

CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Echogenic cardiac mass in the left atrium and associated supraventricular tachycardia in a neonate

Ljiljana Pejčić¹, Radmila Mileusnić-Milenović², Marija Ratković-Janković¹¹Clinical Center of Niš, Clinic for Children's Internal Diseases, Serbia;²Institute of Neonatology, Belgrade, Serbia**SUMMARY**

Introduction Primary cardiac tumors in children are rare and the majority of them are diagnosed before the age of one year. They are mainly rhabdomyomas and have a tendency to regress. The incidence of arrhythmias is not well-defined, depending on the size and location of tumors.

Case outline The authors report a female neonate with ongoing fetal supraventricular tachycardia (SVT). Delivery by urgent cesarean section was performed with neither fetal echocardiography nor fetal antiarrhythmic drug intervention. Electrocardiogram confirmed tachycardia with narrow QRS complex at a rate of 260 beats/min. converting to sinus rhythm after a third dose of intravenous bolus injection of adenosine-5'-triphosphate. But the rhythm reverted to SVT showing refractory supraventricular reentrant tachycardia. Echocardiography performed after conversion to sinus rhythm showed an echogenic, well circumscribed mass in the left atrium, fixed to the primum atrial septum without other structural defects. SVT was treated by a bolus of amiodarone followed by an intravenous infusion. Long-term management with oral amiodarone and beta blocker had a good response. During the one-year follow-up echocardiograms were performed every month showing complete regression of cardiac mass, and there has been no recurrence of tachycardia since neonatal period.

Conclusion Tumor regression and a good long-term outcome in our patient suggest that it was probably a small but unfavorably positioned rhabdomyoma, associated with fetal and perinatal SVT. Prognosis and outcome of the disease depends on timely diagnosis and prompt and adequate treatment of associated life-threatening arrhythmias.

Keywords: cardiac tumor; supraventricular tachycardia; rhabdomyoma; neonate

INTRODUCTION

Primary cardiac tumors are rare in fetuses and neonates, with an incidence ranging between 0.0017% and 0.28 % [1]. Most of these tumors are benign, although sometimes their localization and clinical manifestation can be malignant [2]. Others, in the absence of hemodynamic changes, remain undiagnosed and spontaneously regress with time [3]. Histologically, the most common tumor in the neonatal age is rhabdomyoma [4]. If tumors are multiple, associated eventual tuberous sclerosis should be considered [5]. As rhabdomyomatous tissue can generate myocardial electrical potential and act as an accessory pathway, arrhythmias may develop as the main symptom, even in a fetus, where it could cause hydrops fetalis. Persistent arrhythmia in postnatal life increases the risks of neonatal death and these patients require prolonged anti-arrhythmic therapy [6]. Arrhythmia can sometimes disappear with spontaneous regression of the tumors [5]. Tachycardias most commonly associated with cardiac tumors described in the literature are ventricular tachycardia and supraventricular tachycardia (SVT) [2, 7].

The authors report a neonate with an ongoing fetal SVT and left atrium cardiac mass detected after birth.

CASE REPORT

A two-hour-old newborn was admitted to the neonatal intensive care unit after emergency cesarean section due to antenatal diagnosed SVT at the gestational age of 36/37 weeks. SVT was accidentally discovered during a routine control examination of pregnant primipara women. Delivery by urgent cesarean section was performed with neither fetal echocardiography nor fetal antiarrhythmic drug intervention.

A female baby with Apgar score of 8/1, weighing 3,280 g, with a length of 49 cm, occipitofrontal circumference of 34 cm, oxygen saturation of 95%, and a heart rate of 250 beats/min. (bpm), was immediately referred to pediatric cardiology.

