Supraventricular tachycardia in a neonate with cardiac rhabdomyoma and tuberous sclerosis

In Kug Bang, M.D., Yeo Hyang Kim, M.D., Chun Soo Kim, M.D., Sang Lak Lee, M.D., and Tae Chan Kwon, M.D.

Department of Pediatrics, School of Medicine, Keimyung University, Daegu, Korea

Abstract

Primary tumors of the heart are uncommon among pediatric patients. Rhabdomyoma is the most frequent cardiac tumor in infants and children, which is commonly associated with tuberous sclerosis. Tuberous sclerosis is a neurocutaneous syndrome affecting the brain, heart, skin, and other organs. Cardiac rhabdomyomas are reported in 50-64% of infants with tuberous sclerosis. Tuberous sclerosis involves multiple locations in the atrium, ventricle and septum, and may induce mechanical obstruction of the outflow tract and heart failure depending on the location, number, size, and degree of invasion of tumors. Arrhythmias may also develop in infants with cardiac rhabdomyomas, but only a few of these patients require prolonged anti-arrhythmic therapy because arrhythmia often disappears with spontaneous regression of the tumors, and the ultimate prognosis may be decided by the cerebral manifestations. (Korean J Pediatr 2008;51:766-770)

Key Words: Tuberous sclerosis, Rhabdomyoma, Arrhythmia

Introduction

Tuberous sclerosis is an autosomal dominant disorder with involvement on skin, eyeball, nervous system, kidney and heart. Cardiac involvement may be the first manifestation, and it mainly consists of rhabdomyomas. Rhabdomyomas are the most common primary cardiac tumor in fetus and infants, which 50-64% of patients accompany tuberous sclerosis. It can involve multiple locations of atrium, ventricle and septum. According to the tumor’s location, number, size and degree of invasion, various symptoms such as mechanical obstruction of outflow tract, heart failure and arrhythmia may develop. As rhabdomyomatous tissue can generate myocardial electrical potential and act as an accessory pathway, arrhythmias may be developed as main symptom. However, these cases were uncommon in the previous reports.

The writers report experience of a neonate with cardiac tumor and tuberous sclerosis accompanying supraventricular tachycardia that successfully treated with beta blocker.

Case report

A male newborn infant was born by Cesarean section at 40 gestational weeks from a 35 year-old mother and a 37 year-old father at a local obstetric clinic. On five days of life, tachycardia was developed to him, and so transferred to our hospital.

On family history, his father had suffered from seizure disorder with unknown etiology, but his mother and elder brother were unremarkable.

On physical examination of admission day, his blood pressure and breathing rate were 80/50 mmHg and 64 beats per minute respectively. The heart rate was 260 beats per minute with regular rhythm. Liver was palpable 4 cm below the rib. Arterial oxygen saturation of patient in room air condition was 98%. Serologic laboratory findings were unremarkable except elevated N-Terminal pro-B-type Natriuretic Peptide (25,033 pg/mL). The chest X-ray showed cardiomegaly with increased cardiothoracic ratio (0.61) (Fig. 1). A 12-lead electrocardiogram (ECG) was recorded on the day of admission. ECG finding was supraventricular tachycardia (SVT) with narrow QRS complex at a rate of 220 beats per minute (Fig. 2A). Because the patient had heart failure symptoms such as tachypnea, cardiomegaly and
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Fig. 1. Cardiomegaly is seen on the chest X-ray (cardiorthacic ratio=0.61).

Fig. 2. A) Initial electrocardiogram (ECG) on the day of admission showed supraventricular tachycardia at a rate of 220 beats/min; B) ECG after direct current (DC) cardioversion showed sinus rhythm.

Fig. 3. ECG showing multiple echo-dense masses on the left ventricle. A) Echo-dense masses located on the septum and left ventricular free wall in the parasternal long axis view. B) Echo-dense masses located on septum in left-side tilted parasternal long axis view.

hepatomegaly. Direct current cardioversion (1 J/kg) was taken and then sinus rhythms were recovered again (Fig. 2B).

After conversion to sinus rhythm, echocardiography was performed. Multiple echo-dense masses with homogeneous pattern were observed at left and right ventricular wall and ventricular septum, which size was from 12 mm to 25 mm (Fig. 3). However, there was no definite sign of hemodynamic disturbance suggesting obstruction of cardiac inflow or outflow tract.

