RIGHT ATRIAL MYXOMA WITH RIGHT TO LEFT SHUNT
AND CORONARY ARTERY DISEASE

HIROFUMI SAITO, M.D., HIROSHI KUBOTA, M.D., MIKA TAKESHITA, M.D.
AKIRA MIZUNO, M.D. AND MASARU SUZUKI, M.D.

Surgical treatment of a right atrial myxoma with a right-to-left shunt and coronary artery disease was successfully performed in a 61-year-old man. The interatrial shunt occurred through a patent foramen ovale and was the result of a high central venous pressure due to tricuspid stenosis and regurgitation. Instead of right heart catheterization and cardioangiography, we chose an intraoperative blood-gas study to diagnose the intracardiac shunt. Preoperative polycythemia and hypoxemia were improved after removal of the tumor and closure of the patent foramen ovale. (Jpn Circ J 1994; 58: 76-79)

RIGHT atrial myxoma is a relatively rare lesion that comprises 20 to 25 percent of all cardiac myxomas.1,2 It is very unusual to find myxomas in the right atrium associated with right-to-left shunt, which presents clinically as central cyanosis. This paper describes such a case which is also unusual because of the concurrent presence of coronary artery disease.

CASE REPORT

A 61-year-old man was admitted for further examination of accidentally recognized cyanosis. There was a history of cerebral infarction in 1984. In October of 1990, he was treated for acute myocardial infarction, and an abnormal mass was echocardiographically suspected in the right atrium. At that time he refused any other diagnostic procedures. On physical examination, the blood pressure was 150/90 mmHg, and the pulse rate was 90 beats/min and regular. The lips and finger nails were remarkably cyanotic, but no clubbing was observed. The neck veins were distended. A grade 2/6 systolic murmur was heard along the left sternal border. The liver was felt 2 cm below the right costal margin. Laboratory data revealed polycythemia with a hemoglobin level of 21.8 g/dl, a hematocrit value of 66.5%, and a red blood cell count of 712 x 10^6/mm^3. The serum total bilirubin value was 1.6 mg/dl. All other routine laboratory data were within normal limits. An arterial blood study that was performed while the patient breathed room air showed marked hypoxemia with an oxygen tension (PO_2) of 37.7 mmHg and oxygen saturation (SO_2) of 75.2%. After breathing 100% oxygen for 30 min, the arterial blood study was repeated and gave slightly improved findings: PO_2 to 50.4 mmHg and SO_2 to 88.8%. Chest roentgenography showed moderate cardiomegaly, mainly of the right side. Electrocardiography demonstrated abnormal Q waves and tall peaked P waves in leads II, III, and aVF. A transesophageal echocardiogram revealed a right atrial mass which prolapsed into the right ventricle during diastole (Fig. 1a). Magnetic resonance imaging confirmed the presence of a right atrial mass attached to the interatrial septum (Fig. 1b).

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Department of Cardiac Surgery and Internal Medicine,* Asahi General Hospital, Chiba, Japan
Mailing address: Hirofumi Saitoh, M.D., Department of Cardiac Surgery, Asahi General Hospital, I-1326 Asahi-shi, Chiba-ken 289-25, Japan

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A selective coronary angiogram demonstrated 75% stenosis in segment 6 of the left anterior descending artery and complete occlusion of the circumflex artery with good bridging collateral flow (Fig. 2). A feeding artery to the tumor arose from the right coronary artery. A normal nuclear lung scan excluded the possibility of pulmonary emboli. Therefore, an intracardiac right-to-left shunt, presumably at the interatrial level, was strongly suspected as the cause of cyanosis and polycythemia. Repetitive two-dimensional and color Doppler echocardiography failed to detect an intracardiac defect or shunt signal. Radionuclide angiography confirmed no early visualization of the ascending aorta. Right heart catheterization and cinecardiography were not carried out for fear that they might cause paradoxical emboli. A pressure study and blood gas analysis were performed during surgery before institution of the cardiopulmonary bypass (Table I). The mean pressure of the superior vena cava was elevated to 20 mmHg, in contrast to the normal main pulmonary arterial pressure of 12/2 (6) mmHg. The presence of an intracardiac right-to-left shunt was demonstrated by normal oxygenation in the right upper pulmonary vein (PO₂ 534.7 mmHg, SO₂ 98.3%) on 100% oxygen while the ascending aorta displayed hypoxemia (PO₂ 76.5 mmHg, SO₂ 94.1%). Under total cardiopulmonary bypass, a large lobulated tumor arising from the interatrial septum above and median to the fossa ovalis was re-
moved with a partial thickness resection of the interatrial septum (Fig. 3a). A patent foramen ovale 1.5×1.2 cm in size (Fig. 3b) was closed with Dacron® patch after enlargement for inspection to the left atrium. A tricuspid annuloplasty was performed because the tricuspid annulus was dilated to the maximal diameter of 45 mm and marked tricuspid regurgitation was recognized when cold saline solution was injected under pressure into the right ventricle. Coronary artery bypass grafting was carried out using a saphenous vein graft. The tumor measured 7.5×6.0×4.0 cm, weighed 80 g, and was pathologically consistent with a myxoma (Fig. 4).

The patient followed an uneventful postoperative course. Three months after surgery, arterial blood gas showed normal oxygenation with a PO₂ of 81.1 mmHg and SO₂ of 96.4% on room air. The red blood cell was 436×10⁴/mm³ with a hemoglobin level of 12.7 g/dl and a hematocrit value of 40.8%.

**DISCUSSION**

The available English literature describes 12 cases of right atrial myxoma associated with an atrial septal defect or a patent foramen ovale that caused a right-to-left shunt. However, we were unable to find a case that showed this combination of lesions with the coexistence of coronary artery disease. As in the present case, as suggested by high pressure in the superior vena cava, right atrial myxoma with an atrial septal defect or a patent foramen ovale may produce an interatrial right-to-left shunt when it disturbs blood flow through the tricuspid valve or stretches the tricuspid annulus. The 12 previous cases included 3 men and 9 women who ranged in age from 7 months to 73 years (average 49.4 years). All of the 11 cases in whom these values were described showed moderate to severe preoperative hypoxemia with average arterial PO₂ and SO₂ of 40.6 mmHg and 74.0%, respectively.
This hypoxemia is characteristic in that it shows no substantial improvement following inhalation of 100% oxygen.14 The interatrial right-to-left shunt was caused by an atrial septal defect in 5 of these earlier cases and by a patent foramen ovale in 7 cases. Paradoxical emboli were observed in 2 patients postoperatively.8,10

Eleven reports confirmed the existence of a myxoma and an interatrial right-to-left shunt using right heart catheterization and cardioangiography. Only the latest report by Lee and coworkers recognized a right atrial myxoma echocardiographically, but no intracardiac shunt was detected.14 Because of new diagnostic modalities, including echocardiography, computed tomography, and magnetic resonance imaging, cardioangiography is not always required to diagnose atrial myxoma. Right heart catheterization and cardioangiography may still be effective for proving the existence of an interatrial right-to-left shunt. However, we chose not to use these procedures in the present case because they can cause paradoxical embolism, and the patient had a history of cerebral infarction and myocardial infarction which could have been due to paradoxical embolism. Interastral right-to-left shunt is sufficiently suggested when a patient without pulmonary disease shows hypoxemia or cyanosis, has right atrial myxoma and shows signs of an elevated right atrial or central venous pressure due to tricuspid stenosis and regurgitation. Right heart catheterization and cardioangiography need not be employed with this combination of lesions.

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REFERENCES

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