Korean Journal of Pediatrics Vol. 51, No. 7, 2008
DOI : 10.3345/kjp.2008.51.7.766
■ Case report ■

Supraventricular tachycardia in a neonate with cardiac rhabdomyoma and tuberous sclerosis

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= 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 rhadomyomas 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 rhadomyomas, 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[1].

Rhabdomyomas are the most common primary cardiac tumor in fetus and infants, which 50-64% of patients accompany tuberous sclerosis[2]. 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[3]. 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[3, 4].

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

Received : 8 March 2008, Accepted : 2 May 2008
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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

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 ventriclular 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

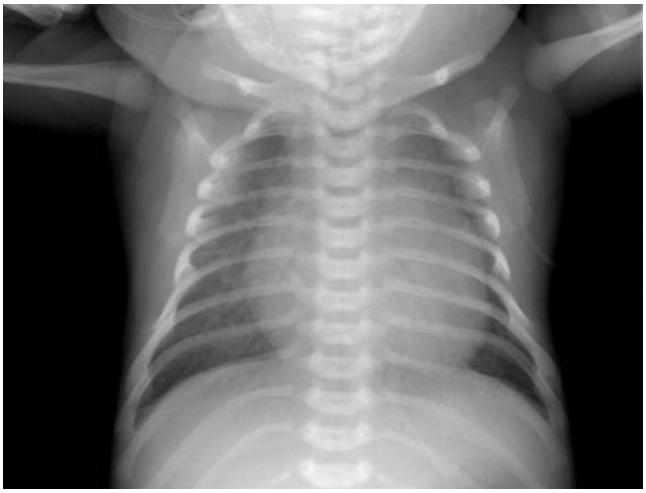

Fig. 1. Cardiomegaly is seen on the chest X-ray (cardiothoracic ratio=0.61).

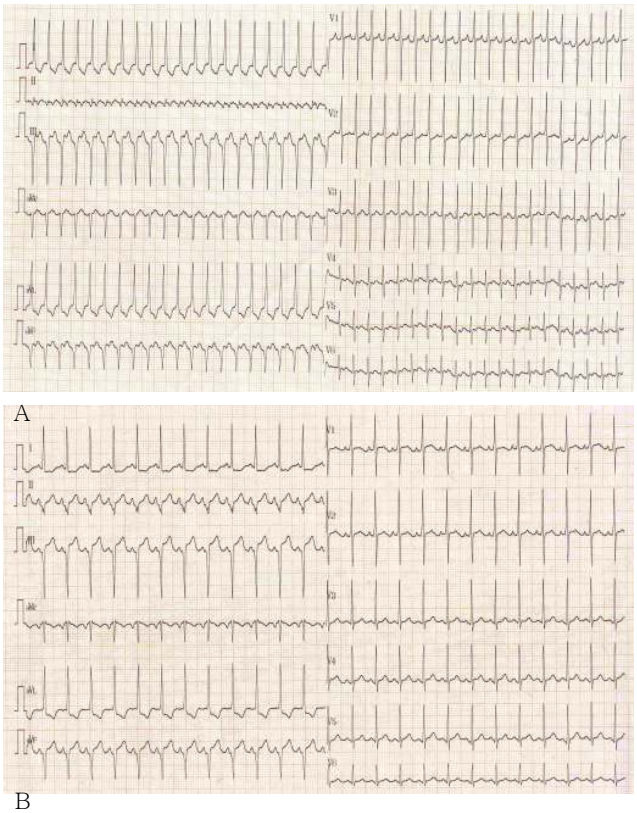

Fig. 2. A) Initial electrocardiogram (ECG) on the day of admission showed supraventriuclar tachycardia at a rate of 220 beats/min; B) ECG after direct current (DC) cardioversion showed sinus rhythm.

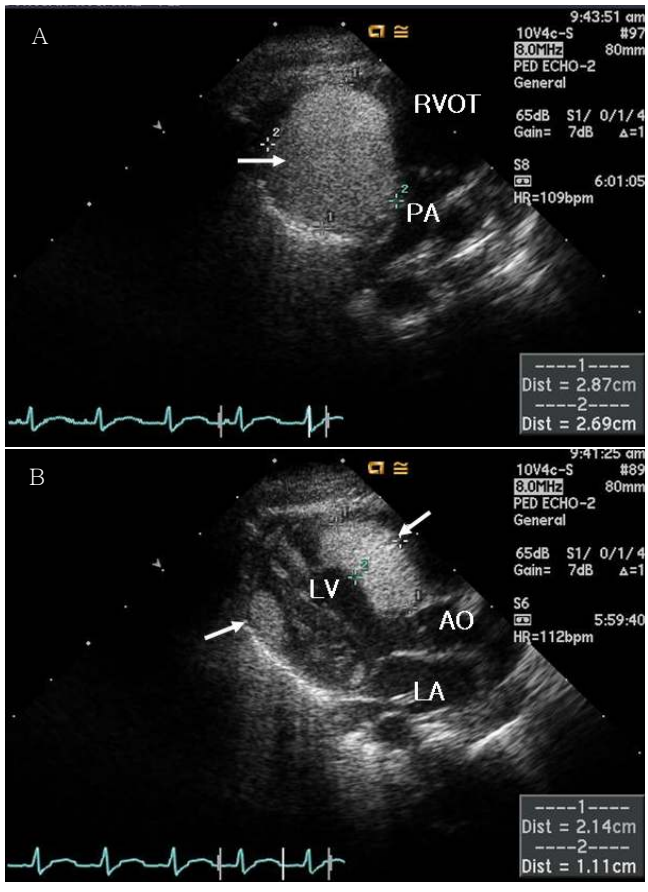

Fig. 3. ECG showing multiple echo-dense masses on the left ventricle. A) echo-dense masses located on the septum and left ventricular free wall in the parasternal long axis view. B) echo-dense masses located on septum in left-side tilted parasternal long axis view.

