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# An Extremely Rare Cause of Wolff-Parkinson-White Syndrome: Rhabdomyoma in Association With Tuberous Sclerosis

CASE REPORT

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ABSTRACT

Rhabdomyomas are the most common primary cardiac tumors in infants and children. They are usually associated with tuberous sclerosis (TS). As the tumors tend to regress spontaneously, surgical intervention is not usually performed unless they become obstructive or cause incessant arrhythmias. We report an extremely rare case of rhabdomyoma serving as a substrate for Wolff-Parkinson-White (WPW) syndrome and intractable supraventricular tachycardia accompanied by TS. Our case is particularly interesting because it was diagnosed prenatally. The signs of WPW syndrome disappeared from the electrocardiogram with the regression of the tumor.

**Keywords:** Wolff-Parkinson-White Syndrome, child, rhabdomyoma

## INTRODUCTION

Rhabdomyomas are the most common cardiac tumors in infants and children, and they are closely related with tuberous sclerosis (TS). A significant part of rhabdomyomas is asymptomatic, and they regress on follow-up. However, symptoms of cardiac failure, arrhythmias, and obstruction can be observed depending on their location in the heart. They require urgent medical or surgical treatment (1, 2).

Rhabdomyoma-related arrhythmias are reported as premature atrial and ventricular contractions, supraventricular and ventricular tachycardia, sinus node dysfunction, atrioventricular block, and Wolff-Parkinson-White (WPW) syndrome (1, 3, 4). There are only a few studies of WPW syndrome occurring in association with TS, with and without rhabdomyoma. Furthermore, to the best of our knowledge, only one case has been reported about rhabdomyoma that functions as a substrate for WPW as the tumor location site. In this case, no accompanying TS was found (5–7).

We described a very rare cause of a WPW syndrome occurring in association with TS and rhabdomyoma diagnosed prenatally. The signs of WPW syndrome disappeared from the electrocardiogram (ECG) with the regression of the tumor.

## CASE REPORT

A 26-year-old G2P1 pregnant woman was referred to our clinic at 38 weeks 4/7 days of gestational age for fetal echocardiography because fetal cardiac mass and tachycardia were detected. Fetal echocardiography showed multiple rhabdomyomas within the right and left ventricle cavities and in the right atrium that were protruded to the right ventricle through the tricuspid valve. Supraventricular tachycardia (SVT) diagnosis was made upon observing that fetal heart rate was 240 beats/min, and M-mode and pulse wave Doppler echocardiography showed regular 1:1 atrioventricular conduction. Since the fetus was term, birth of the baby was decided. On physical examination postnatally, birth weight was 3380 g, heart rate was 230 beats/min, blood pressure was 70/40 mm Hg, respiratory rate was 40 breaths/min, and $SpO_2$ was 96%. There were four hypomelanotic macules on the back. ECG revealed a narrow QRS tachycardia of 230 beats/min with regular RR interval (Fig. 1a).

Echocardiography showed a homogeneous, echo-dense mass that was compliant with rhabdomyoma, measuring approximately 7x9 mm in the right atrium extending into the right ventricle through the tricuspid annulus. Additionally, multiple masses localized at the left ventricular apex in the right and left ventricle cavities were observed (Fig. 2). Rhabdomyomas did not cause any obstruction, and left ventricle systolic function was normal.

Since initial treatments with vagal maneuvers and intravenous adenosine dosage of 0.2 mg/kg and 0.3 mg/kg were ineffective, esmolol was started at 150 µg/kg bolus followed by continuous infusion starting at 100 µg/kg/