On examination, the newborn had livid skin and expressed acrocyanosis, breathing rate of 58 cycles/min., blood pressure of 85/55 mmHg, liver was palpable 2 cm below the rib. The heart rate was 250 bpm, there was no murmur and peripheral pulses were palpable. Electrocardiogram (ECG) confirmed SVT with narrow QRS complex at a rate of 260 bpm (Figure 1a). Tachycardia was treated by intravenous bolus injection of adenosine-5'-triphosphate (ATP). SVT was converted to sinus rhythm after a third ATP dose of 0.3 mg/kg. No serious side effects of ATP injection occurred. However, the

Received • Примљено:

February 20, 2017

Accepted • Прихваћено:

March 13, 2017

Online first: March 17, 2017

Correspondence to:

Ljiljana PEJČIĆ
Vizantijski bulevar 98/36
18000 Niš, Serbia
ljiljasonja@yahoo.com

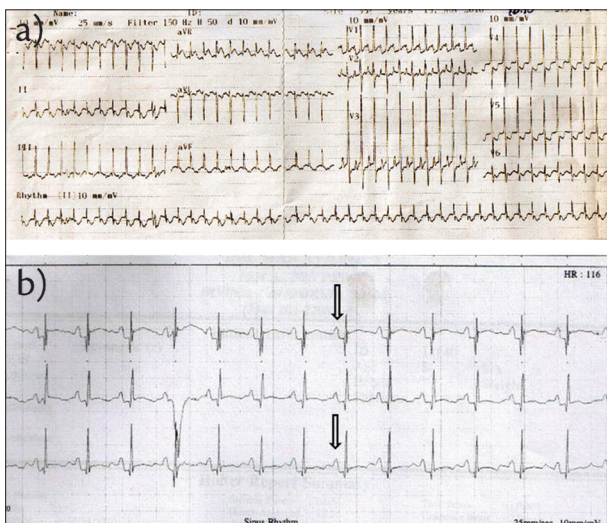


Figure 1. (a) Initial electrocardiogram showing supraventricular tachycardia; (b) electrocardiogram after converting to sinus rhythm showing giant P waves and the first degree AV block

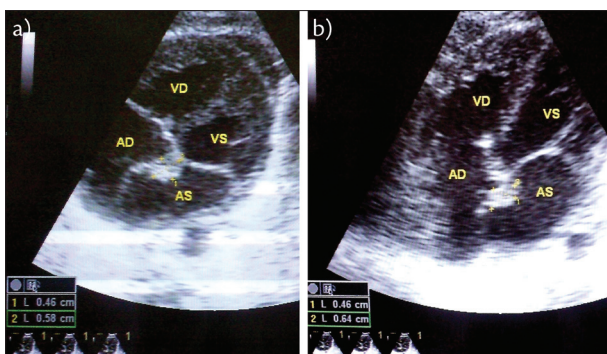


Figure 2. Neonatal echocardiogram (a) in subcostal four-chamber view and (b) long-axis four-chamber view; cardiac mass is visible in the left atrium, fixed to the primum atrial septum

rhythm frequently reverted to SVT, showing refractory supraventricular re-entrant tachycardia. During continuous ECG monitoring, when newborn was in a sinus rhythm, ECG revealed giant P waves and the first degree AV block (Figure 1b).

Echocardiography performed after the conversion to sinus rhythm showed an echogenic, well-circumscribed mass (6 × 5 mm) in the left atrium, fixed to the primum atrial septum (Figure 2). There were neither other structural defects nor signs of hemodynamic disturbance. SVT was thought to be secondary to a small cardiac tumor, especially because of its localization.

The family history did not show any birth defects, no data of benign or malignant tumors, especially no history of tuberous sclerosis. The brain ultrasonography was normal and there were no other clinical markers of tuberous sclerosis at birth.

SVT was successfully treated by a bolus of amiodarone followed by an intravenous infusion. Long-term management with oral amiodarone and propranolol had a good response.