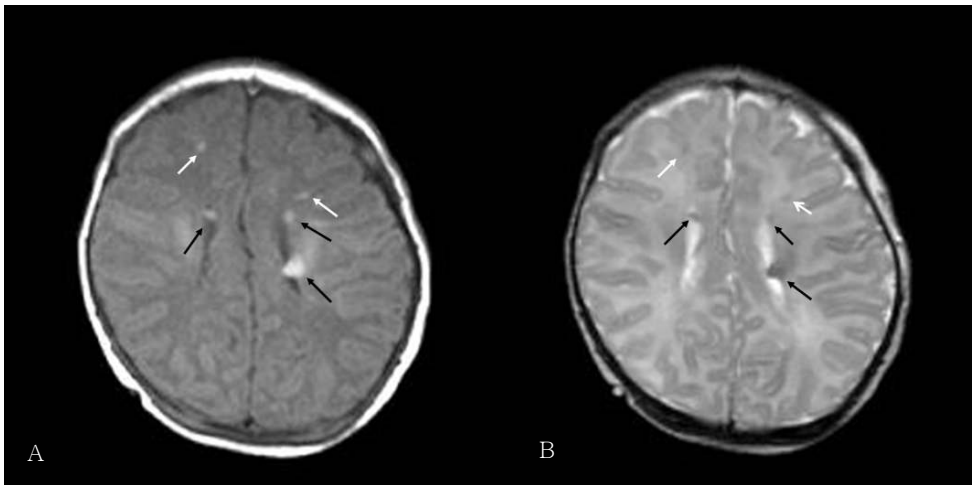

Fig. 4. Axial nonenhanced T1-weighted A) and T2-weighted B) magnetic resonance (MR) images show several small subependymal nodules (black arrow) projecting into the lumen of lateral ventricles and hamartomatous lesions (white arrow) in the white matter of both the frontal lobes.

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 convulsion 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[5]. Congenital rhabdomyomas are known to be associated with other congenital anomalies and a high correlation with tuberous sclerosis[5]. It was reported in 50-64% of infants with tuberous sclerosis[2].

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

The development of cardiac rhabdomyomas in tuberous sclerosis was first reported in 1862 by Von Recklinghause[6]. Thereafter, according to development of echocardiography, asymptomatic cardiac rhabdomyomas could be diagnosed and found about 50% of the patients with tuberous sclerosis[7, 8].

They can involve multiple locations of atrium, ventricle and septum. Ventricular septum and wall have frequent occurrence over 90% of tumor[3]. 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[3]. Arrhythmias could also be developed in infants with cardiac rhadomyomas but were uncommon manifestation in the previous reports[9]. Arrhythmias could include atrioventricular block (left bundle branch block), supraventricular tachycardia, ventricular tachycardia and atrial flutter[3, 6]. Especially, preexcitation syndrome and combination of two or more arrhythmias were more frequent than in normal population[8, 10].

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 harmatoma originating from embryonic myoblasts and this is a widely accepted theory in the present[3]. The other reported that the cardiac rhabdomyomas took origin from the Purkinje cell[4]. Therefore, rhabdomyomatous tissue could generate myocardial electrical potential and might act as an accessory pathway associated with the preexcitation syndrome.

Rhabdomyomas may invade into the myocardium or protrude into the cardiac cavity[3]. 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[11]. 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 electrocardiaographic 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 ventriuclar 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[12, 13], a small number of patients require antiarrhythmic therapy[14, 15]. Surgical removal is indicated only for patients with refractory arrhythmia which has poor response to medical management or significant intracardiac obstruction[16, 17]. 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.

한 글 요 약

상심실성 빈맥과 심장 종양으로 진단된 결절성 경화증 1예

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심장의 원발성 종양은 소아에서는 매우 드문 질환이다. 횡문근종은 태아, 영아 및 소아 연령에서 가장 흔한 원발성 심장 종양으로 결절성 경화증이 동반되는 것으로 보고되고 있다. 결절성 경화증은 뇌, 심장, 피부 및 기타 장기들을 침범하는 신경피부 증후군으로, 결절성 경화증을 가진 영아의 50-64%에서 동반된다. 심장 횡문근종은 종괴는 심방을 비롯하여 심실 중격과 심실벽에 다발성으로 존재하며 종양의 위치와 크기, 침범 정도에 따라 심장 유출로 협착 및 심부전과 같은 여러 가지 임상 양상을 보일 수 있다. 드물지만 부정맥을 주증상으로 발현하는 경우도 보고되고 있다. 하지만 대부분 심장의 횡문근종이 특별한 문제를 일으키지 않고 성장과 함께 쇠퇴되는 경향을 보이므로 극히 일부분에서만 장기적인 항부정맥제 사용이 요구된다. 환자의 최종적인 예후는 동반되는 뇌병변에 의해 결정되는 경향이 있다. 저자들은 상심실성 빈맥으로 입원치료 중 진단된 심장 횡문근종을 가진 결절성 경화증 1예를 경험하였기에 보고하는 바이다.

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