**Case Report**

A Rare Presentation of Cardiac Rhabdomyoma in Children: 
Sudden Cardiac Arrest

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**Abstract**

Cardiac rhabdomyomas are mostly asymptomatic. Although the mechanism has not been well characterised, cardiac rhabdomyomas can lead to ventricular tachycardia (VT). We report a case of cardiac rhabdomyoma in which the initial presentation was sudden cardiac arrest (SCA) due to VT. Previously healthy 16-year-old boy collapsed after he worked at farm. After successful resuscitation and cardioversion, he was admitted to the hospital. Non-sustained VT was detected on 24-hour Holter monitoring. Transthoracic echocardiography and magnetic resonance imaging showed a mass on the left ventricle apex. After surgical resection, Holter tests were completely normal. The histopathologic study confirmed the diagnosis of rhabdomyoma. Data regarding arrhythmias associated with cardiac rhabdomyoma are limited with small series. Management strategies include antiarrhythmics, cardioverter-defibrillator and surgery, but as to which strategy can be considered as optimal remains unclear. Benign cardiac tumours should be kept in mind as a cause of SCA in children and resection seems to be effective for treating associated VT.

**Key words** Cardiac rhabdomyoma; Children; Sudden cardiac arrest; Ventricular tachycardia

**Introduction**

Sudden cardiac arrest (SCA) in children is a rare, but dramatic condition. The incidence of SCA is approximately 0.013%, of which 30% of cases have present cardiac diseases.¹ Common causes of SCA are structural cardiac lesions [cardiomyopathies, coronary artery diseases, myocarditis, congenital heart diseases, arrhythmogenic right ventricular dysplasia (ARVD), etc.], electrical abnormalities (long QT syndrome, short QT syndrome, catecholaminergic polymorphic ventricular tachycardia, Wolf-Parkinson-White syndrome Brugada syndrome, complete heart block, etc.) and acquired conditions (drugs/stimulants, chemicals, pulmonary hypertension).¹²

Primary cardiac tumours are usually benign and very rare in children with an incidence of 0.2%. However, they have the potential for haemodynamic compromise and life-threatening dysrhythmias.³ Herein; we present a case of benign cardiac tumour of which the initial presentation was ventricular tachycardia and SCA.

**Case Report**

A 16-year-old previously healthy boy collapsed on his way home by a tractor after he worked on a farm. The family immediately called emergency medical service (EMS).
Ventricular tachycardia (VT) was observed on the monitor of the cardioverter-defibrillator. After 10 minutes of resuscitation and cardioversion, the patient was admitted to the hospital.

The patient was intubated at admission. Heart rate, blood pressure, oxygen saturation values were within normal limits. After initial stabilisation and treatment in the paediatric intensive care unit (PICU), the patient was evaluated for possible cardiac diseases and arrhythmias.

Detailed medical history and physical examination results were unremarkable. There were no complaint of palpitation or palpitation-associated symptoms such as dizziness, shortness of breath, sweating, headaches and chest pain before and during the attack. Family history revealed no clues for SCA.

The electrocardiography (ECG) of the VT attack was seen on the monitor of the cardioverter-defibrillator but it was not documented during resuscitation. Chest X-ray and 12-lead ECG were evaluated at the PICU. Cardiothoracic index was 0.48 and there were not any clues of long QT syndrome, Brugada syndrome, short QT syndrome or other arrhythmias.

Past medical history, physical examination and baseline ECG of the patient were unremarkable. Pharmacologic treatment was initiated with propafenone and bisoprolol due to the suspicion of VT.

Twelve-lead twenty-hour Holter recording showed multiple ventricular ectopic beats and two unifocal monomorphic non-sustained VT attacks with a rate of 165 beats per minute, which did not cause haemodynamic compromise. The VT was compatible with left sided VT (right bundle branch block pattern, superior axis, QRS transition beyond V3).

Transthoracic echocardiography (TTE) was performed especially for the detection of HCM, ARVD, and congenital heart diseases, which are the most common causes of VT. Echocardiography demonstrated a homogeneous intramural left ventricular mass in the apex. There were not any echocardiographic findings of inflow or outflow obstruction of ventricles.