The brain ultrasonography (USG) and magnetic resonance imaging (MRI) revealed multiple nodules with various size along the part of frontal graymatter and lateral ventricle. These nodules showed high signal on T1 weighted image and low signal on T2 weighted image and did not enhanced by contrast medium (Fig. 4). Other organs including skin did not show any definite abnormal features and/or symptoms. The diagnosis of tuberous sclerosis was confirmed by MRI of the brain showing typical subependymal and cortical calcifications.

On nine days of life, patient had recurrent SVT with rate
Fig. 4. Axial nonenhanced T1-weighted A) and T2-weighted B) magnetic resonance (MR) images show several small subependymal nodules (black arrow) projecting into the lumen of lateral ventricles and hamartomatous lesions (white arrow) in the white matter of both the frontal lobes.

of about 250 beats per minute on ECG monitoring system, and this arrhythmia did not respond to adenosine and digoxin therapy. This intractable and recurrent SVT of patient was controlled successfully with high dose of propranolol (6 mg/day).

Currently he is one year old. Most of cardiac tumors were regressed on the follow-up echocardiographic findings, there was no additional episode of arrhythmia. And also he is healthy without any neurologic symptoms such as convolution or developmental delay.

Discussion

The primary tumors of the heart are uncommon in the pediatric population and usually histologically benign. In the pediatric population, the most common primary tumor of the heart is rhabdomyomas. Congenital rhabdomyomas are known to be associated with other congenital anomalies and a high correlation with tuberous sclerosis. It was reported in 50–64% of infants with tuberous sclerosis.

Tuberous sclerosis is an autosomal dominant disorder with a frequency of 1 in 15,000 to 30,000. It is a neurocutaneous syndrome characterized by central nervous system hamartomas, seizure, developmental delay and skin changes. Cardiac involvement may be the first manifestation and mainly consists of rhabdomyomas.

The development of cardiac rhabdomyomas in tuberous sclerosis was first reported in 1862 by Von Recklinghausen. Thereafter, according to development of echocardiography, asymptomatic cardiac rhabdomyomas could be diagnosed and found about 50% of the patients with tuberous sclerosis.

They can involve multiple locations of atrium, ventricle and septum. Ventricular septum and wall have frequent occurrence over 90% of tumor. Symptoms and manifestations depend on the tumor’s location, number, size and degree of invasion and include outflow tract mechanical obstruction, heart failure and arrhythmia. Arrhythmias could also be developed in infants with cardiac rhabdomyomas but were uncommon manifestation in the previous reports. Arrhythmias could include atrophicventricular block (left bundle branch block), supraventricular tachycardia, ventricular tachycardia and atrial flutter. Especially, pre-excitation syndrome and combination of two or more arrhythmias were more frequent than in normal population.

The specific cell that gives rise to the cardiac rhabdomyomas is controversial. Among several reports, one demonstrated that the cardiac rhabdomyomas were a type of hamatoma originating from embryonic myoblasts and this is a widely accepted theory in the present.

Rhabdomyomas may invade into the myocardium or protrude into the cardiac cavity. Rhabdomyomas invaded into the myocardium may induce ventricular hypertrophy and
cause the electrocardiographic changes. The electrocardiogram could show features of ventricular hypertrophy depending on the size and location of the tumors. A recent report in a patient with rhabdomyoma described ventricular hypertrophy on electrocardiogram. This report demonstrated that the presence of cardiac rhabdomyomas in patients with tuberous sclerosis might explain the ventricular hypertrophy seen on the electrocardiogram through its electrically active tissue without ventricular pressure overload or enlargement. These changes also tend to disappear concomitant with regression of the tumors with increasing age. On the other hand, patients with giant cardiac rhabdomyomas do not necessarily show the electrocardiographic changes of ventricular hypertrophy. In our case, a giant tumor was located in the septum of left ventricle, and extended to the direction of the RVOT. The patient did not show any electrocardiographic changes of ventricular hypertrophy.

Treatment modalities of patients with cardiac tumors and arrhythmias are dependent on clinical symptoms. Because most patients are asymptomatic and their tumors and arrhythmias are regressed spontaneously, surgical removal is indicated only for patients with refractory arrhythmia which has poor response to medical management or significant intracardiac obstruction. In our case, patient showed recurrent SVT and poor feeding and treatment with digoxin and oral beta blocker was started. The patient was maintained on medication for 10 months and had no further episodes of SVT. On follow-up, the patient had no cardiac symptoms, and tumors regressed spontaneously.

In conclusion, only a few patients with tuberous sclerosis and rhabdomyomas require prolonged antiarrhythmic therapy because most of the tumors regress spontaneously. The cardiac outcome is usually favorable and cerebral manifestations determine the ultimate prognosis in these patients.

References


