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LARGE MALIGNANT CARDIAC TUMOR IN A 7-YEAR-OLD CHILD

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Primary malignant cardiac tumors are extremely rare in childhood and among them, sarcomas are most frequent. We present clinical, echocardiographic and magnetic resonance imaging findings of a 7-year-old child in whom an invasive malignant cardiac tumor was detected at the level of the right atrium and ventricle, with the presence of pulmonary metastases at the time of diagnosis.

Descriptors: HEART NEOPLASMS – complications, diagnosis, radiography; CHILD

INTRODUCTION

Primary cardiac tumors are extremely rare, especially in the pediatric age group. Autopsy studies in children report an incidence of 0.027% to 0.08% and echocardiographic studies report an incidence of 0.0017% to 0.003% of the total number of hospital admissions. Primary malignant cardiac tumors are very uncommon in children, being mostly represented by sarcomas. Secondary cardiac tumors, although more frequently found in adults, are also extremely rare in children (1-6). Rhabdomyosarcoma is a malignant tumor of mesenchymal origin, which infiltrates the myocardium diffusely, invades the cardiac cavities and the pericardium, and has a poor prognosis (7, 8).

We present clinical, echocardiographic and magnetic resonance imaging findings of the invasive malignant cardiac tumor in a 7-year-old boy. The tumor was detected at the level of the right heart cavities.

CASE REPORT

A 7-year-old male patient, with a recent history of acute pneumonia (when systolic murmur and enlargement of the heart were found on x-ray), was hospitalized in our service for suspicion of congenital heart disease. On admission, the child was in good condition, completely asymptomatic, with normal height and mild weight deficit (10th percentile of body mass index). On physical examination, he presented skin paleness, arrhythmic cardiac sounds, heart rate of 80 beats

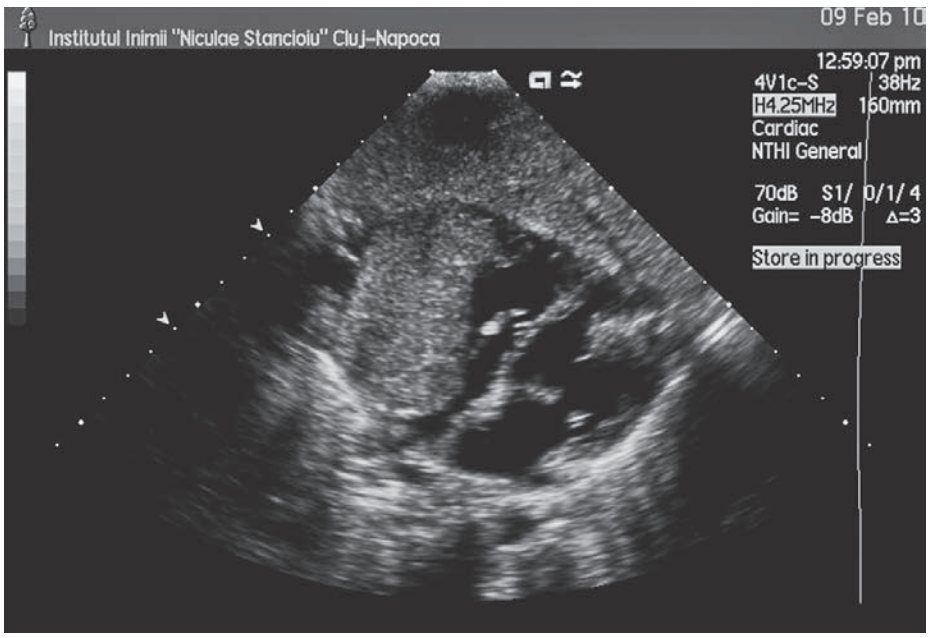

Figure 1. *Echocardiography: giant, non-homogeneous tumor located in the anterior and lateral wall of the right ventricle and right atrium*
Slika 1. *Ehokardiografija: veliki nehomogeni tumor u prednjem i bočnom zidu desne klijetke i desne pretklijetke*

