Injury to the Tricuspid Valve and Membranous Atrioventricular Septum Caused by Huge Calcified Right Ventricular Myxoma —— Report of a Case ——

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A 64-year-old female, admitted because of severe dyspnea on exertion and facial edema, showed echocardiographic findings of a large tumor in the right ventricle (RV). Echocardiography revealed a cardiac mass extending from the RV across the tricuspid valve into the right atrium, synchronized with the cardiac cycle, and severe tricuspid regurgitation was apparent. The mass was removed under cardiopulmonary bypass. It measured 7x5x5 cm with diffuse superficial calcification and arose from the posterior wall of the RV, just under the tricuspid valve ring, with a short pedicle. During the same procedure, after the successful excision of the tumor, small atrial and ventricular septal defects were found that had been caused by the tumor and these were closed directly. The tricuspid valve was repaired with valvuloplasty, chordoplasty and annuloplasty. The microscopic findings were of typical myxoma; however, a right ventricular myxoma protruding into the right atrium is exceedingly rare. (Circ J 2004; 68: 799–801)

Key Words: Atrial septal defect; Right ventricular myxoma; Tricuspid valve regurgitation; Ventricular septal defect

Cardiac myxomas are the most frequent primary tumor of the heart, but being located in the ventricle is extremely infrequent and only approximately 2–5% of myxomas arise from the right ventricle (RV). We describe a case of RV myxoma protruding into the right atrium and evoking severe tricuspid valve regurgitation, as well as atrial and ventricular septal defects (ASD, VSD).

Case Report

A 64-year-old Japanese female was referred to hospital for severe dyspnea on exertion for 2 weeks, facial edema, lower extremity edema and 8 kg weight gain. She had had dyspnea on effort for 2 weeks and was initially admitted to the practitioner in October 2001. At that time she was diagnosed with a RV tumor and was referred to hospital in the same month because of deterioration of her symptoms, which corresponded to New York Heart Association grade III. Physical findings showed a regular pulse of 80 beats/min and her blood pressure was 146/92 mmHg at the time of admission. She had facial edema, jugular venous distension, hepatomegaly, pretibial edema and ascites. Auscultation showed fixed splitting of the second sound with a mild systolic murmur along the right sternal border of the third intercostal space and normal vesicular sounds with no rales. Hematological and biochemical examinations revealed no abnormalities, and tumor marker levels were within normal limits. A chest X-ray revealed an increased cardiothoracic ratio (0.63), pulmonary congestion and pleural effusion. An electrocardiogram revealed a normal sinus rhythm with low voltage R-waves in leads V1–4. Transesophageal echocardiography revealed a huge polypoid tumor mass attached to the posterior wall of the dilated RV with a pedicle just below the tricuspid valve ring. The mass was moving toward the right atrium through the tricuspid valve ring in systole and relocating in the RV in diastole (Fig 1). Color Doppler echocardiography showed massive tricuspid valve regurgitation. The right heart angiogram from the inferior vena cava (IVC) showed a moving shadow in the RV that crossed the tricuspid valve during systole. Pulmonary ventilation and perfusion scintigraphy demonstrated no abnormalities. The family history was unremarkable.

Based on these findings, a RV tumor with massive tricuspid valve regurgitation was diagnosed and the patient underwent surgery. Total cardiopulmonary bypass was instituted with only direct superior vena cava (SVC) cannulation for venous uptake. The aorta was cross-clamped with cardioplegic arrest, and the right atrium was incised longitudinally. Open direct IVC cannulation via the IVC orifice was done to prevent pulmonary tumor embolism by the cannula. The right atrium was occupied by a huge, whitish-yellow polypoid tumor with a calcified surface (Fig 2) that was attached by a short stalk on the free wall of the posterior RV, underneath the posterior leaflet of the tricuspid valve. The tumor was excised together with a cuff of endocardium adjacent to its attachment. The septal and anterior leaflets of the tricuspid valve had been remarkably damaged by collision with the RV tumor, and the main chordae of the medially papillary muscle was ruptured. The destruc-
tion of the perimembranous portion and the anteroseptal comissure located in the other side of the tumor insertion was remarkable, and a small ASD and VSD were found, each of which was closed directly, and the tricuspid valve was repaired by chordoplasty, valvuloplasty and annuloplasty using the DeVega method. The annulus of the tricuspid valve was reduced from 35 mm to 27 mm (Fig 3). The patient was weaned from cardiopulmonary bypass without difficulty and the postoperative course was excellent, with sinus rhythm maintained throughout. The right heart failure was improved noticeably; however, mild tricuspid valve regurgitation was still evident at the postoperative examination.

On gross examination, the tumor was an irregular lobulated, short, pedunculate, gelatinous, diffusely calcified mass measuring 50×50×65 mm (Fig 4). Microscopic examination showed typical myxomatous tissue with general calcification of the surface of the tumor (Fig 5).

Discussion

Myxoma is the most common cardiac tumor and constitute 40% of benign cardiac masses. Ventricular myxomas
appears to be a major risk factor.\textsuperscript{9,10} The present patient did obstruction of both the RV outlet and inlet.\textsuperscript{6} In cases such as the inflow portion of the RV septum, it will result in from the anterior free wall of the RV papillary muscle or between the right atrium and the RV or obstructs RV the posterior free wall of the RV, the RV myxoma moves the present patient in which the RV myxoma projects from between the right atrium and RV or obstructs RV inflow.

There are 2 types of cardiac myxomas, the friable polypoid type and the smooth-surface rounded type! The polypoid type is associated with a higher incidence of embolism because of its obvious fragility\textsuperscript{7,8} and tumor size also appears to be a major risk factor.\textsuperscript{9,10} The present patient did not have evidence of pulmonary embolism in the pre-operative examinations, although the RV myxoma was big and polyloid. Catastrophic embolic complications are known to occur during induction of anesthesia and sternotomy;\textsuperscript{4} one report describes a fragment of the myxoma embolizing around aortic cross-clamp\textsuperscript{8} Because the pre-operative study had shown that the right atrium during systole, we could not to destroy the tumor by fragmentation when the venous cannula was placed via the right atrium during establishment of extracorporeal circulation. In this situation it is best to first cannulate the SVC directly and then the IVC should be cannulated through an internal right atrial exposure under direct vision with suction venous uptake. Using this surgical technique, the operation was performed safely in the present patient without pulmonary tumor embolism.

This case of RV myxoma had severe surface calcification, which is more common in right atrial myxomas\textsuperscript{2} and the continuous friction during ventricular systole between the prolapsing tumor and the right-sided atrioventricular portion, including the tricuspid valve leaflets, must have been responsible for the damage to the leaflets and the septal membranous portion. Damage to aortic, mitral and tricuspid valve leaflets by myxomas has been documented\textsuperscript{11,12} but there are no known reports of myxoma producing tricuspid valve regurgitation, as well as ASD and VSD through injury to the tissue.

References
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