Case Reports

Divided Right Atrium Associated with Extensive Coronary Vein Abnormalities

Y. Inoue, T. Tomomasa, Y. Okada, A. Morikawa
Department of Pediatrics, Gunma University School of Medicine, 3-39-15 Showa-machi, Maebashi, Gunma 371-8511, Japan

Abstract. This report describes a case of a divided right atrium associated with coronary vein abnormalities. An 18-year-old woman who had a past history of surgery for repair of a divided right atrium and atrial septal defect developed exertional dyspnea 8 years after the surgery. Selective coronary angiography showed a dilated right coronary artery with a fistulous communication to the right atrium, tortuous coronary veins draining directly into the cardiac chambers, and the absence of the coronary sinus. Embryologically, regression failure of the right venous valve is hypothesized to have created both the divided right atrium and extensive coronary vein abnormalities.

Key words: Cor triatriatum dexter — Coronary sinus — Eustachian valve — Thebesian vein

Although the variability in the morphology and function of the divided right atrium has been well documented [11], no mention has been made of coexisting coronary vein abnormalities either in autopsy cases or in living patients. In this report, we describe an 18-year-old woman with divided right atrium, atrial septal defect, and extensive coexisting coronary vein abnormalities.

Case Report

A 10-year-old girl was first evaluated in 1990 because of a history of cyanosis and dyspnea on exertion. The plain chest x-ray and electrocardiography were normal. Cardiac catheterization showed that the right and left atrial pressures were 7 mm and 6 mmHg, respectively. The right ventricular pressure was 26/0 mmHg, with a pulmonary artery pressure of 22/8 mmHg. There was no step-up in oxygen saturation between the right atrium and the pulmonary artery. The oxygen saturation in the femoral artery was 97% and the calculated $Q_p/Q_s$ was 1.0. A catheter entered the left atrium through an atrial septal defect. Although most of the contrast medium injected into the inferior vena cava passed into the left atrium through the atrial septal defect, contrast medium from the superior vena cava primarily passed into the hypoplastic right ventricle through the tricuspid valve (Fig. 1). Based on the catheterization data, the diagnosis of divided right atrium and atrial septal defect was made. At the time of surgery, a dilated right coronary artery and dilated coronary veins were identified. There was a large second-order type atrial septal defect and a large, thin membranous eustachian valve in the right atrium, which drained blood flow from the lower right atrium into the left atrium. The atrial septal defect was closed directly and the membranous eustachian valve was excised. The right atria cavity was small and was enlarged using a glutaraldehyde-preserved bovine pericardial patch. The patient was weaned from bypass without difficulty and required only minimal inotropic support in the postoperative period. The postoperative course was uneventful and no heart murmurs were audible afterward. She did well and had no symptoms for almost 8 years.

In 1998, she was referred for the evaluation of exertional dyspnea. She did not develop cyanosis on exercise. Physical examination revealed a blood pressure of 100/70 mmHg, a regular pulse of 55 beats per minute, and no audible murmur. The chest radiograph showed mild cardiomegaly. The electrocardiogram showed sinus rhythm with a QRS axis of 120° and anterior T wave inversion. Transthoracic echocardiography showed a markedly dilated ostium of the right coronary artery. Quantitative gated single photon emission tomography (QGS) [2] using $^{99m}$Tc-tetrofosmin showed mildly decreased regional perfusion and decreased regional wall motion in the anterior wall and septum. The QGS-derived left ventricular end diastolic volume, end systolic volume, and ejection fraction at rest were 105 ml, 47 ml, and 55%, respectively. A treadmill exercise stress test was performed using the modified Bruce protocol, during which the patient went 24 seconds into stage 3 with a heart rate of 144 beats per minute. The patient had no chest pain and there were no ST segment changes on the electrocardiogram.

Cardiac catheterization revealed a pulmonary artery pressure of 20/6 mmHg. The femoral artery oxygen saturation was 95%, the pulmonary oxygen saturation was 77%, and the calculated $Q_p/Q_s$ was 1.0. Aortography revealed a dilated right coronary artery. Selective coronary angiography revealed no atherosclerotic changes. However, the right coronary artery was dilated with a fistulous communication to the upper right atrium (Fig. 2A). The left coronary artery was not dilated, but in the very early angiographic phase a majority of part of the blood from the left coronary artery was drained directly into the left ventricle through tortuous cardiac veins (Figs. 2B and 2C). Despite obtaining additional angiographic projections, it was impossible to demonstrate a cor-

Correspondence to: T. Tomomasa
Fig. 1. Preoperative angiogram (anteroposterior view). (A) Contrast medium injected into the inferior vena cava passes into the left atrium through an atrial septal defect. (B) Contrast medium from the superior vena cava passes into the hypoplastic right ventricle through the tricuspid valve.

A coronary sinus or its ostium. A persistent left superior vena cava was not identified.

Discussion

A divided right atrium, or cor triatriatum dexter, is a rare cardiac abnormality in which the right atrium is subdivided into two distinct chambers [11]. This ab-

Fig. 2. Selective coronary angiogram (right anterior oblique view). (A) Selective right coronary angiogram showing a dilated right coronary artery with a fistulous communication to the upper right atrium. (B, C) Selective left coronary angiogram showing tortuous cardiac veins directly draining into the left ventricle.