Case Report

Congenital Systemic and Coronary-to-Pulmonary Artery Fistulas

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Congenital systemic-to-pulmonary artery fistulas are very rare, with the exception of congenital heart disease and pulmonary sequestration. We describe the first reported case of left internal mammary and bronchial artery to pulmonary artery fistulas associated with bilateral coronary arteries to pulmonary artery fistulas. (Ann Thorac Cardiovasc Surg 2010; 16: 203–206)

Key words: systemic-to-pulmonary artery fistula, coronary-to-pulmonary artery fistula, multidetector-computed tomography

Introduction

Systemic-to-pulmonary artery fistulas (S-PAFs) are very rare, with the exception of congenital heart disease and pulmonary sequestration.[1)] We treated a patient who had systemic and coronary arteries to pulmonary artery fistulas (C-PAFs) fed by the left internal mammary artery (LIMA), the bronchial artery (BA), and both coronary arteries.

Case

A 52-year-old-man was admitted to our hospital for the evaluation of a continuous heart murmur and progressive dyspnea on effort. He had no history of trauma or lung disease and had not undergone coronary artery bypass grafting (CABG). Transthoracic echocardiography revealed abnormal blood flow into the main pulmonary trunk and anterior left ventricular wall hypokinesis. Cardiac catheterizations confirmed the presence of a left-to-right shunt of 26% at the main pulmonary artery with a pulmonary-to-systemic flow ratio of 1.40:1.00. The pulmonary artery pressure was 31/16 mmHg, the mean pulmonary capillary wedge pressure was 17 mmHg, and the right atrial pressure was 13 mmHg. Tecnetium-43 myocardial scintigraphy showed a decrease in ejection fraction from 53% at rest to 46% at exercise. Selective coronary angiography demonstrated a bilateral C-PAF and poor visualization of the left anterior descending artery. Furthermore, multidetector computed tomography (MD-CT) (Fig. 1, A, B, and C) showed a bilateral C-PAF and S-PAFs from LIMA and BA into the pulmonary artery.

Operative procedure was performed through median sternotomy (Figs. 2, A and B). The large left C-PAF and small right C-PAF were visible on the surface of the main pulmonary trunk and were dissected up to their origins. The LIMA-to-pulmonary artery fistula was identified at the same site and was exposed at the level of the third intercostal space. The patient was then placed on a standard cardiopulmonary bypass. The main pulmonary trunk was incised longitudinally, and the common orifice of the fistulas was found in the left sinus of Valsalva. The fistula was ligated both proximally and distally. The orifice of the fistula was closed by suturing the wall of fistulas from the outer aspect of the pulmonary artery.

Postoperative selective angiography and MD-CT (Fig. 3, A, B, and C) revealed occlusion of all fistulas. Cardiac catheterization revealed a left-to-right shunt of 6.4% at the main pulmonary artery with the pulmonary-to-systemic flow ratio of 1.10:1.00, and improvement of the pulmonary artery pressure (18/5 mmHg), the mean pulmonary capillary wedge

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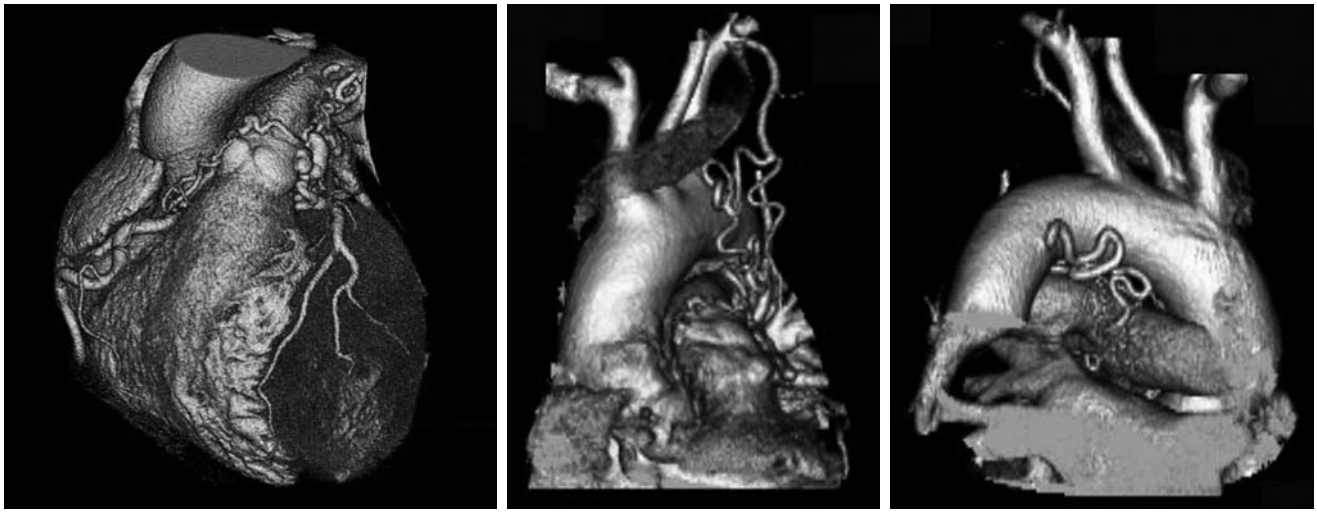

