Disappearance of a Cardiac Rhabdomyoma Complicating Congenital Mitral Regurgitation as Observed by Serial Two-Dimensional Echocardiography

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SUMMARY. A cardiac tumor was diagnosed by two-dimensional (2D) echocardiography in a four-month-old infant who also had congenital mitral regurgitation. At 18 months of age, histological examination of a biopsy specimen of the cardiac tumor, obtained during surgery for mitral valve replacement, revealed a rhabdomyoma. In serial postoperative 2D echocardiograms, this tumor decreased in size until total disappearance after 6 months.

KEY WORDS: Cardiac rhabdomyoma — Spontaneous disappearance — Mitral regurgitation — Two-dimensional echocardiography

Most primary cardiac tumors in children are histologically benign, a high percentage being cardiac rhabdomyomas. Although occasional spontaneous disappearance is an interesting characteristic of cardiac rhabdomyoma [6, 7], its cause remains to be clarified. Only a few cases of this tumor have been reported in association with congenital heart disease. Recently, we treated a young child with congenital mitral regurgitation complicated by this tumor and observed the disappearance of the tumor by serial 2D echocardiography.

Case Report

On admission the patient was 4 months old. She had been born at term by spontaneous delivery, weighing 3400 g. About 1 month after birth, stridor developed. After 3 months she was admitted to our department because of tachypnea, poor feeding, and an inadequate increase in body weight. On admission she had a labored respiration at a rate of 60/min. The pulse rate was 116/min. The body weight was then 4.5 kg. A left precordial bulge was noted. Harsh rales and stridor were heard bilaterally over the lower lung. A grade 3/6 systolic regurgitant murmur and a grade 2/6 diastolic rumble were heard at the apex. The second sounds were shortly split, and the pulmonary sound was accentuated. The liver was palpated 3.5 cm below the right subcostal margin. A chest x-ray film showed enlargement of the left atrium, left ventricle, and the pulmonary artery with pulmonary congestion and cardiomegaly (cardiothoracic ratio of 66%). The ECG revealed combined ventricular hypertrophy with more marked left ventricular and left atrial hypertrophy. Two-dimensional echocardiography (Fig. 1A,B) showed marked enlargement of the left atrium and ventricle, a thickened anterior mitral leaflet and its prolapse to the left atrium, as well as hyperkinetic movement of the ventricular septum and left ventricular posterior wall. In the lower ventricular septum, in the long axis view of the left ventricle, and in the left ventricular lateral wall, in the short axis view of the left ventricle, a mass suggestive of a cardiac tumor was observed [9, 10]. Pulsed Doppler echocardiography revealed a systolic reverse flow signal in the left atrium. Based on these findings, a diagnosis of cardiac tumor complicating congenital mitral regurgitation was made.

The patient was treated with digitals and diuretics in the outpatient clinic to observe the course, but was admitted several times when respiratory and cardiac failure worsened. Since growth was also markedly delayed, cardiac catheterization and angiography were performed at the age of 14 months. The pulmonary arterial pressure was 50/20 (mean, 30) mmHg. The v wave and mean pressure of the left atrium were elevated: a wave, 11 mmHg; v wave, 27 mmHg (mean, 20 mmHg). Oxygen saturation was similar throughout the right heart. Left ventriculography (Fig. 2A,B) demonstrated severe mitral regurgitation and marked enlargement of the left atrium and ventricle. There was no ventricular septal defect or persistent ductus arteriosus. A frontal left ventriculogram (Fig. 2A) showed a spherical filling defect near the left ventricular lateral wall, and a lateral ventriculogram (Fig. 2B) revealed a semispherical filling defect protruding into the left ventricular cavity in the middle to lower septum. The filling defect, demonstrated on the frontal image and lateral images suggested a tumor. Thus these ventriculograms showed the same mass as that observed in each of the two echocardiographic views, originating in the left ventricular lateral wall and extending to the ventricular septum. Since cardiac failure persisted, the mitral valve was replaced by a 19-mm St. Jude Medical Valve prosthesis at the age of 18 months, when her body weight was 6.2 kg. An ash-colored nodule, slightly stiffer than

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normal myocardial tissue, was seen at the site of the left ventricu-
lar lateral wall corresponding to the filling defect on the left
ventriculograms. A part of this nodule was resected for histologi-
cal evaluation. This revealed a tumor composed of cells with
large vacuoles (Fig. 3). The cells exhibited a spider-cell picture
typical of cardiac rhabdomyoma, with myocardial fibers running
in a radial and spider-web pattern [5].

The patient’s postoperative course was satisfactory. Serial
postoperative 2D echocardiography revealed reduction in the
tumor echo 2 months after surgery and disappearance of the

Fig. 1. Preoperative 2D echocardiogram. A A long-axis view of the left ventricle. B A short-axis view of the left ventricle. White
arrow, tumor mass; LA, left atrium; LV, left ventricle.

Fig. 2. Left ventriculogram. A Anteroposterior view. B Lateral view. Abbreviations as in Fig. 1.