

Multiple cardiac rhabdomyoma with severe obstruction of the right ventricle outflow tract in a neonate

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Background

Cardiac rhabdomyoma is the most common cardiac tumor in fetus and newborn, and can be associated with tuberous sclerosis in the brain. It's often multiple, asymptomatic, and usually regresses spontaneously in a few years. However, when the tumor is big enough to obstruct major cardiac inflow or outflow tract or it causes some critical arrhythmias on the infant, an immediate intervention will be demanding. We present a neonate of multiple cardiac rhabdomyomas with its largest one obstructs remarkably on the RVOT.

Case report

A 5-day-old female newborn was transferred to our hospital due to occasional cyanosis after birth. Echocardiographic study showed multiple tumors in the myocardium with hyperechogeneity. The largest tumor occupied the RVOT and even pushed out of the pulmonary valve. Doppler investigation revealed a systolic pressure gradient across the RVOT by 55 mmHg. There were also positive findings in brain MRI, brain echo, and renal echo, suggesting an association with Tuberous sclerosis. Surgical excision of the RVOT tumor was performed on her 11th day of age. Immediately after disconnecting from the total cardiopulmonary bypass, hypoxemia (SaO₂ <80%) and hypotension (sBP < 60 mmHg) occurred, in spite of high ventilator setting and strong inotropic agents. CXR showed cardiomegaly and lung congestion. She had to be managed with HFO ventilators and NO inhalation. The heart was supported with dopamine and bosmin infusion for one week. Also, a tachycarrhythmia of JET was managed with amiodarone infusion. She was transferred to SBR on the 17th op. day, and discharged 5 days later.

Discussion

This severe complication of lung edema and heart failure immediately after the big, obstructive RVOT tumor was surgically removed from this neonate is considered a phenomenon similar to a postoperative complication when a large amount of pericardial effusion is rapidly drained in a patient of cardiac tamponade.

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