During the one-year follow-up echocardiograms were performed every month showing regression of the cardiac mass. By the eighth month of age the tumor had completely regressed (Figure 3). Amiodarone was withdrawn

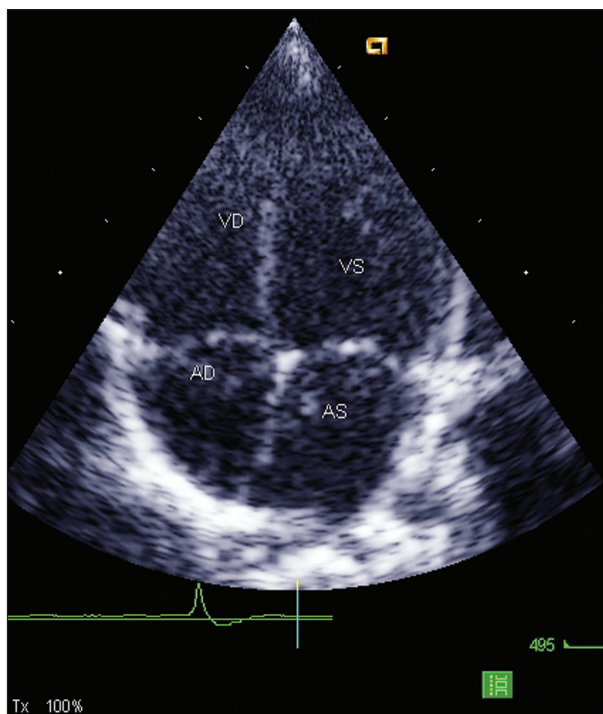


Figure 3. Echocardiogram performed in the eighth month showing that the tumor had completely regressed

at the age of six months (propranolol at eight months) and serial Holter studies documented that there had been no recurrence of tachycardia since the neonatal period.

Currently, she is a healthy six-year-old girl with no cardiac symptoms.

DISCUSSION

Primary pediatric cardiac tumors are uncommon and usually histologically benign. Rhabdomyomas are most common, followed by teratomas, myxomas, and fibromas, and the majority of them are diagnosed before the age of one year [8]. Symptoms, clinical manifestations and treatment depend on tumor size, location, number, and histological type. They can be very different and unpredictable. Thus a giant tumor in the silent zone can be asymptomatic, while a small tumor close to the conduction system, can cause arrhythmia resistant to therapy and even sudden cardiac death [2]. This report precisely describes a situation where a small tumor at a critical location caused refractory SVT that required emergency treatment and long-term management, fortunately with a good response. Our dilemma was related to the histologic type of the tumor. Rhabdomyomas appear on ultrasound as round, homogeneous, hyperechogenic mass, similar to the echocardiographic picture that we have seen in our patient. However, differential diagnosis between rhabdomyoma, fibroma, or myxoma using ultrasonography for a single cardiac mass remains very difficult [3]. Recent reports describe mostly fetuses and newborns with rhabdomyomas and associated SVT with different outcomes. The diagnosis of rhabdomyoma was confirmed either after surgical tumor removal or by autopsy [2, 4, 9]. Other authors had similar experiences

to the one we describe: tumor regressed spontaneously, and the patient had no further episodes of SVT [5].

Tumor regression and a good long-term outcome in our patient suggest that it was probably a small but unfavorably positioned rhabdomyoma, associated with fetal and perinatal SVT. Antenatal detection of fetal cardiac tumors ensures better prenatal and postnatal management [10].

In conclusion, rhabdomyomas are mostly benign and have tendency to regress, but their prognosis depends on the fact that they can be associated with life-threatening arrhythmias and tuberous sclerosis. Adequate and prompt management of arrhythmias and potential cerebral events directly affect the prognosis in these patients.

