

A Case of Bilateral Coronary to Pulmonary Artery Fistulas Associated With Severe Aortic Regurgitation

Seung-Jae Lee, MD, Sung-Ho Her, MD, Seung-Won Jin, MD, Jong-Min Lee, MD, Hee-Jeoung Yoon, MD, Hee-Yeon Lee, MD, Hee-Yeon Kim, MD, Gun-Min Kim, MD and Cheol-Hong Park, MD

Division of Cardiology, College of Medicine, The Catholic University of Korea, Daejeon, Korea

ABSTRACT

Coronary artery fistula (CAF) is a rare form of congenital anomalies of the coronary arteries, and this is usually discovered by chance during coronary angiography. However, this type of fistula can cause important coronary morbidity and mortality leading to angina, syncope, congestive heart failure, myocardial infarction and sudden death. Bilateral CAFs are even rarer, and especially when combined with valvular heart disease. The coincidence of CAF with aortic regurgitation is relatively rare and this might sometimes cause myocardial ischemia. We present here a case of bilateral coronary-pulmonary artery fistulas that arose from the first diagonal branch of the left anterior descending artery and the conal branch of the right coronary artery combined with severe aortic regurgitation, and this all caused myocardial ischemia. (**Korean Circ J 2008;38:331-334**)

KEY WORDS: Arteriovenous fistula; Coronary vessels; Aortic regurgitation.

Introduction

Congenital anomalies of the coronary arteries are rare and they have been discovered in 0.67% of the patients who undergo coronary arteriography and in 0.8% of patients at autopsy.^{1,2)} Although rare, they can cause important coronary morbidity and mortality leading to angina, syncope, congestive heart failure, myocardial infarction and sudden death.³⁾

Coronary artery fistula (CAF) is an abnormal communication between an epicardial coronary artery and the cardiac chambers, the major vessels (the vena cavae, sub-pulmonary veins and pulmonary artery) or other structures.^{4,5)} Congenital CAFs joining into the pulmonary artery are rare cardiac anomalies. CAFs arising from two coronary arteries are even rarer, especially when they are combined with valvular heart disease.⁶⁻⁹⁾ Furthermore, a few cases of coronary-pulmonary artery fistula combined with aortic regurgitation have been previously reported.^{6,9)}

To the best of our knowledge, the present report

may be the first Korean case of 55-year-old woman with severe aortic regurgitation and she also had bilateral coronary-pulmonary artery fistulas.

Case

A fifty-five year old female patient was admitted with a squeezing type chest pain of 5 days duration, and this symptom was not effort related. She did not have any atherosclerotic risk factors or any significant past medical history. There was no family history of aortic, collagen, vascular or congenital heart disease.

She had a regular pulse rate of 71 beats/min, a blood pressure of 140/30 mmHg and a respiratory rate 16 breaths/min.

On physical examination, cardiac auscultation revealed 4 to 6 grade diastolic murmurs at the aortic and pulmonary areas radiating to the upper back.

Her electrocardiography demonstrated left ventricular hypertrophy by the voltage. No ST segment and T wave changes were apparent.

The blood chemistries, including the cardiac enzyme and lipid profiles, were within normal limits. On chest X-ray, cardiomegaly was noted (cardiothoracic ratio: 0.6). Both transthoracic echocardiography and transesophageal echocardiography relatively revealed a mildly dilated left ventricle and good regional wall motion with a left ventricular ejection fraction of 65%, but she had se-

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Correspondence: Sung-Ho Her, MD, Division of Cardiology, College of Medicine, The Catholic University of Korea, Daejeon St. Mary's Hospital, 520-2 Daeheung-dong, Jung-gu, Daejeon 301-723, Korea

Tel: 82-42-220-9504, Fax: 82-42-253-9505

E-mail: hhhsungho@naver.com

vere aortic regurgitation with incapacitated, minimally calcified cusps of the aortic valve (Fig. 1). The thallium 201 myocardial perfusion scintigraphy was normal.

On coronary angiography, the bilateral CAFs arising from the first diagonal branch of the left anterior descending artery and the conal branch of the right coronary artery were detected. Both CAFs drained into the pulmonary artery (Fig. 2). There was no obstructive lesion in all of the coronary arteries and the spasm studies were negative. On the aortogram, severe aortic regurgitation was detected and the shunt ratio (Q_p/Q_s) was 1.15 (Fig. 3).