Fig. 1. Multidetector computed tomography shows a bilateral coronary-to-pulmonary artery fistula (**A**) and systemic-to-pulmonary artery fistula (arrow) from the left internal mammary artery and the bronchial artery into the pulmonary artery (**B** and **C**).
LAD, left anterior descending artery; Lt. C-PAF, left coronary-to-pulmonary artery fistula; RCA, right coronary artery; Rt. C-PAF, right coronary-to-pulmonary artery fistula; LIMA, left internal mammary artery; BA, bronchial artery.

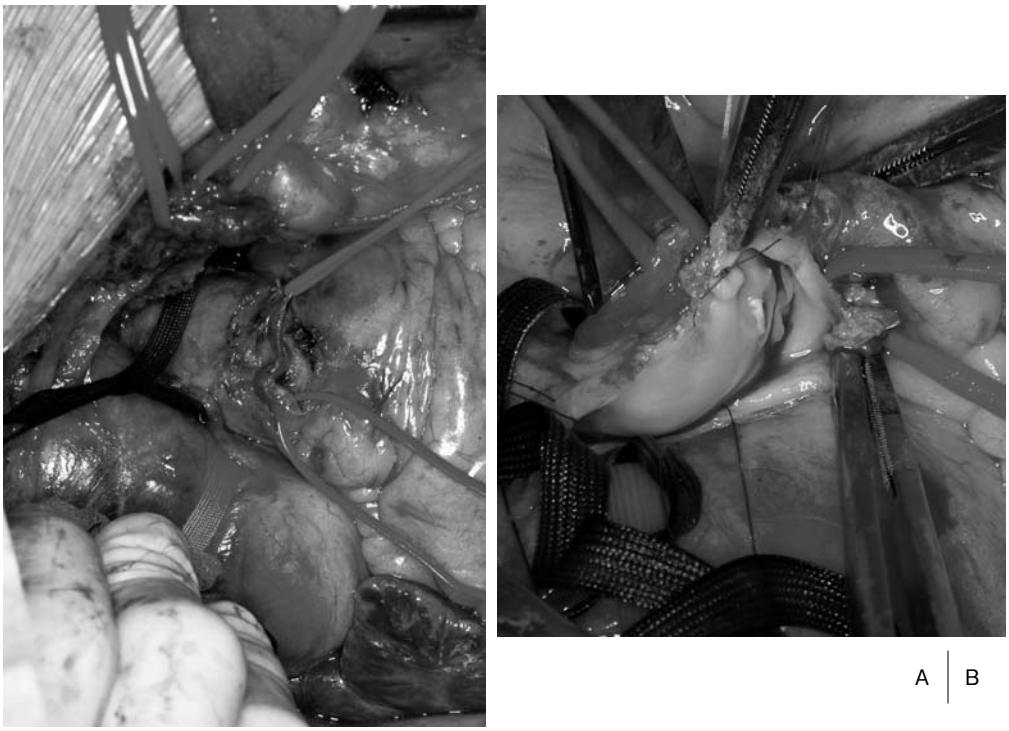

Fig. 2. Operative photograph demonatrates both coronary and left internal mammary artery-to-pulmonary artery fistulas (**A**). The common orifice of the fistulas can be seen in the main pulmonary trunk (arrow) (**B**).
LIMA, left internal mammary artery; PAF, pulmonary artery fistula; Rt. C-PAF, right coronary-to-pulmonary artery fistula; Lt. C-PAF, left coronary-to-pulmonary artery fistula; As-Ao, ascending aorta; PA, pulmonary artery.