Cardiac magnetic resonance imaging (MRI) revealed a 2.5x1.5x1 cm, well-demarcated, firm intramural mass unrelated to the intraventricular structures. After gadolinium injection, late contrast-enhanced cardiac MRI images showed a homogeneous and intense bright mass, suggesting rhabdomyoma (Figure 1).

Ventricular tachycardia morphology suggested an origin at the left ventricular apex, coinciding with the cardiac tumour's location. The decision was taken to perform a complete surgical resection of the tumour and hence the mass was resected completely (Figure 2).

Holter recordings after surgery were normal and resection of the mass seemed to be effective for the elimination of VT. The histopathologic evaluation of frozen sections confirmed the diagnosis of rhabdomyoma. After surgery, the patient was evaluated by a paediatric neurologist for tuberous sclerosis and was found to be normal. Holter recording and treadmill test at the 6th month period after the operation were also found to be completely normal.

**Discussion**

SCA is an uncommon and critical condition. For effective management of SCA, clarifying the underlying problem is as important as a successful resuscitation. Cardiac causes of SCA are generally grouped into three categories: structural cardiac defects, primary cardiac electrical abnormalities and acquired diseases. Electrocardiography, TTE, and 24-hour Holter monitoring must be performed after a detailed medical history and physical examination. In the presence of problematic past medical history and ECG clues of arrhythmias, advanced genetic tests of arrhythmias should be planned to diagnose the underlying disease.

Initial TTE generally focusses on systolic cardiac functions, major cardiac defects, HCM and especially ARVD in case of VT suspicion. The echocardiographic evaluation of the patient was normal in terms of these diseases. Although the first 24 hours were uneventful in PICU, 24-hour Holter monitoring showed non-sustained left-sided VT attacks. Therefore, a second TTE was performed carefully and a homogeneous intramural left ventricular mass protruding to inferior was shown in the apex.

Biopsy and histologic assessment evaluation are the gold standards for confirmation of the diagnosis of tumours; although TTE or cardiac MRI imaging is usually adequate to facilitate the diagnosis of cardiac tumours. We planned a cardiac MRI to define the shape, diameter and location of the lesion. Cardiac MRI revealed a 2.5x1.5x1 cm, well-demarcated, firm intramural mass suggesting rhabdomyoma.

Primary cardiac tumours are rare with an incidence of 0.03-0.3% in children. Although the majority are asymptomatic, they can result in cardiac failure,
Figure 1  Protruding mass at the apical portion of left ventricle at MRI (white arrows).

Figure 2  Intraoperative pictures of cardiac rhabdomyoma.
intracardiac obstruction depending on the size and the location of the mass. Data regarding arrhythmias associated with cardiac tumours are limited with small series and case reports.\textsuperscript{6-8} With cardiac tumours, 24\% of patients have clinically significant arrhythmias (16\% ventricular tachycardia, 2\% sudden cardiac arrest).\textsuperscript{9} Surprisingly, the patient did not show palpitation, dizziness, presyncope-syncope, chest pain or any other cardiac symptoms before.

Management strategies for arrhythmias associated with cardiac tumours are pharmacologic treatment, cardioverter-defibrillator implantation and surgical excision; but as to which treatment strategy is optimal remains unclear.\textsuperscript{9} We started propafenone and bisoprolol to control cardiac rhythm before a decision of surgical procedure. Holter tests showed that non-sustained left-sided VT attacks persisted even though the patient was not sensing these. The ineffectiveness of antiarrhythmic treatment obligated a surgical resection. Arrhythmic events associated with cardiac tumours are known to be reversible with resection of the tumour.\textsuperscript{5}

Surgical resection was performed successfully. Holter tests after surgery were completely normal and resection of the mass seemed to be effective for the elimination of VT. The histopathologic evaluation of frozen sections confirmed rhabdomyoma diagnosis.

Finally, it can be stated that SCA in children is a rare, but dramatic, public health problem. Clarifying the cardiac cause is as important as resuscitating the patient. Unusually, a benign cardiac tumour could result in SCA especially as the first symptom. Surgical resection should be performed for serious VT or SCA related with cardiac tumours. Physicians should keep cardiac masses in mind as a cause of SCA and VT in children which can be treated successfully by the excision of the mass.

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### Conflict of Interest

All authors declare that they have no conflicts of interest.

### References