Since aortic valve replacement was required in this case, we recommended CAF ligation during valvular surgery. Yet she wanted to be operated on in the other hospital, and she was then transferred. When we called

her by the telephone, she was being treated with medical therapy in the other hospital.

Discussion

The embryogenesis of CAF is uncertain. There are six coronary enlargements in the embryo: three in the developing aorta and three in the developing pulmonary artery. These enlargements normally involute, except for the two from the right and left aortic sinuses. A coronary to pulmonary artery fistula may result from persistence of one or more of the pulmonary arterial enlargements, hence the term accessory coronary artery.¹⁰⁾ The relatively high incidence of bilateral CAFs to the pulmonary artery is in accord with this theory.¹⁰⁾ Our case also showed that the bilateral CAFs drained into

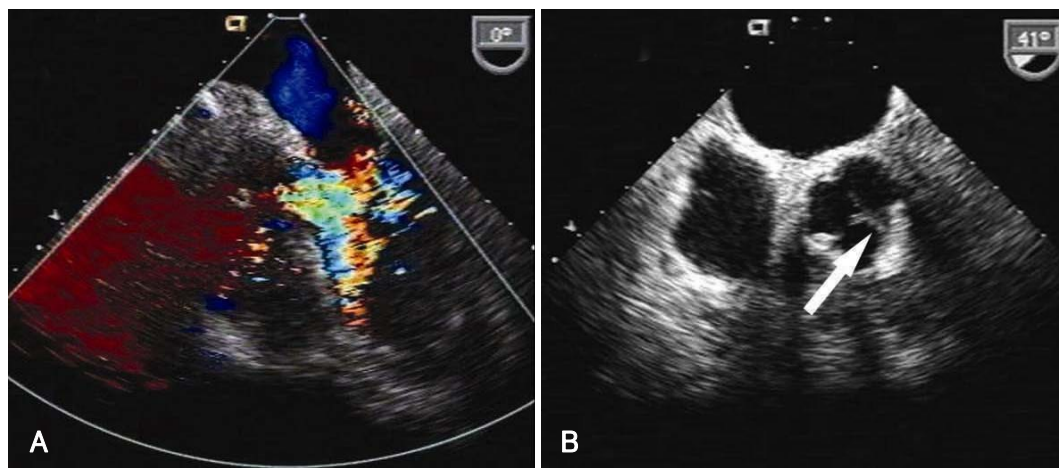


Fig. 1. The aortic valve revealed by the color flow and 2-dimensional echocardiogram of the TEE. TEE shows severe aortic regurgitation (A) and incapacitated cusps (white arrow) of the aortic valve during the diastolic phase (B). TEE: transesophageal echocardiography.