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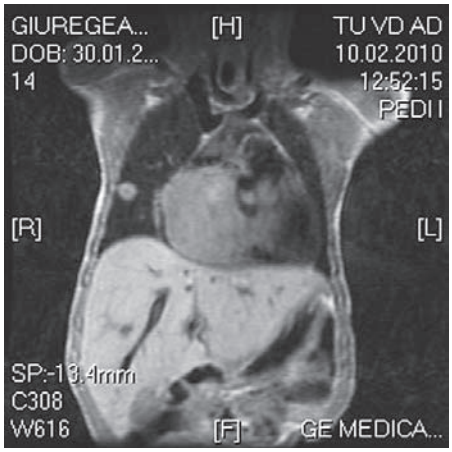

Figure 2. *Cardiac MRI: giant cardiac tumor*
Slika 2. *MRI srca: veliki srčani tumor*

per minute, loud 1st and 2nd heart sounds, third degree systolic murmur and enlargement of the liver.

Electrocardiogram evidenced the presence of a right bundle branch block and extrasystolic ventricular arrhythmia. Echocardiography showed a non-homogeneous polypoid mass, measuring 85x56 mm, located in the anterior and lateral wall of the right ventricle and right atrium, causing compression at the level of the right atrium and of the receiving chamber of the right ventricle. The ventricular septum had abnormal motion. The inferior vena cava was dilated, without respiratory variations. A low amount of pericardial collection was also found. This mass was interpreted as a giant cardiac tumor (Figure 1).

Cardiac magnetic resonance imaging (MRI) evidenced an aspect suggestive of malignant tumor (Figure 2).

Chest radiography evidenced cardiac enlargement and pulmonary opacities, varying from several millimeters to 2.5 cm in size (Figure 3). These lung metastases were confirmed by chest MRI.

