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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FIGURE 1. A, Pulmonary artery (PA) angiogram before bidirectional Glenn anastomosis showing that the left PA originated from the right PA and formed the sling around the trachea. No stenosis but hypovascularity was observed in the left pulmonary artery. B, Angiogram 2 years after Fontan operation showing reasonable left PA growth.

palliation. No stenosis was found in the left PA on angiography (Figure 1, B), and the nuclear pulmonary perfusion scan showed that the right-to-left perfusion ratio was 2.3 to 1 at the follow-up period of 4 years.

DISCUSSION

This is the first reported patient with PA sling and HLHS who successfully had staged Fontan completion. Because confluence and absence of distortion in the branch pulmonary arteries are essential for successful Fontan completion, PA sling exerts a much greater impact on patients with a single-ventricle physiology than those with a biventricular physiology. There is no question that a symptomatic neonate with PA sling should be repaired at the time of diagnosis even if any kind of cardiac anomalies are associated; however, the fundamental question raised in this patient was whether asymptomatic PA sling should be repaired, and if necessary, the timing of the repair.

PA sling in the present case was untreated for the following reasons. First, this neonate had a small body weight, less than 2.0 kg, which posed technical difficulty as well as risks of late stenosis/occlusion in the implanted left PA. In fact, PA sling repaired in neonatal or early infantile period results in high stenosis or occlusion rate.^{3,4} Second, because the main PA was used for aortic arch reconstruction, there was no appropriate site to reimplant the left PA. Finally, concomitant

Norwood procedure and PA sling repair might result in prolonged CPB and ischemic time, exposing the patient to risks.

Concerns over untreated PA sling in single-ventricle physiology include turbulence or stenosis/occlusion and subsequent energy loss in Fontan circulation; however, the Fontan circulation has successfully been sustained over 4 years with the evidence of reasonable left PA growth and acceptable pulmonary blood flow distribution. In addition, it is possible that the use of RV-PA shunt as well as the bidirectional BDG tends to provide a symmetric growth pattern of branch pulmonary arteries, and the presence of an additional pulmonary blood flow through the RV-PA shunt after BDG might have, in part, played some roles to facilitate left PA growth in this patient.⁵

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