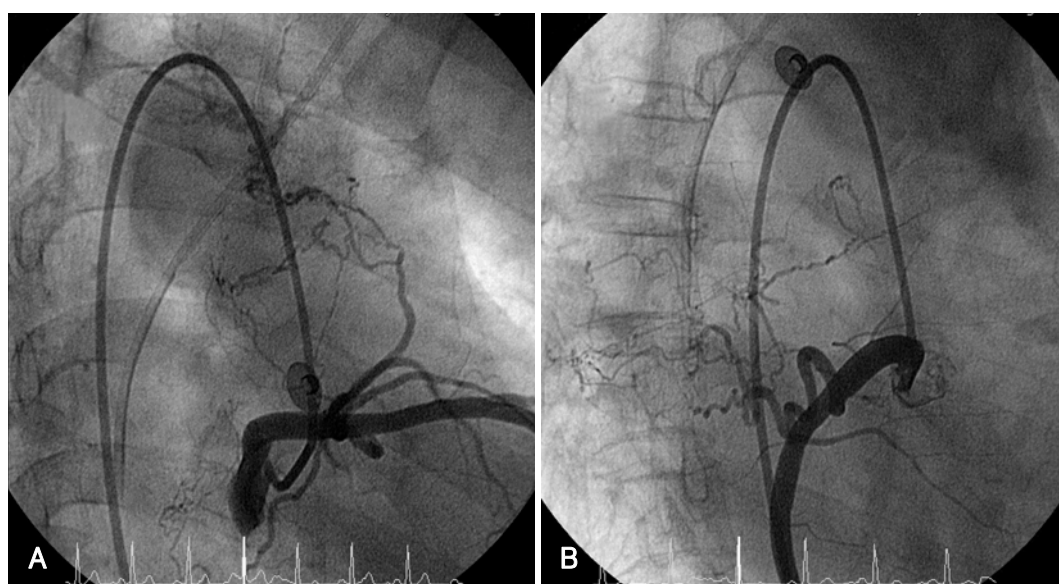


Fig. 2. The left coronary angiogram and right coronary angiogram revealed by the 6 Fr Judkins catheter. Coronary angiography shows that the bilateral coronary pulmonary artery fistulas arise from the first diagonal branch of the left anterior descending artery (A) and the conal branch of the right coronary artery (B).

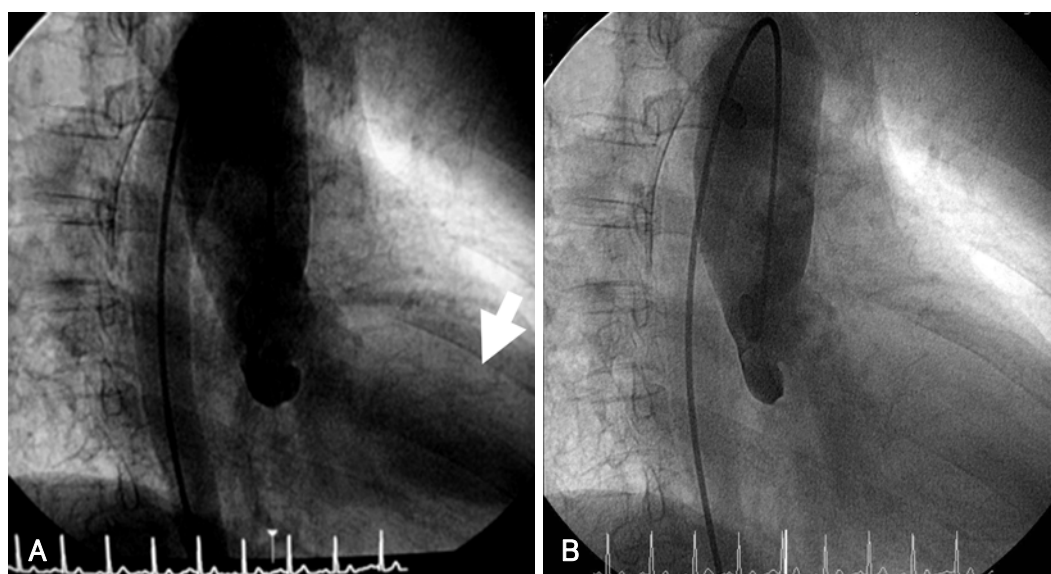


Fig. 3. The aortogram revealed by the pig-tail catheter. The aortogram showed that aortic regurgitant flow reached to the left ventricular apex via backflow (white arrow) of the aortic valve during the diastolic phase (A), but any backflow was not visualized in the left ventricle during the systolic phase (B).

the pulmonary artery, and the pulmonary-systemic flow ratio was 1.15.

CAFs are usually discovered by chance at coronary angiography. In addition, they can be diagnosed during the evaluation of a continuous murmur at the precordium, which is atypical for patent ductus arteriosus, myocardial ischemia and congestive heart failure, and this is rarely found during investigating the etiology of bacterial endocarditis.¹¹⁾¹²⁾ The most frequent complaints of patients with coronary artery fistula are angina pectoris, atypical chest pain and lethargy. The pathophysiologic mechanisms of the symptoms are volume overload as a result of the shunt, coronary steal that causes a decreased myocardial oxygen supply and the lack of capillary formation.¹²⁾ However, small fistulas, and especially those that drain into the main pulmonary artery and the left ventricle, are usually benign. A pulmonary-systemic flow ratio exceeding 1.5 : 1 is recommended as a basic criterion for intervention, but aneurysmal degeneration, coronary rupture or side-branch obstruction may also necessitate surgery.¹³⁾ In addition, the first manifestation may be sudden death due to myocardial infarction or rupture of its aneurysm or a coronary aneurysm. Moreover, endocarditis, intra-cardiac shunt and dyspnea can be observed.¹⁴⁾¹⁵⁾

