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Andrzej Szydłowski[1], Jacek Kusa[2], Katarzyna Gruszczyńska[3], Agnieszka Skierska[1], Ewa Moric-Janiszewska[4], Zbigniew Olczak[5], Anna Szydłowska[1]

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A giant heart tumour diagnosed accidentally in a 6-year-old boy with wide QRS complex tachycardia

Olbrzymi guz serca rozpoznany przypadkowo u 6-letniego chłopca z częstoskurczem z szerokimi zespołami QRS

[1] Department of Paediatric Cardiology, Upper Silesian Child Health Centre, Katowice, Poland
[2] Department of Paediatric Cardiology, Medical University of Silesia, Katowice, Poland
[3] Department of Diagnostic Imaging, Department of Radiology and Nuclear Medicine, Faculty of Medical Sciences in Katowice, Medical University of Silesia, Katowice, Poland
[4] Department of Biochemistry, Faculty of Pharmaceutical Sciences in Sosnowiec, Medical University of Silesia, Sosnowiec, Poland
[5] Department of Diagnostic Imaging and Interventional Radiology, The Independent Public Clinical Hospital No. 6, Upper Silesian Child Health Centre, Katowice, Poland
Correspondence: Andrzej Szydłowski, Upper Silesian Child Health Centre in Katowice, Medyków 16, 40-752 Katowice, Poland, tel.: +48 32 207 18 57, e-mail: szydlowskil@interia.pl

ORCID iDs
1. Andrzej Szydłowski https://orcid.org/0000-0002-0883-5454
2. Jacek Kusa https://orcid.org/0000-0003-2607-1491
3. Katarzyna Gruszczyńska https://orcid.org/0000-0002-0863-4492
4. Ewa Moric-Janiszewska https://orcid.org/0000-0003-1181-2824
5. Anna Szydłowska https://orcid.org/0000-0001-8288-5487

Abstract

The paper presents a case report of a 6-year-old boy with broad QRS tachycardia, who was accidentally diagnosed with a cardiac tumour. The tachycardia occurred twice, 10 months apart, and was quickly interrupted in a night medical care unit by performing a carotid sinus massage. Electrocardiography was performed, but its low quality did not allow for a detailed analysis, except for the heart rate, which was 180 bpm. Cardiological diagnosis was performed in a reference centre. No laboratory abnormalities were found. Holter ECG recorded only 374 single ventricular beats, while a routine echo revealed a very large 4.2 × 3.1 cm tumour in the interventricular septum, which did not impede intracardiac blood flow. The presence of the tumour was confirmed by magnetic resonance imaging of the heart, in which a fibroma was suspected. A beta-blocker (metoprolol) was included in the treatment and further cardiac monitoring was recommended. The boy was discharged home with a recommended follow-up in 2 months. He did not report for the appointment, while his parents requested for full imaging documentation.

Keywords: tachycardia, heart tumour, echocardiography, magnetic resonance imaging

Streszczenie

W pracy przedstawiono historię choroby 6-letniego chłopca z częstoskurczem o szerokich zespołach QRS, u którego przypadkowo wykryto guz w sercu. Częstoskurcz wystąpił dwukrotnie w odstępie 10 miesięcy i został szybko przerwany w gabinecie nocnej pomocy medycznej poprzez wykonanie masażu zatoki szyjnej. Wykonano zapis elektrokardiograficzny (EKG), ale z uwagi na jego niską jakość nie nadawał się on do szczegółowej analizy, określono jedynie częstość rytmu na 180/min. Diagnostykę kardiologiczną wykonano w ośrodku referencyjnym. Nie stwierdzono nieprawidłowości w badaniach laboratoryjnych. W badaniu EKG metodą Holtera zarejestrowano jedynie 374 pojedyncze pobudzenia komorowe, natomiast w trakcie rutynowego badania echokardiograficznego uwidoczniono w przegrodzie międzykomorowej bardzo duży guz o wymiarach 4,2 × 3,1 cm, nieutrudniający wewnątrzsercowego przepływu krwi. Obecność guza potwierdzono w rezonansie magnetycznym serca, w którym wysunięto podejrzenie włókniaka. Do leczenia włączono beta-adrenolityk (metoprolol) i zalecono dalszą obserwację kardiologiczną. Chłopiec został wypisany do domu z zaleceniem kontroli za 2 miesiące. Nie zgłosił się na wizytę w terminie, a rodzice poprosili o wydanie pełnej dokumentacji badań obrazowych.

