

Multimodality imaging of a subtotally obstructive right ventricular myxoma in an asymptomatic 10-year-old girl

Gesa Wiegand, Ludger Sieverding, Ulrich Kramer¹, Susanne Haen², Michael Hofbeck

Department of Pediatric Cardiology, University Children's Hospital, ¹Department of Diagnostic and Interventional Radiology, ²Department of General Pathology, Institute of Pathology and Neuropathology, Eberhard-Karls-University, Tuebingen, Germany

ABSTRACT

A 10-year-old girl presented with a supraventricular tachycardia. A heart murmur was detected during a clinical examination; therefore, echocardiography was performed. This revealed a giant right ventricular myxoma that subtotally obstructed the right ventricular outflow tract. A myxoma in the right ventricle is uncommon in children. This rare intracardiac tumor was examined using multimodality imaging, and the patient underwent surgical resection. The pathologic features confirmed our suspicion and revealed an encapsulated tumor with myxomatoid stroma containing focal hemorrhages.

Keywords: Pediatric cardiology, cardiac tumor, myxoma, right ventricle

CASE REPORT

A 10-year-old girl was referred for an episode of sustained tachycardia while diving in a swimming pool. The electrocardiogram registered supraventricular tachycardia at a rate of 250 beats/min, which was successfully treated with adenosine. A heart murmur was detected; therefore, transthoracic and transesophageal echocardiography was performed. This revealed a smooth tumor located below the tricuspid valve that occupied almost the entire cavity of the right ventricle and obstructed its outflow tract [Figure 1a and b]. Cardiac magnetic resonance imaging (MRI) confirmed these results; the relatively mobile tumor protruded into the right ventricular outflow tract and appeared to be pedunculated with an attachment to the right ventricular free wall [Figure 2a and b]. While the function of the tricuspid valve was normal, the septum showed paradoxical motion. Structural inhomogeneities of the tumor, which represented intratumoral hemorrhages, were obvious upon MRI [Figure 2a and b]. These hemorrhages were later examined by histopathology. Preoperative cardiac catheterization confirmed

obstruction of the right ventricular outflow tract [Figure 3a and b] and identified vascular supply of the tumor by an abnormal vessel originating from the right coronary artery.

While the patient was on cardiopulmonary bypass, surgical resection was performed through a right atrial approach and revealed a $5.5 \times 3.7 \times 3.1$ cm smooth elastic tumor with gelatinous content and distinct hemorrhages. Histopathology confirmed the diagnosis of a capsulated myxoma [Figure 4]. Postoperative recovery was uneventful, and there was no recurrence of the tumor during 3.5-year follow-up. Supraventricular tachycardias recurred; therefore, an electrophysiological examination was performed, which revealed atrioventricular node reentry tachycardia that was unrelated to the cardiac myxoma. This was treated by cryomodulation of the slow pathway. During 2 years of follow-up, the patient did not exhibit symptoms of tachycardia, and she resumed competitive biathlon sports.

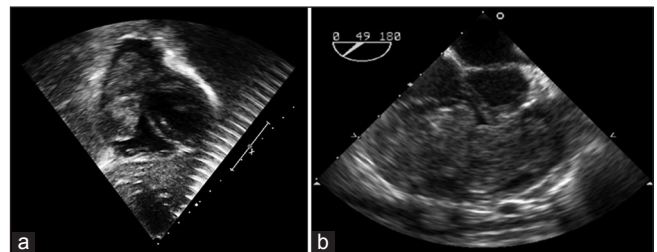


Figure 1: The right ventricular outflow tract in a transthoracic echocardiographic subcostal view (a) and a transesophageal echocardiographic view at 49° (b)

Access this article online	
Quick Response Code:	Website: www.annalspc.com
	DOI: 10.4103/0974-2069.132519

Address for correspondence: Dr. Gesa Wiegand, Department of Pediatric Cardiology, University Children's Hospital, Hoppe-Seyler-Strasse 1, Tuebingen - 72076, Germany. E-mail: gesa.wiegand@med.uni-tuebingen.de

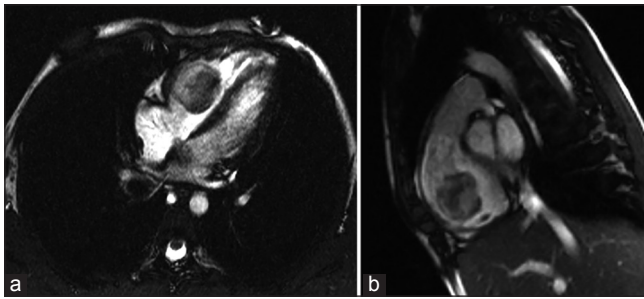


Figure 2: Cardiac MRI of the axial (a) and sagittal (b) view of the right ventricle. MRI = Magnetic resonance imaging