REFERENCES

1. Uzun O, Wilson DG, Vujanac GM, Parsons JM, De Giovanni JV. Cardiac tumours in children. *Orphanet J Rare Dis.* 2007; 2(1):11–24.
2. Myers KA, Wong KK, Tipple M, Sanatani S. Benign cardiac tumours, malignant arrhythmias. *Can J Cardiol.* 2010; 26(2):58–61.
3. Burke A, Virmani R. Pediatric heart tumours. *Cardiovasc Pathol.* 2008; 17(4):193–8.
4. Sadoh WE, Obaseki DE, Amuabunos EA, Eregie CO, Isah IA, Idemudia E, et al. Cardiac Rhabdomyoma in a Neonate with Supraventricular Tachycardia. *World J Pediatr Congenit Heart Surg.* 2014; 5(1):110–3.
5. Bang I, Kim YH, Kim CS, Lee SL, Kwon TC. Supraventricular tachycardia in a neonate with cardiac rhabdomyoma and tuberous sclerosis. *Korean J Pediatr.* 2008; 51(7):766–70.
6. Chao AS, Chao A, Wang TH, Chang YC, Chang YL, Hsieh CC, et al. Outcome of antenatally diagnosed cardiac rhabdomyoma: case series and a meta-analysis. *Ultrasound Obstet Gynecol.* 2008; 31(3):289–95.
7. Miyake CY, Del Nido PJ, Alexander ME, Cecchin F, Berul CI, Triedman JK, et al. Cardiac Tumors and Associated Arrhythmias in Pediatric Patients, With Observations on Surgical Therapy for Ventricular Tachycardia. *J Am Coll Cardiol.* 2011; 58(18):1903–9.
8. Sallee D, Spector ML, van Heeckeren DW, Patel CR. Primary pediatric cardiac tumors: a 17 year experience. *Cardiol Young.* 1999; 9(2):155–62.
9. Padalino MA, Vida VL, Bhattarai A, Reffo E, Milanese O, Thiene G, et al. Giant Intramural Left Ventricular Rhabdomyoma in a Newborn. *Circulation.* 2011; 124(20):2275–7.
10. Amelia A, Mohd Nizam MB. Perinatal Management of Cardiac Tumors: A Case Series. *Med J Malaysia.* 2013; 68(4):374–5.

Хиперехогено ткиво у левој преткомори и суправентрикуларна тахикардија код новорођенчета

Љиљана Пејчић¹, Радмила Милеуснић-Миленовић², Марија Ратковић-Јанковић¹

¹Клинички центар Ниш, Клиника за деље интерне болести, Србија;

²Институт за неонатологију, Београд, Србија

САЖЕТАК

Увод Примарни тумори срца код деце су ретки и већина њих се дијагностикује до краја прве године живота. Најчешће су то рабдомиоми који имају склоност ка регресији. Учесталост аритмија код ових тумора није позната и њихова појава углавном зависи од величине и локализације самог тумора.

Приказ болесника Приказујемо женско новорођенче са суправентрикуларном тахикардијом (СВТ), која је регистрована пренатално. Порођај је завршен хитним царским резом, при чему није урађена фетална ехокардиографија, нити је покушана медикаментна конверзија тахикардије. Електрокардиограм је потврдио тахикардију (260/мин.) са уским QRS комплексом која је конвертована у синусни ритам након треће интравенске болус дозе аденозин-5'-трифосфата. Синусни ритам се тешко одржавао због упорне СВТ. На ехокардиографском прегледу, после конверзије у синусни ритам, виђено је хиперехогено, добро ограничено ткиво у

левој преткомори, фиксирано за дистални део преткоморске преграде. Није било других морфолошких промена. СВТ је успешно је заустављена болусом, а затим интравенском инфузијом амиодарона. Тахикардија је надале успешно контролисана перорално комбинацијом амиодарона и бета блокатора. Током једногодишњег ехокардиографског праћења, које је спровођено једном месечно, регистрована је постепена и потпуна регресија тумора. Тахикардија се није више поновила и девојчици је укинута терапија.

Закључак Регресија тумора и добра дугорочна прогноза код нашег болесника указују на то да се вероватно радило о малом, али „лоше постављеном“ рабдомиому, који је узроковао СВТ код фетуса и након рођења код новорођенчета. Прогноза и исход болести код оваквих болесника директно зависе од правовремене дијагнозе и брзог и адекватног лечења тумором узрокованих аритмија опасних по живот.

Кључне речи: тумори срца; суправентрикуларна тахикардија; рабдомиом; новорођенче