Słowa kluczowe: częstoskurcz, guz serca, echokardiografia, rezonans magnetyczny

INTRODUCTION

Cardiac malignancies are very rare and can develop as primary tumours, i.e. arise directly from the heart, or as a metastasis from another site. Their prevalence estimated in autopsy studies is 0.002–0.33%[1].
Primary cardiac tumours are mostly benign, although they can exhibit a locally malignant nature due to their location or size, when they obstruct blood flow or movement of the heart valves. They include rhabdomyomas, fibromas, lipomas, myxomas, and teratomas. Rhabdomyoma is the most common cardiac tumour in children, whereas myxoma is the most common neoplasm in adults[2–4].
Rhabdomyoma can be diagnosed already in utero and is one of the manifestations of tuberous sclerosis complex (TSC), an autosomal dominant disease[2,5]. In recent years, TSC has been shown to be caused by a mutation in the *TSC1* or *TSC2* genes. This molecular defect disrupts cell development, which promotes tumorigenesis not only in the heart muscle, but also in the liver, skin, fundus of the eye, as well as in the lungs and brain. Café-au-lait spots identified by a family physician or a paediatrician on the child's skin should always arouse vigilance and prompt the search for the clinical features of TSC. It is known that while cardiac rhabdomyomas are capable of spontaneous regression, they can lead to epilepsy and developmental delay. The introduction of mTOR (mammalian target of rapamycin) pathway inhibitors, i.e. rapamycin and everolimus, which inhibit the development of cancer cells, was a breakthrough in the treatment of TSC. In Poland, the Children's Memorial Health Institute in Warsaw is currently conducting the world's first project to assess the efficacy of modern rapamycin in drug-resistant epilepsy in children with TSC[6,7].
Fibroma, which may occur in Gorlin syndrome (multiple endocrine adenomatosis), is another most common benign heart tumour in children under 2 years of age. It is most often located in the interventricular septum or in the left ventricular wall, although it can also develop in the right ventricle and right atrium. It is usually a single well-delineated tumour with regular, smooth surface. In some cases, it can reach enormous sizes already during foetal life, leading to foetal death as a consequence of significant haemodynamic disturbances that obstruct intracardiac blood flow. It is usually asymptomatic in children, except when it causes arrhythmias or obstructs intracardiac blood flow. Other primary cardiac tumours are extremely rare in children[1,4,8].
Cardiac tumours are usually diagnosed in children accidentally, except when the child develops typical clinical manifestations of TSC and epilepsy. Then the patient is referred by the neurologist for a cardiological consultation to rule out/confirm cardiac tumour[9]. Echocardiography, computed tomography (CT) and magnetic resonance imaging (MRI) of the heart are the main diagnostic tools. Heart MRI is a reference method in the diagnosis of cardiac tumours. It allows for a reliable assessment of the location and nature of the tumour tissue, and is also useful in the differentiation of the tumour, e.g. with a thrombus or a vegetation. However, the final diagnosis is always based on histopathological analysis of a section of the resected tumour or post-mortem findings[10,11]. Large heart tumours reduce the capacity of the left, right or both ventricles, resulting in circulatory failure, which may be an indication for complete or partial tumour resection. Postoperative tumour recurrence is reported in about 6% of patients. Primary and metastatic malignant tumours are associated with unfavourable prognosis from the moment of diagnosis[12,13].

CASE REPORT

A 6-year-old boy was admitted by a paediatrician in a night medical care unit due to increased heart rate during physical activity, chest pain and malaise. A similar incident had occurred 10 months earlier, but resolved spontaneously. This time, electrocardiography (ECG) was performed as part of night medical care, supraventricular tachycardia was diagnosed with a heart rate of about 180 bpm, and a carotid artery massage was successfully performed, resulting in tachycardia resolution. The patient was transferred to the paediatric ward in the regional hospital for diagnosis, but the parents had the boy discharged on day 2 and reported to the hospital emergency department (ED) at the Upper Silesian Child Health Centre in Katowice, and then urgently to the Department of Paediatric Cardiology in the same hospital. On admission, the child was in good condition, circulatory and respiratory efficient, with regular heart rate of 84 bpm, a venous humming murmur with changing loudness depending on the position of the head, disappearing on jugular compression, correctly accented heart sounds, and symmetric heart rate. Blood pressure was 120/68 mm Hg. The provided copy of the night medical care ECG indicated a wide-complex tachycardia of approximately 180 bpm, but detailed interpretation of the ECG record was impossible due to its poor technical quality.
Laboratory findings were normal. High sensitivity troponin T (hs TnT) was 5.12 pg/mL (normal), free thyroxine (fT4) was 1.38 ng/dL (normal), and inflammatory markers were negative. ECG showed a leading sinus rhythm of about 91 bpm, normal heart axis, PQ interval of 150 ms, QRS of 80 ms; QTc of 394 ms, non-specific ST segment depression by 1 mm in leads II, III, aVF and negative T wave in leads II, III and V4–V5. A 24-hour Holter ECG showed 374 premature, single, monomorphic ventricular beats (Fig. 1).
Two-dimensional echocardiography (2DE) revealed a giant 4.2 × 3.1 cm tumour with a surface area of 12 cm^2, communicating with the interventricular septum, and protruding into the right and left ventricle (Fig. 2). Three-dimensional echocardiography (3DE) showed that the tumour was located within the interventricular septum and was well delineated (Fig. 3).
It was decided to extend the imaging diagnosis with MRI, which confirmed the presence of the 3.84 × 5.07 × 4 cm tumour in the interventricular septum, which protruded into

