An Unusual Case of Dual Coronary Artery Fistulas to Main Pulmonary Artery

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Coronary artery fistula is an anomaly in which a coronary artery directly connects to a cardiac chamber or great vessel. Its incidence is around 0.1 to 1% in the adult population. Dual coronary artery fistulas are far less common and their incidence is estimated to be around 5% in patients with this anomaly. Closure of the fistulas is indicated in patients with myocardial ischemia, large left to right shunt, congestive heart failure or other complications. Herein, we report a 64 year-old man with dual coronary artery fistulas presenting with exertional chest pain. The fistulas were initially suspected on transesophageal echocardiogram because of abnormal flow with a mosaic pattern between the left anterior descending and main pulmonary arteries. Selective coronary angiogram confirmed the diagnosis and revealed fistulous connections from the proximal left anterior descending and ostial right coronary arteries to the main pulmonary artery. The patient became symptom-free after surgical closure of the fistulas. (Chang Gung Med J 2002;25:51-5)

Key words: coronary artery fistulas, echocardiography, coronary angiography.

CASE REPORT

A 64 year-old man was admitted to our hospital because of progressive chest tightness and breathlessness for 2 months. He had been well in the past and there was no previous history of chest trauma. The only risk factor for coronary artery disease was advanced age. His chest discomfort was effort-related, and relieved by rest, and was accompanied by diaphoresis and radiation to the bilateral jaws and shoulders. The duration was less than 10 minutes. On examination, the only positive finding was a grade II/VI continuous murmur loudest at the left 2nd intercostal space. His baseline electrocardiogram revealed non-specific ST-T changes. He was then referred for a Treadmill exercise test (Cornell’s protocol) for the possibility of ischemic heart disease. However, it was non-diagnostic because of inadequate exercise load. Transesophageal echocardiography in the short axis view showed abnormal
flow with a mosaic pattern between the left anterior descending artery and the main pulmonary artery (Fig.1A), which was continuous on continuous-wave Doppler (Fig.1B). Under the suspicion of coronary artery fistula, he underwent cardiac catheterization. Coronary angiogram revealed bilateral coronary artery fistulas from the ostial right coronary artery and proximal left anterior descending artery draining into the main pulmonary artery (Fig. 2). Left ventricular function was normal (ejection fraction 78%). No O$_2$ step-up was found during a pull-back tracing of the right heart channel. Due to persistent symptoms even with anti-angina therapy, he was referred to cardiovascular surgery for fistula closure. The orifices of the fistulas were closed with 4-0 prolene via pulmonary arteriotomy under partial cardiopul-

Fig. 1  (A) Transesophageal echocardiogram in the short axis view showing abnormal flow with mosaic patterns on color Doppler between the left anterior descending artery and the main pulmonary artery (arrowhead). Ao = aorta; PA = pulmonary artery, LA = left atrium; RV = right ventricle; RA = right atrium. (B) Continuous-wave Doppler echocardiogram revealing a continuous jet at the drainage site.

Fig. 2  Selective coronary angiograms showing 2 fistulas connecting from the ostial right coronary artery (A) and proximal left anterior descending artery (B) to the main pulmonary artery in a 30-degree right anterior-oblique projection. F = fistula; RCA = right coronary artery; LAD = left anterior descending artery; LCX = left circumflex artery; PA = pulmonary artery.
Coronary artery fistula, a direct connection of the coronary artery with a cardiac chamber or great vessel, may develop due to enlargement of the capillary network during cardiac embryogenesis or main coronary arteries being attached to the pulmonary trunk at the time of separation. The fistulas originate slightly more often from the right than the left coronary artery with only 5% originating from both. The right side of the heart is the drainage site in more than 90% of cases of single coronary artery fistula. Furthermore, the main pulmonary artery is the site of drainage in more than 50% of dual coronary fistulas as was the case in our patient.

It is noteworthy that one fistula originates from ostium of right coronary artery in our patient. Coronary steal phenomenon may be prominent in this situation resulting from proximal location of the fistulas and its bilaterality.

Most patients with coronary artery fistula are asymptomatic at diagnosis and it is often an incidental finding on coronary angiogram. Some patients are referred for cardiac evaluation for a continuous murmur at the site of drainage. But complications do occur and patients may present with heart failure, chest pain, infective endocarditis, arrhythmia, aneurysmal changes or fistula rupture. As found in our patient, coronary steal phenomenon is the main manifestation of fistula. It should be noted that patients with dual coronary artery fistulas presenting with angina pectoris usually have other associated lesions, such as significant stenosis due to coronary atherosclerosis, aortic stenosis, hypertrophic obstructive cardiomyopathy or others, all of which may also cause coronary ischemia. This is the first reported case of dual coronary artery fistulas presenting with exertional chest pain without definite coronary artery stenosis or other associated lesions.

Surgical management of coronary artery fistula is clearly indicated in patients with evidence of myocardial ischemia, a large left to right shunt or congestive heart failure. It has been reported that the fistulas can be repaired without cardiopulmonary bypass with low morbidity and mortality. Recently, successful transcatheter coil embolization of the fistula has also been reported. Some authors advocate fistula closure even in asymptomatic patients for prevention of complications because of the high success rate and low risk of complications. We believe that fistula closure was clearly indicated in this patient because of symptomatic coronary steal. On the other hand, non-surgical modalities such as transcatheter coil embolization of the fistula may be promising. They may become an attractive alternative for surgical intervention after the procedures and devices are standardized.

REFERENCES

11. Wax DF, MaGee AG, Nykanen D, Benson LN. Coil embolization of a coronary artery to pulmonary artery fistula from an antegrade approach. Cathet Cardiovasc
