Hemangioma as a rare cause of a neonatal cardiac tumor resulting in inflow obstruction of the tricuspid valve

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Received: 31 August 2010 / Accepted: 2 December 2010 / Published online: 17 December 2010
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Sirs:

Cardiac tumors are rare in children. Prevalence varies between 0.0017 and 0.027% in autopsy studies [1]. Approximately, 90% of the pediatric primary cardiac tumors are benign. Rhabdomyomas are the most common subtype accounting for about 60% while hemangiomas are rare with about 5% [2]. Diagnosis in the neonatal age group may be difficult because of nonspecific symptoms such as murmur, dysrhythmia, hypoxia, or congestive heart failure. Transthoracic echocardiography is the most important diagnostic tool allowing rapid diagnosis and assessment for the need for surgical intervention.

A male infant was born at 36 + 6 weeks of gestation after an uneventful pregnancy by caesarean section because of pathologic cardiotokography. He was noted to have cyanosis, tachypnea, and signs of poor perfusion in the first hour of life. On chest radiograph, an enlarged heart with a prominent right atrium was evident (Fig. 1). Echocardiography revealed a large mass in the right atrium adhering in large parts to the atrial wall causing severe inflow obstruction to the tricuspid valve (Fig. 2). Low cardiac output and metabolic acidosis were resistant to fluid supplementation. Emergent cardiac surgery was performed using cardiopulmonary bypass. The tumor could be removed in toto by peeling it off the atrial wall. Surgical reconstruction of the dilated atrium was performed. The tumor had an extension of $4.5 \times 3.5 \times 2.8$ cm. Histologically, it consisted mostly of blood, lined by a thin strip of tissue that was diagnosed as benign cavernous hemangioma.

The postoperative course was complicated by a multifocal atrial tachycardia resulting in a ventricular heart rate of 270–290/min refractory to medical treatment and cardioversion. Because of severe hemodynamic compromise extracorporeal membrane oxygenation support was required for 5 days, until sufficient heart rate control could be achieved by amiodarone and overdrive pacing. Cardiac function and rhythm normalised in the following weeks. The patient was discharged home without residual arrhythmias and normal cardiac function. Until now, there are no signs of relapse or hemangiomas in other locations.

Hemangiomas are a known but rare entity of primary cardiac tumors in childhood [1]. There are reports of tumors diagnosed prenatally in screening ultrasound examinations

Fig. 1 Initial radiograph demonstrating an enlarged heart with a prominent right atrium without signs of pulmonary congestion

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Fig. 2 2-D echocardiogram, subxiphoidal view, showing a large tumor in the dilated right atrium attached to the atrial-free-wall with inflow obstruction to the tricuspid valve. LA left atrium, LV left ventricle, RV right ventricle, T tumor

[3]. Pregnancy in our patient had been unremarkable. Retrospectively, the pathologic cardiokotography just before birth may have been the first sign of hemodynamic compromise in this patient. It is likely that an initially rather small hemangioma ruptured just before birth resulting in a rapid expansion of the tumor, as it was filled with blood, that subsequently caused the significant inflow obstruction to the tricuspid valve.

Immediate surgical intervention was necessary due to imminent low output failure of the right heart. The tumor was peeled off the atrial wall causing a large area of endothelial and myocardial trauma. The intractable atrial arrhythmia that developed postoperatively was probably caused by this necessary procedure. Arrhythmias due to right atrial surgery are a known but often unavoidable complication. This tachycardia required initial extracorporeal membrane oxygenation support [4]. Subsequently, it

could be controlled by amiodarone and overdrive pacing, and finally it resolved completely after 2 months.

As in all patient described so far, there are no signs of recurrence of the hemangioma in our patient up to date.

This case report describes a hemangioma as a rare cause of a neonatal intracardiac tumor. In this patient, the large atrial mass resulted in severe inflow obstruction to the tricuspid valve requiring emergent surgical relief. The postoperative course was complicated by rapid atrial arrhythmias probably due to large atrial endothelial and myocardial trauma.

Acknowledgments No grants or other financial support was received for this work.

Conflict of interest No conflict of interest exists for any of the authors.

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