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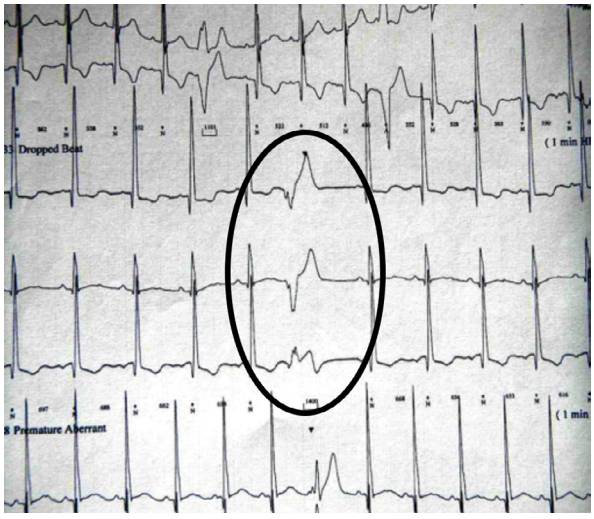

Fig. 1. ECG recording. Leading sinus rhythm disturbed by single ventricular beats (circled)

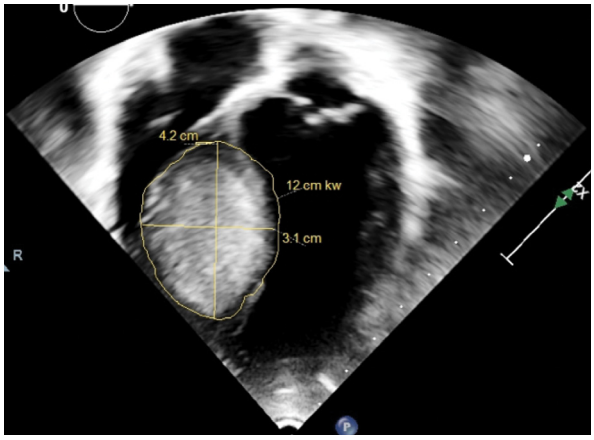

Fig. 2. Two-dimensional echocardiography, four-chamber projection. A 4.2 × 3.1 cm tumour with a surface area of 12 cm², protruding into the right and left ventricle is present in the interventricular septum

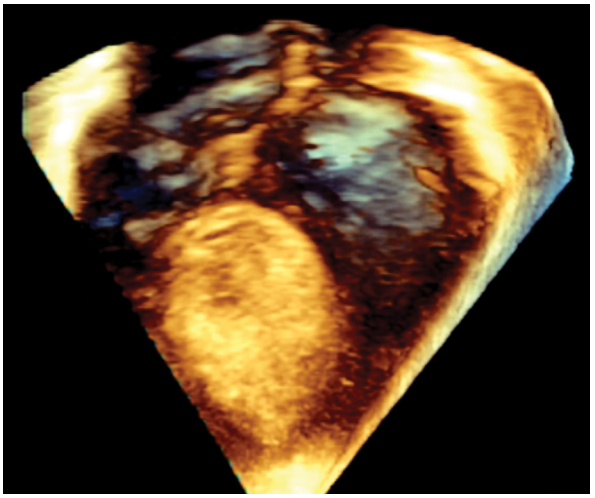

Fig. 3. Three-dimensional echocardiography, four-chamber projection. A tumour protruding into both ventricles, not obstructing the blood flow within the ventricles is seen in the interventricular septum