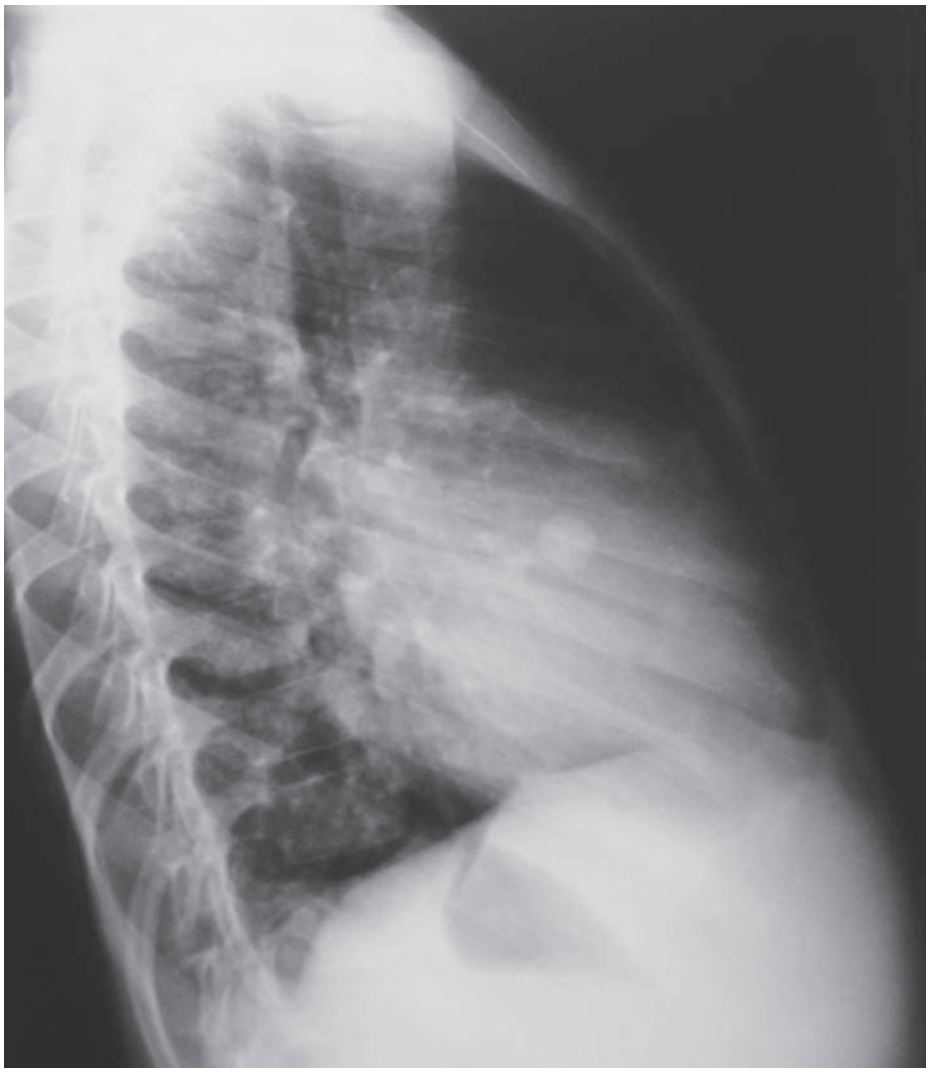

Figure 3. *Chest radiography: pulmonary metastases*
Slika 3. *Rendgenska slika toraksa: plućne metastaze*

Laboratory findings showed mild inflammation (ESR=20 mm/h), anemia (hemoglobin=10.7g/dL,hematocrit=33%) and increased lactate dehydrogenase value (735 IU/L).

After the diagnosis, the patient was transferred to Pediatric Oncology for specialized treatment. Biopsy of the cardiac mass was not performed because of the risk of the procedure involved.

DISCUSSION

Most primary cardiac tumors in the pediatric age group are benign. Fewer than 10% of these tumors are malignant. The most frequent malignant tumors are angiosarcoma, rhabdomyosarcoma, mesothelioma, fibrosarcoma, lymphoma and liposarcoma (1, 2, 8, 9). Rhabdomyosarcoma is the most common malignant cardiac tumor of the child. This tumor extends to the heart cavities, invades the valves and the pericardium, and is more frequently found in the right cavities (10). It seems that tumors from the right heart have a higher risk of being malignant (11-13). Clinical features depend on the tumor size and localization and include the classic triad: symptoms that show heart obstruction, signs that reveal the presence of embolisms and general signs, including fever, weight loss and myalgia (7, 12, 14). Arrhythmias and pericardial collection may also be present (12). In our patient, clinical finding was extremely poor, including only systolic murmur and ventricular extrasystoles.

Transthoracic or transesophageal ultrasound examination confirms the diagnosis, shows the location and the possible complications (3, 15). In this case, the tumor was very large (85x56mm), causing compression at the level of the right atrium and of the receiving chamber of the right ventricle.

The imaging examination is completed by cardiac MRI, which provides additional data on tumor extension, detection of the possible metastases, and tumor resectability (3, 5, 7). There are several criteria for malignancy that can be evidenced by MRI: rapid growth, location in the right cavities, invasion of the myocardium and cavities, extension to the pulmonary veins, presence of metastases, and hemorrhagic pericardial collection (13). In the reported case, four of the men-

tioned criteria were present, the aspect being suggestive of rhabdomyosarcoma.

At presentation, rhabdomyosarcomas are usually large and surgical removal is very frequently impossible (7, 16).

The treatment is usually chemotherapy. The results are dismal because of the large tumor volume and the presence of metastases at the time of diagnosis. Patient survival ranges between 6 months and 2 years, with a mean of 9.6 months, depending on the histologic degree of malignancy (11, 17). A therapeutic alternative in chemotherapy-resistant cases without metastases is heart transplantation (7).

In this case, chemotherapy was initiated, but chest MRI after 3 months of treatment was unfortunately unchanged compared to the initial evaluation. The clinical condition was worsening, so biopsy was not considered.

CONCLUSION

Rhabdomyosarcoma is a very rare malignant cardiac tumor, which usually has large size and metastases at the time of diagnosis. The prognosis is poor.

Authors declare no conflict of interest.
Autori izjavljuju da nisu bili u sukobu interesa.