CAFs can be associated with valvular heart disease.⁵⁻⁹⁾ Several cases of coronary to pulmonary artery fistula combined with valvular heart disease, principally mitral stenosis, have been reported.⁶⁻⁸⁾ CAFs combined with severe aortic regurgitation are relatively rare in adults.⁷⁾⁹⁾ To the best of our knowledge, this is the first Korean case of an adult who had bilateral coronary-pulmonary artery fistulas associated with severe non-rheumatic aortic regurgitation. Although CAFs can cause myo-

cardial ischemia by the mechanism of coronary steal, the coexistence of severe aortic regurgitation might augment the myocardial ischemia. Most of these cases that showed valvular heart disease with the coronary artery fistula were not treated with artificial valves, but the fistula was surgically occluded and the patients revealed a good prognosis.⁵⁻⁹⁾ Therefore, if cardiac pathologies with the concomitant coronary artery fistula require surgical treatment, then we recommend the fistula be surgically occluded, as was done in our case with severe aortic regurgitation.

REFERENCES

- 1) Reul RM, Cooley DA, Hallman GL, Reul GJ. *Surgical treatment of coronary artery anomalies report of a 37.5: year experience at the Texas Heart Institute. Tex Heart Inst J* 2002;29:299-307.
- 2) Alexander RW, Griffith GC. *Anomalies of the coronary arteries and their clinical significance. Circulation* 1956;14:800-5.
- 3) Taylor AJ, Rogan KM, Virmani R. *Sudden cardiac death associated with isolated congenital coronary artery anomalies. J Am Coll Cardiol* 1992;20:640-7.
- 4) Waller BF. *Nonatherosclerotic coronary heart disease. In: Fuster W, Alexander RW, O'Rourke RA editors. The Heart. 11th ed. New York: McGraw Hill; 2004. p.1180-1.*
- 5) Choi JJ, Park CG, Choi CW, et al. *A case of coronary arteriovenous fistula from diagonal branch of left anterior descending coronary artery to left ventricle. Korean Circ J* 2002;32:438-41.
- 6) Gunduz H, Akdemir R, Binak E, Ayarcan Y, Kurtoglu N, Uyan C. *A case of coronary artery fistula with mitral stenosis. Int J Cardiovasc Imaging* 2003;19:533-6.
- 7) Iida H, Sugita Y, Okamura Y, Mochizuki Y, Mori H, Shimada K. *Three cases of coronary to pulmonary arterial fistulae combined with valvular disease. Kyobu Geka* 1996;49:384-7.
- 8) Lin CL, Chen YF, Chiu CC, Voon WC, Lee CS, Lin CC. *Coexistent mitral stenosis and coronary artery fistula presenting as myocardial ischemia: case report. Kaohsiung J Med Sci* 1997;13:117-22.
- 9) Kono T, Kitahara H, Kuroda H, Amano J. *Coronary arteriovenous*

- fistula with annuloaortic ectasia and aortic regurgitation. Jpn J Thorac Cardiovasc Surg* 2001;49:135-7.
- 10) Perloff JK. Congenital coronary artery fistula. In: Perloff JK editor. *Clinical Recognition of Congenital Heart Disease*. 5th ed. Philadelphia: Saunders; 2003. p. 443-5.
 - 11) Kim MS, Ahn YK, Bae Y, et al. Clinical characteristics of coronary arteriovenous fistula in Korean adults. *Korean Circ J* 1997;27: 900-6.
 - 12) Sapin P, Frantz E, Jain A, Nichols TC, Dehmer GJ. Coronary artery fistula: an abnormality affecting all age groups. *Medicine* 1990;69:101-13.
 - 13) Angelini P, Velasco JA, Flamm S. Coronary anomalies: incidence, pathophysiology, and clinical relevance. *Circulation* 2002; 105:2449-54.
 - 14) Rajs J, Brodin LA, Hertfeld I, Larsen FF. Death related to coronary artery fistula after rupture of an aneurysm to the coronary sinus. *Am J Forensic Med Pathol* 2001;22:58-61.
 - 15) Hobbs RE, Millit HD, Raghavan PV, Moodie DS, Sheldon WC. Coronary artery fistula: a 10 year review. *Cleve Clin Q* 1982;49: 191-7.