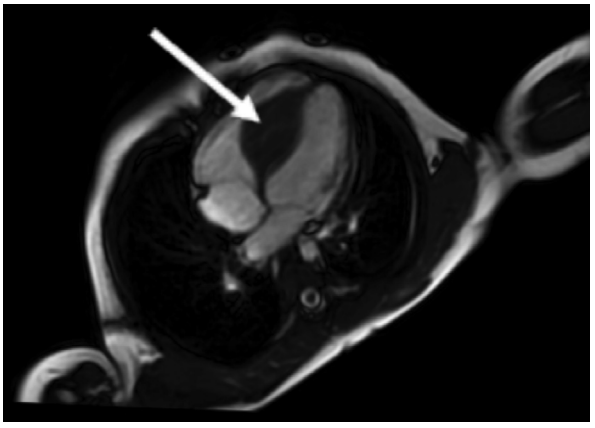

Fig. 4. MRI of the heart., a 3.84 × 5.07 × 4 cm tumour, most likely a fibroma, is present in the wall of the interventricular septum

both the right and the left ventricle, reducing their volume. The lesion had regular and smooth contours, did not impair cardiac function, and the left ventricular ejection fraction was normal (LVEF = 61.6%). The MRI confirmed the cardiac tumour, indicating a benign, fibroma-like lesion (Fig. 4). A beta-blocker (metoprolol) was started at 25 mg/day for ventricular arrhythmia. The boy was discharged home with a recommendation to report for a follow-up at the cardiology clinic in 2 months. The parents did not report with the child at the arranged date, but instead requested full imaging documentation. They most likely decided to consult or seek further treatment of their child in another centre.

DISCUSSION

Benign heart tumours are mostly asymptomatic, but they can cause arrhythmias, which is the reason for visits to outpatient cardiac clinics. In the described boy, tachycardia was the first symptom of the disease. Tachycardia is an alarming symptom and requires detailed diagnosis to identify its aetiology and choose further treatment. In the presented case, the tachycardia diagnosed in the night medical care setting was about 180 bpm, but the poor technical quality of ECG did not allow for determining whether it was wide-complex ventricular tachycardia or atrial tachycardia with aberrant conduction. Distinguishing between these two types of tachycardia is often very difficult and requires a 12-lead Holter ECG for a thorough analysis of P-waves, QRS duration, and precordial R- and S-waves[14]. Unfortunately, the personnel of emergency rooms or even emergency departments not always remember that a correct ECG recording often determines further treatment, as it may turn out that it is the only record on the basis of which the patient can be qualified for further treatment, including radiofrequency (RF) ablation of an accessory pathway or an arrhythmogenic focus.

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Cardiac tumour can be diagnosed with echocardiography (foetal and postnatal), including two- (2DE) and three-dimensional (3DE) projections. Cardiac MRI, which can determine the type of abnormal tumour tissue with high probability, is recommended to confirm the diagnosis. This was the case of our patient, in whom 2DE and 3DE echocardiography allowed for a detailed identification of the tumour mass and its location in the interventricular septum, including its protrusion into the right and left ventricle, while the MRI determined the structure of abnormal tumour tissue, which corresponded most to a fibroma (Figs. 2–4). Paediatric cardiac fibromas with a confirmed histopathological picture that are described in the world literature resemble the tumour identified in our patient. At this stage, the management strategy for the described patient assumes a wait-and-see attitude and further cardiac follow-up. Recurrent cardiac arrhythmias, especially tachycardias, are the main problem. The treatment with a beta-blocker seems to be effective, as no arrhythmias were found in follow-up. Frequent and repeated Holter ECG and clinical follow-up are the primary diagnostic methods for arrhythmias. The boy's episodes of rapid heart rate were very early and correctly identified by his parents. Unfortunately, they did not report the boy to the cardiology clinic on the appointed date, with no information on the reason for their resignation from the follow-up visit. Instead, they requested the patient's medical records, most likely for the purposes of further follow-up and treatment in another centre, which is not unusual or unheard of in such a situation.

CONCLUSIONS

Primary cardiac tumours are very rare in children and their diagnosis is usually accidental. Fibroma is the second most common benign heart tumour in children after rhabdomyosarcoma. The differential diagnosis of any intracardiac mass should include a tumour, a thrombus, and a vegetation. Complementary CT or MRI of the heart is a very useful diagnostic tool in the differentiation of cardiac tumours.

Conflict of interest

The authors do not declare any financial or personal links to other persons or organisations that could adversely affect the content of this publication or claim rights thereto.

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