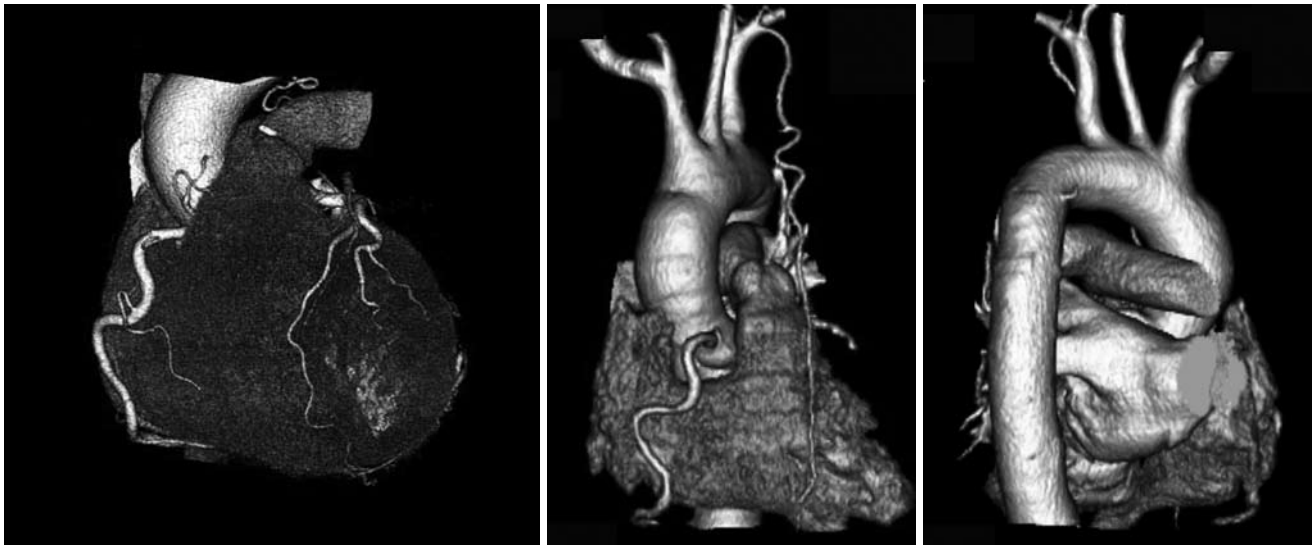

Fig. 3. Postoperative multidetector computed tomography (**A**, **B**, and **C**) show occlusion of all fistulas.
LAD, left anterior descending artery; RCA, right coronary artery; LIMA, left internal mammary artery; BA, bronchial artery.

pressure (4 mmHg), and the right atrial pressure (2 mmHg). Transthoracic echocardiography revealed no abnormal blood flow into the main pulmonary trunk and normal anterior left ventricular wall motion, and tecnetium-43 myocardial scintigraphy demonstrated no reduction of ejection fraction at exercise. The patient recovered with no complications and was discharged on the 19th postoperative day.

Discussion

Congenital systemic-to-pulmonary artery fistulas are very rare, except for pulmonary sequestration and congenital heart disease.[1,2] Among them, only 21 cases of LIMA to pulmonary artery fistula have been reported.[3] Meanwhile, C-PAF occurs in 0.15% to 0.60% of the population, and bilateral C-PAF constitutes about 5% of them.[4] Our patient, to the best of our knowledge, is the first case of BA and LIMA to pulmonary artery fistulas associated with bilateral coronary arteries to pulmonary artery fistulas.

The etiology of fistulas is classified as acquired, e.g., infection, trauma, neoplasm, and surgery, or congenital. Our patient had no history of any disease that could have led to such fistula. Therefore the fistulas in this case may be thought to be congenital. Although indications for the treatment of fistulas have not been established because their natural course is unknown,[5] several authors recommended that intervention was necessary for asymptomatic patients, not to mention for symptomatic patients, being in danger of endocarditis, angina pectoris, and congestive heart failure.[6]

The selection of therapeutic option is controversial. Transcatheter embolization has been reported, and results have improved recently.[7] However, the anatomy and characteristics of some fistulas do not allow catherter intervention. Although several authors reported that fistulas can successfully be ligated without extracorporeal circulation,[4] surgical intervention with cardiopulmonary bypass was applied in our patient because recurrence may occur in simple ligation cases. As described by Huang and colleagues,[8] we also recommend that exploration of the pulmonary artery be carried out for complete closure of the orifice of the fistula and to prevent recurrence. Postoperative selective angiography and MD-CT revealed occlusion of the fistula fed by BA, though closure of bronchial artery-to-pulmonary artery fistula was not undertaken in this patient. This suggests that a complete closure of fistula orifice led to an occlusion of the bronchial artery-to-pulmonary artery fistula.

Recent advances in diagnostic modality, e.g., MD-CT, might make it possible to detect systemic-to-pulmonary artery fistulas, and it may be expected that the natural course, prognosis, therapeutic indication, and optimal treatment method in such cases will be better understood.

Conclusion

In summary, we experienced successful treatment of S-PAFs associated with bilateral C-PAFs. We recommend pulmonary artery exploration using cardiopulmonary bypass to prevent a recurrence of fistulas in such complex cases.

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