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CARDIAC RHABDOMYOSARCOMA DIAGNOSED BY ECHOCARDIOGRAPHY*

*F. Ayşenur Paç MD***, *Zuhal Akçören MD****, *Melda Çağlar MD*****
*Süheyla Özkutlu MD******

SUMMARY: Paç FA, Akçören Z, Çağlar M, Özkutlu S. (Cardiology and Pathology Units, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Cardiac rhabdomyosarcoma diagnosed by echocardiography. Turk J Pediatr 1995; 37: 257-261.

Primary, malignant, cardiac tumors are extremely rare in infants and children. Only a few cases of cardiac rhabdomyosarcoma have been reported in childhood. Echocardiography greatly increases the rate of correct diagnosis of cardiac tumors. There are some cases in the literature which were diagnosed by echocardiography and confirmed histopathologically.

In this report, a 13-year-old girl with the clinical findings of cardiac tamponade and who was diagnosed as having a cardiac tumor by two-dimensional echocardiogram is presented An echodense, solid tumor with irregular borders extending especially toward the left ventricular cavity from the apex was demonstrated on echocardiography. The diagnosis was confirmed histopathologically on postmortem examination. *Key words: cardiac tumor, rhabdomyosarcoma, echocardiography.*

Primary, malignant, cardiac tumors are extremely rare in infants and children. Only a few cases of cardiac rhabdomyosarcoma have been reported in children[1-4].

Echocardiography greatly increases the possibility of correctly diagnosing cardiac tumors. There have been some reported cases which were diagnosed by echocardiography and confirmed histopathologically[5, 6].

In this report, we present a 13-year-old girl with a malignant, cardiac tumor diagnosed by echocardiography which proved to be rhabdomyosarcoma on histopathologic examination.

Case Report

A 13-year-old girl was admitted to Hacettepe Children's Hospital with a two-month history dyspnea, fatique and edema. The physical examination revealed tachycardia, tachypnea, orthopnea, cyanosis, jugular venous distension,

* From the Cardiology and Pathology Units, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara.

** Associate Professor of Pediatrics and Pediatric Cardiologist, Atatürk University Faculty of Medicine, Erzurum.

*** Assistant Professor of Pediatrics, Hacettepe University Faculty of Medicine.

**** Professor of Pediatrics and Pediatric Pathologist, Hacettepe University Faculty of Medicine.

***** Professor of Pediatrics and Pediatric Cardiologist, HacettepeUniversity Faculty of Medicine.

edema in the extremities, and hepatomegaly. On auscultation, the heart sounds were muffled. The pulse rate was 120/min with marked pulsus paradoxus. The arterial blood pressure was 100/75 mmHg. The chest roentgenogram showed marked enlargement of the cardiac silhouette. There were low voltage QRS complexes in the EKG, which also displayed massive pericardial effusion. Emergency pericardiocentesis was performed with the diagnosis of cardiac tamponade, and 600 ml serosanguinous fluid was drained. The subsequent echocardiogram revealed the presence of an echodense mass in the ventricular apexes with irregular borders and extending to both ventricular cavities, especially to the left (Fig. 1). Minimal pericardial fluid was detected in the apical portion. B-bump mitral valve was demonstrated by M-mode echocardiography.

