophageal echo and MDCT and treated by surgical closure.

References

Figure 1. Transesophageal echocardiogram picture showing the left coronary artery draining to pulmonary artery.

Figure 2. Transesophageal echocardiogram picture right coronary artery draining to pulmonary artery.