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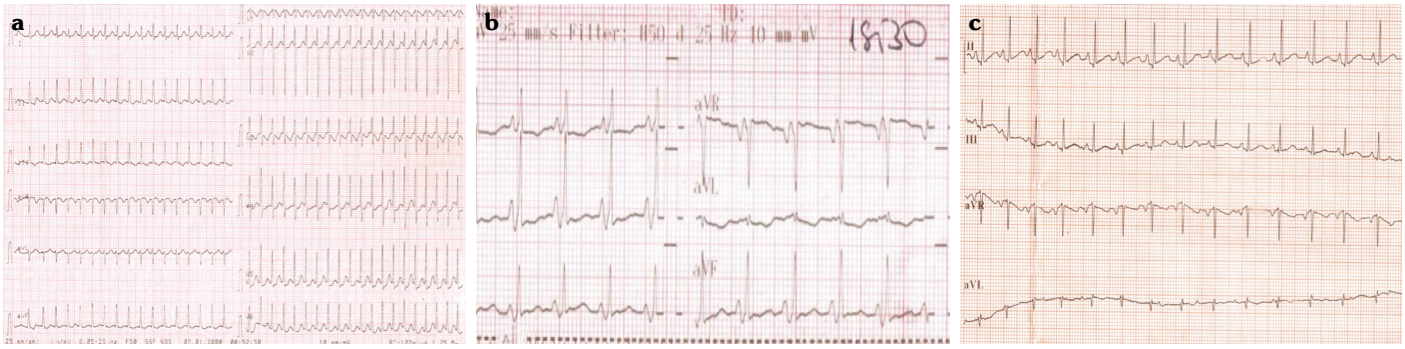

**Figure 1. a–c.** **(a) ECG showing narrow QRS tachycardia at postnatally. (b) ECG taken in sinus rhythm show that properties of Wolff-Parkinson-White syndrome. (c) ECG showing normal sinus rhythm and disappearance of the Wolff-Parkinson-White syndrome after the regression of rhabdomyoma acting as a substrat for WPW syndrome**

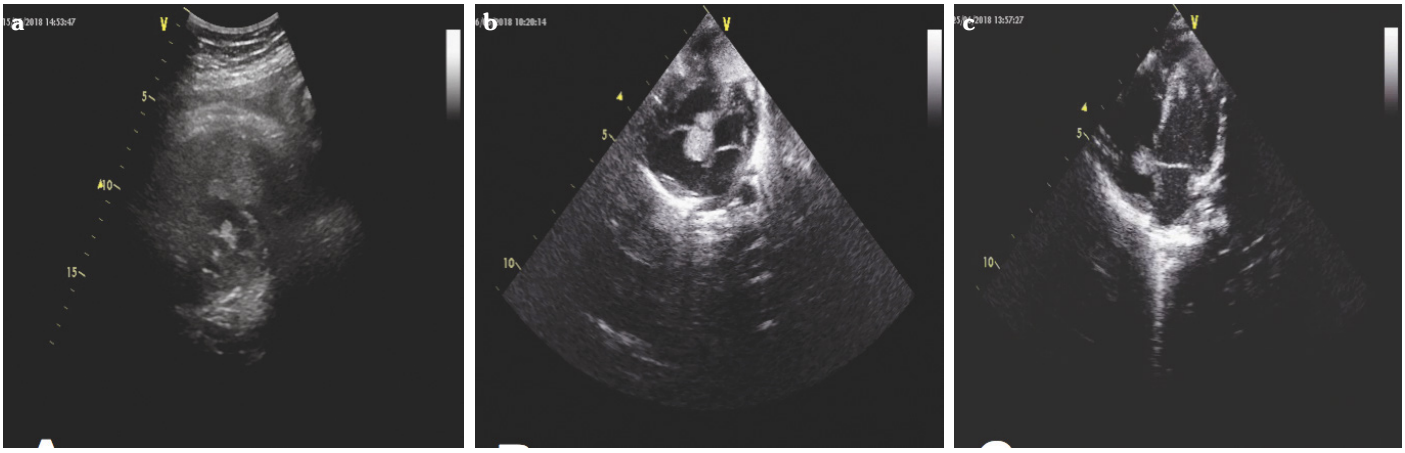

**Figure 2. a–c.** **(a) Fetal echocardiography showing multiple cardiac rhabdomyomas. (b) Echocardiographic appearance of the patient at the time of diagnosis: a rhabdomyoma in the right atrium extended into right ventricle through the tricuspid annulus and different sizes of multiple rhabdomyomas localized in the left ventricular apex, right and left ventricle cavity in apical 4-chamber view. (c) Echocardiographic views of rhabdomyomas after 3-month follow up. Note the regression of rhabdomyomas**