REFERENCES

1. Marx GR, Moran AM. Cardiac Tumors, in Moss and Adams' Heart Disease in Infants, Children and Adolescents: Including the Fetus and Young Adult. 7th ed. Philadelphia: Lippincot Williams & Willkins, 2008;1480-95.
2. Bojan V, Dragan B, Svetozar S, Miroslav B, Dejan V. Slobodan D. Rhabdomyosarcoma of the heart. Herz. 2006;31:798–800.
3. Fresneau B, Oberlin O, Brugières R, Valteau-Couanet D, Patte C. Les tumeurs cardiaques primitives malignes de l'enfant et de l'adolescent. Arch Péd. 2010;17:495-501.
4. Odim J, Reehal V, Laks H, Mehta U, Fishbein MC. Surgical pathology of cardiac tumors. Two decades at an urban institution. Cardiovasc Pathol. 2003;12:267–70.
5. Uzun O, Wilson DG, Vujanic GM, Parsons JM. Cardiac tumours in children. Orphanet J Rare Dis. 2007;2:11.
6. Manasia R. Tumori cardiace. In: Manasia R, editor. Tratat elementar de cardiologie pediatrica. Cluj-Napoca: Todesco, 2002;189-92.
7. Neragi-Miandoab S, Kim J, Vlahakesy GJ. Malignant tumours of the heart: A review of tumour type, diagnosis and therapy. Clin Oncol. 2007;19: 748-56.
8. Lima RC, Mendes A, Bezzera E, Oliveira W. Surgical treatment of primary cardiac rhabdomyosarcoma. Rev Bras Cir Cardiovasc. 2009;24:242-4.
9. Manasia R, Grigorescu-Sido P, Barsan M, et al. Cardiac fibroma in child – consideration regarding a clinical case. The Heart. 1997;1:82-4.
10. Selvaraj T, Kapoor PM, Kiram U. Large rhabdomyosarcoma of the right ventricle obstructing tricuspid valve, pulmonary valve and left ventricular outflow tract. Ann Card Anaesth. 2009;12:81-2.
11. Thomas-de-Montpréville V, Nottin R, Dulmet E, Serraf A. Heart tumors in children and adults: clinicopathological study of 59 patients from a surgical center. Cardiovasc Pathol. 2007;16:22–8.
12. Menga Q, Laib H, Limac J, Tongc W, Qiana Y, Lai S. Echocardiographic and pathologic characteristics of primary cardiac tumors: a study of 149 cases. Int J Cardiol. 2002;84:69–75.
13. Villacampa VM, Villarreal M, Ros LH, Álvarez R, Cózar M. Fuertes MI. Cardiac rhabdomyosarcoma: diagnosis by MR imaging. Eur Radiol. 1999; 9:634-7.
14. Fernandes F, Soufen HN, Ianni BM, Arteaga E, Ramires FJA, Mady C. Primary neoplasms of the heart. Clinical and histological presentation of 50 cases. Arq Bras Cardiol. 2001;76:235-7.
15. Skopin II, Serov RA, Makushin AA, Sazonenkov MA. Primary rhabdomyosarcoma of the right atrium. Interact Cardiovasc Thor Surg. 2003;2: 316–8.
16. Tavil Y, Turkoglu S, Tacoy G, Cemri M. Huge biatrial cardiac rhabdomyosarcoma resulting in bilateral atrioventricular valve obstruction. Cardiovasc Pathol. 2006;15:354–5.
17. Bouzas-Mosquera A, Flores-Rios X, Aldama G. Primary cardiac rhabdomyosarcoma causing obstruction to the right ventricular outflow. Eur J Cardiol. 2007;8:406-7.

S a ž e t a k

VELIKI SRČANI MALIGNI TUMOR U 7-GODIŠNJEG DJETETA

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Primarni zloćudni tumori srca iznimno su rijetki u djetinjstvu, a među njima su najčešći sarkomi. Prikazujemo kliničke, ehokardiografske i nalaze magnetske rezonancije u sedmogodišnjeg djeteta kod kojega je invazivni zloćudni tumor srca otkriven na razini desne klijetke i pretklijetke, uz prisutnost plućnih metastaza u vrijeme postavljanja dijagnoze.

Deskriptori: NEOPLASMI SRCA – komplikacije, dijagnoza, radiografija; DIJETE

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