Mitral valve lipomatous hamartoma infiltrating myocardium

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Primary cardiac neoplasms are very rare. The incidence is reported to be 0.001% to 0.03% in autopsy series. Of these, about 10% are valvular and the majority are benign. Only a few malignant valvular tumors have been described in the literature.¹ Most valvular tumors are small and asymptomatic but may present with a variety of symptoms including arrhythmias, particulate or thrombotic embolizations, valvular stenosis or regurgitation with congestive heart failure, and even sudden death.^{2,3}

Primary valvular lipomatous hamartomas have been described until now in only 9 patients: 5 patients with mitral hamartomas, 3 with tricuspid hamartomas, and 1 with aortic hamartoma.^{4,5}

CLINICAL SUMMARY

Our patient was a 28-year-old healthy woman. She had had 4 uncomplicated pregnancies and during the last had disturbing amount of monofocal premature ventricular contractions without any other symptoms. Subsequent echocardiogram revealed a huge tumor in the left ventricle occupying about 85% of the ventricular cavity. It was fixed to the ventricular side of anterior mitral leaflet and the apex. No cardiac murmurs were heard and no clinical signs of any type of valvular disease could be detected. Catheterization revealed normal coronaries. All measured pressured were within normal limits: left ventricular (LV) ejection fraction was 0.45 and cardiac index was $4.1 \text{ L} \cdot \text{min}^{-1} \cdot \text{m}^{-2}$. Because LV angiogram could not be done, a levophase ventriculogram was performed and revealed the tumor occupying almost the whole LV cavity (Figure 1).

The patient was operated using cardiopulmonary bypass and whole-blood antegrade and retrograde cardioplegia during the crossclamp time. A longitudinal left ventriculotomy was done between the left anterior descending coronary artery and the circumflex coronary artery branches. The tumor was attached to the anterior leaflet with a broad base and fixed into the apical region of the left ventricle. The tumor was removed first from the anterior leaflet but required so

FIGURE 1. Levophase left ventriculogram showing the ventricular tumor.

much of leaflet tissue to be removed that no repair of the valve was possible. Removing the tumor from the apex required removal of a piece of LV wall. A mechanical mitral valve prosthesis was inserted. The patient's recovery was uneventful, and she was discharged home on postoperative day 4. Pathologic examination revealed a lipomatous hamartoma fixed into the mitral valve tissue and myxomatous degeneration of the valve. In the apical region, examination revealed that the lipoma infiltrated myocardium (Figure 2) and had been removed with healthy margin.

The patient is now 9 years postoperative and has had yearly echocardiographic examinations. No recurrences have been noted and the patient has normal functional capacity.

FIGURE 2. Photomicrograph of mature fat cells invading the myocardium.

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DISCUSSION

Intracardiac lipomas usually present with encapsulation and have no potency of infiltrating adjoining tissues. In this case, the tumor had no encapsulation and theoretically was able to infiltrate the myocardium, which it did.

Lipomas are very slow-growing tumors, and this case also shows the enormous adaptive capacity of a normal heart as the patient had no cardiac symptoms except the premature ventricular contractions with a huge tumor in the left ventricle.

We believe this is the first report of a benign cardiac tumor behaving as a malignant tumor infiltrating myocardium.

All valvular tumors should be removed regardless of size or symptoms.¹ In this case, the surgery was curative as the

tumor was completely resected and no recurrences have been seen.

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Fontan completion in patient with pulmonary artery sling associated with hypoplastic left heart syndrome

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Pulmonary artery (PA) sling is an uncommon congenital anomaly in which the left PA arises from the posterior aspect of the right PA, forming a sling around the trachea.¹ Current surgical approach (ie, reimplantation of the left PA via median sternotomy under cardiopulmonary bypass [CPB]), offers favorable early and late outcomes.^{2,3} Although approximately one-third of patients with PA sling have associated cardiac anomalies, only 1 patient reported in late 1950s had single-ventricle anomaly¹; therefore, surgical implications of this entity in patients with functional univentricular heart have never been discussed. We herein describe a patient with asymptomatic PA sling associated with hypoplastic left heart syndrome (HLHS) who had staged Fontan completion.

CLINICAL SUMMARY

A 2-day-old girl with the body weight of 1.9 kg was diagnosed with HLHS with persistent left superior vena cava.

The existence of PA sling was suspected on preoperative echocardiography. No respiratory symptoms were noted. She had modified Norwood procedure using a right ventricle-to-pulmonary artery (RV-PA) shunt at 3 days of age. The left PA originating from the right PA was detected. The main PA was divided and was used for aortic arch reconstruction. The distal side of the RV-PA shunt was anastomosed to the right PA with a noncuffed 5-mm polytetrafluoroethylene graft (Gore-Tex expanded polytetrafluoroethylene graft, W. L. Gore & Associates, Inc, Flagstaff, Ariz). The postoperative course was uneventful. Cardiac catheterization at 4 months of age revealed mean PA pressure of 13 mm Hg in the right PA and 9 mm Hg in the left PA. The right PA was 6 mm in diameter and the left was 3.1 mm in diameter with hypovascularity in its lung field (Figure 1, A). The Nakata index was 163 mm²/m². PA resistance was 2.75 W/U. Bidirectional Glenn (BDG) anastomosis on the right side was performed at 6 months of age through a median sternotomy under CPB. Arterial desaturation persisted after surgery, and we additionally performed BDG on the left through a left thoracotomy, resulting in increase in SpO₂. The RV-PA shunt was left open. Fontan operation was performed at the age of 2 years and 2 months, at an earlier timing than expected, due to a restrictive interatrial communication. The right PA was 7.2 mm in diameter and the left was 6.3 mm in diameter. The Nakata index was 218 mm²/m². The left PA was untreated and the patient has had no respiratory symptoms throughout the staged

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