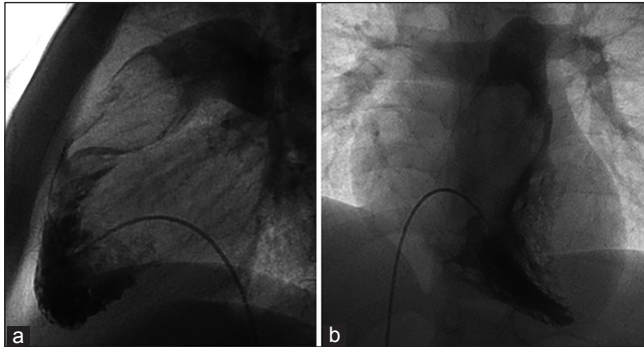


Figure 3: Angiography of the right ventricle in the lateral (a) and anteroposterior (b) projection



Figure 4: Macroscopic presentation of the encapsulated myxoma

DISCUSSION

Primary cardiac tumors are rare in children; the most common are rhabdomyomas and teratomas, whereas myxomas and fibromas are less common.^[1] Myxomas occur in all age groups, most frequently between the third and sixth decades of life, and there is a female predominance.^[2] About 75% of myxomas develop in the left atrium in both children and adults; 15-20% of myxomas develop in the right atrium and myxomas rarely arise in the ventricles.^[2,3] Myxomas that arise in the right ventricle and obstruct the right ventricular

outflow tract are extremely rare in children, and there are only isolated reports of such cases.^[4-6]

The macroscopic appearance of myxomas is most often polypoid; however, in rare cases, such as that presented here, they have a smooth surface. Many large blood vessels derived from the subendocardium are often located at the base of a myxoma. As in the patient presented here, myxomas often contain hemorrhages and cysts, as well as foci of extramedullary hematopoiesis, which may cause hemorrhages. In rare cases, such tumors contain regions of calcification, making them detectable by chest radiography^[2] and MRI.

Depending on the size and location of the myxoma, patients can be asymptomatic or exhibit various degrees and types of cardiac insufficiency.^[2] Typically, atrial myxomas protrude into and obstruct the atrioventricular valves and can cause syncopal episodes and sudden death. Transthoracic echocardiography is useful to describe the size and location of the tumor, whereas transesophageal echocardiography is helpful to determine where the tumor is attached.

Our patient presented owing to a first episode of supraventricular tachycardia. However, in this case, the supraventricular tachycardia was probably unrelated to the tumor and its right ventricular outflow tract obstruction. Therefore, it underlines the requirement to perform echocardiography to exclude structural heart disease in all children with cardiac arrhythmias.

REFERENCES

1. Bielefeld KJ, Moller JH. Cardiac tumors in infants and children: Study of 120 operated patients. *Pediatr Cardiol* 2013;34:125-8.
2. Reynen K. Cardiac myxomas. *N Engl J Med* 1995;333:1610-7.
3. Reddy SN, Sunil GS, Kumar RK. Surgical removal of a left ventricular myxoma in an infant. *Ann Pediatr Cardiol* 2013;6:179-81.
4. Buheitel G, Hofbeck M, Singer H, Lindemann Y, von der Emde J. Kardiale Myxome bei Kindern und Jugendlichen. *Monatsschr Kinderheilkd* 1996;144:1087-91.
5. Kern JH, Aguilera FA, Carlson DL, Galantowicz M. Right ventricular myxoma obstructing the right ventricular outflow tract. *Circulation* 2000;102:E14-5.
6. Kumagai K, Sai S, Endo M, Tabayashi K. Right ventricular myxoma obstructing the pulmonary artery during early childhood. *Gen Thorac Cardiovasc Surg* 2008;56:351-3.

How to cite this article: Wiegand G, Sieverding L, Kramer U, Haen S, Hofbeck M. Multimodality imaging of a subtotally obstructive right ventricular myxoma in an asymptomatic 10-year-old girl. *Ann Pediatr Card* 2014;7:158-9.

Source of Support: Nil, **Conflict of Interest:** None declared