min. However, SVT persisted although the infusion rate increased up to 200 µg/kg/min. Hence, amiodarone was administered at 5 mg/kg bolus followed by infusion at 10 µg/kg/min. Since SVT did not show any regression with these treatments, synchronized cardioversion was performed with restoration of normal sinus rhythm. ECG in sinus rhythm demonstrated signs of WPW syndrome (Fig. 1b). Laboratory test results, including arterial blood gas analysis, complete blood count, serum electrolytes, thyroid function tests, C-reactive protein levels, and procalcitonin levels, were normal. Owing to non-sustained SVT episodes, oral propranolol (1 mg/kg for 8 h) was added to the treatment on day 4 of the patient's follow-up. A cranial magnetic resonance imaging (MRI) performed in view of onset myoclonic jerks at 15 days of life showed subependymal tubers confirming TS. On follow-up assessment of the patient at month 3, echocardiography showed significant regression of rhabdomyomas, and ECG and 24-hour Holter examination revealed sinus rhythm without WPW syndrome (Fig. 1c).

## DISCUSSION

TS is a neurocutaneous disorder that affects the brain, heart, skin, and other organs. The most common cardiac presentation of the disease is cardiac rhabdomyoma, which is presumed to occur in at least 60% of children diagnosed with TS. The rate of TS is higher in the presence of multiple rhabdomyoma (5). Since our case had multiple cardiac rhabdomyomas, hypopigmented macules, and myoclonic seizures, cranial MRI was obtained, and TS diagnosis was confirmed upon finding subependymal nodules.

Rhabdomyomas form approximately 60%–65% of cardiac tumors in children. While the diagnosis can be made prenatally, most of the patients are diagnosed in the newborn and infancy period. Cases that are diagnosed prenatally should be monitored closely with respect to cardiac obstruction or resistant arrhythmia-induced hydrops fetalis (5, 8). Our case had been diagnosed prenatally, and since the gestational week was suitable, the birth of the fetus was decided due to incessant SVT.

Rhabdomyoma-related arrhythmias have been reported as premature atrial and ventricular contractions, supraventricular and ventricular tachycardia, sinus node dysfunction, atrioventricular block, atrial flutter with non-conducted premature atrial contraction, and WPW syndrome (1, 3, 4, 9). De Rosa et al. reported their 18-year-long experiences related with cardiac rhabdomyoma, and they found intractable arrhythmias in five cases during the newborn period. These arrhythmias were determined as

atrial tachycardia, ventricular tachycardia, and WPW syndrome-related SVT with bradycardia due to blocked atrial extrasystole. Arrhythmias were controlled with drugs, including propranolol, sotalol, and amiodarone (4). Miyake et al. reported that cardiac arrhythmias accompany 13% of rhabdomyomas, and intractable SVT develops in only 2 of the 10 cases who were found to have pre-excitation (1).

As rhabdomyomas tend to regress spontaneously in the first years of life, surgical intervention is indicated only in patients whose arrhythmias are refractory to medical treatment (10). In our case study, since intractable SVT was controlled with medical treatment, surgery was not performed.

There are only a few isolated studies reporting the comorbidity of TS, rhabdomyoma, and WPW syndrome, and its prevalence is not fully known. Furthermore, to our knowledge, only one case has been reported about rhabdomyoma that functions as a substrate for WPW as the location site. TS was not found in this case (5–7). The etiology of WPW syndrome in TS has not been fully explained. It is known that some cells in cardiac rhabdomyomas of patients with TS are identical with Purkinje cells structurally. For this reason, it is thought that rhabdomyomatous tissue acts as a pathway bypassing atrioventricular node (5). In our case study, rhabdomyoma extending from the right atrium via the tricuspid valve to the right ventricle acted as an accessory pathway and caused resistant SVT. With the regression of the patient's rhabdomyoma in the clinical follow-up, WPW symptoms disappeared from the ECG.

In conclusion, necessary planning should be made to prevent potential hemodynamic and arrhythmic complications in babies diagnosed with rhabdomyoma prenatally. Rhabdomyomas very rarely act as accessory pathways and cause life-threatening arrhythmias. In patients with resistant arrhythmia, due to the spontaneous regression feature of the tumor, medical treatment should be attempted first, if possible. Since TS symptoms can appear any time, careful monitoring is also required with respect to this.

**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

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**Author Contributions:** Conceived and designed the experiments or case: ÖE, CK, Dİ. Performed the experiments or case: ÖE, CK, Dİ. Analyzed the data: ÖE, CK. Wrote the paper: ÖE. All authors have read and ap- proved the final manuscript.

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