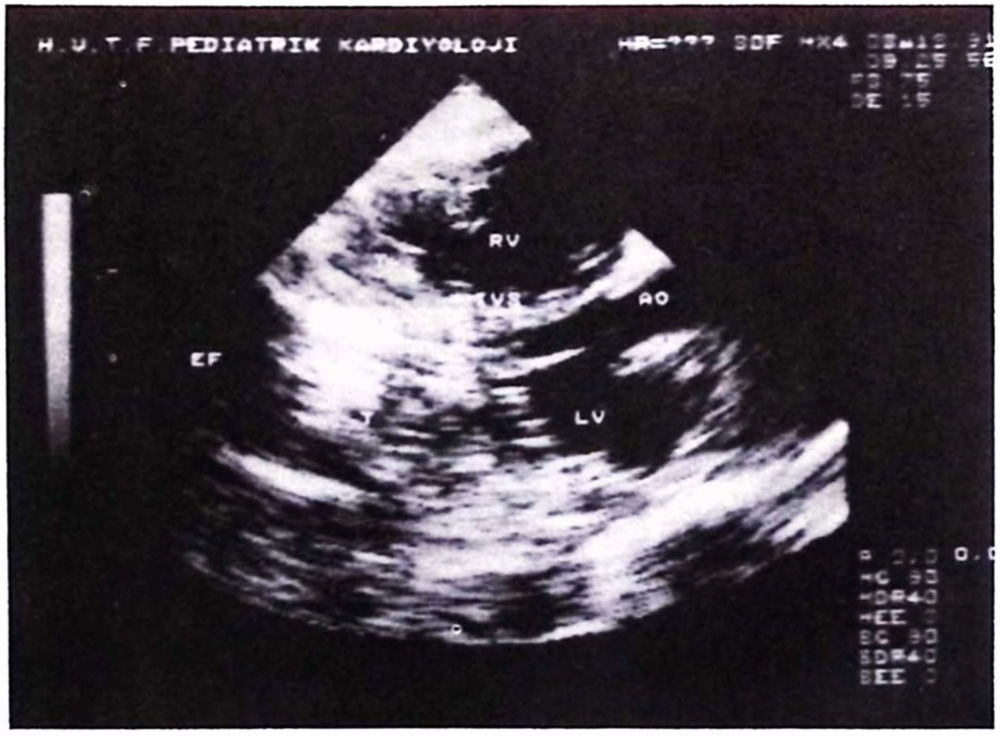

Fig. 1: Two-dimensional echocardiogram showed an echodense mass with irregular borders in the ventricular apexes and pericardial effusion. T: tumor, LV: left ventricle, RV: right ventricle, Ao: aorta, EF: effusion.

Congestive heart failure, pericardial effusion and the echocardiographic appearance suggested a cardiac tumor. The patient underwent tube drainage and pericardial biopsy. She was given digoxin for heart failure. She died two days after admission. Neither chemotherapy nor any further radiologic evaluation could be applied.

Postmortem examination revealed thickening of the myocardium, especially of the left ventricle, irregularity of the endocardium and budlike appearance of the visceral pericardium. Histopathologic study of the specimens showed hypertrophy

of the myocardial fibers and diffuse infiltration of the myocardium and pericardium by tumor cells. The tumor seemed to arise from the base of the left ventricular cavity and was mostly composed of undifferentiated round cells and spindle-shaped rhabdomyoblasts (Fig. 2). There were few strap-shaped cells. No cross-striation was identified even with phosphotungstic-acid hematoxylin stain. These microscopic findings were all compatible with embryonic-type rhabdomyosarcoma. Although the other organs could not be evaluated on postmortem examination, all histopathological and clinical findings strongly suggested that the tumor was a primary rahbdomyosarcoma of the heart.

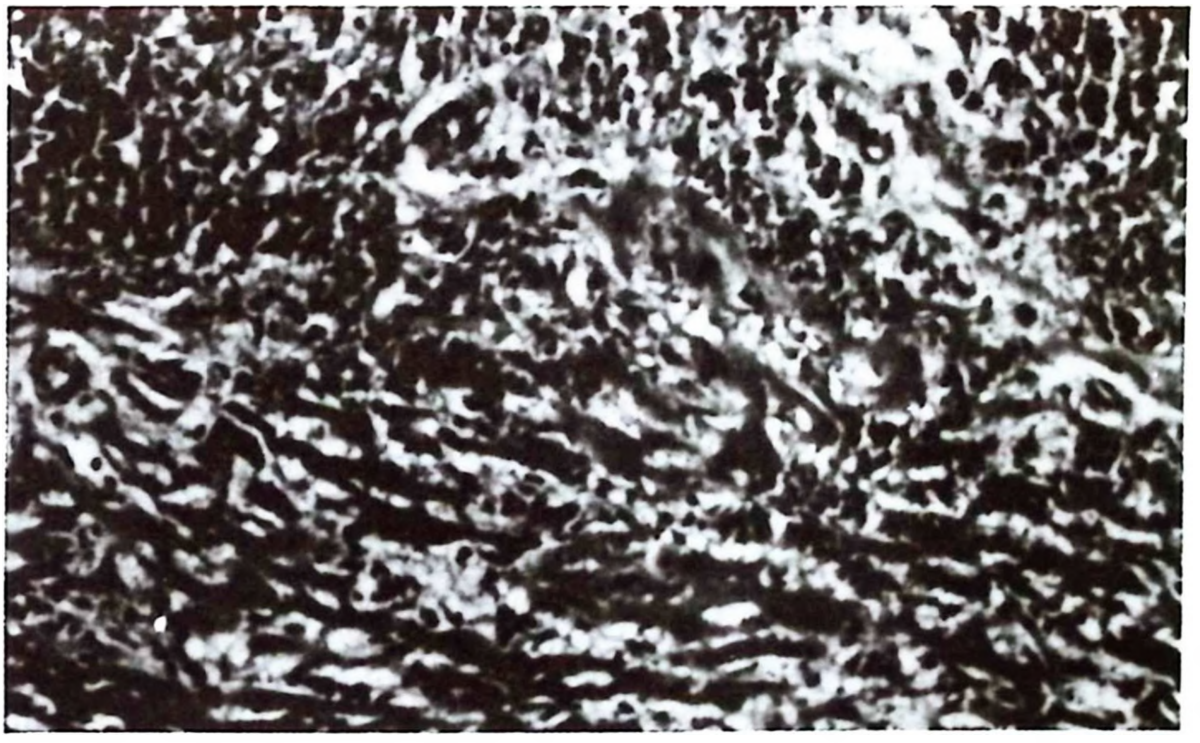

Fig. 2: Infiltration of the myocardium by round and a few spindle-shaped tumor cells (HE, x33).

Discussion

The incidence of primary cardiac tumors on autopsy is reported to be 0.0017-0.28 percent in children[1]. Only nine cases of primary cardiac tumor were described in 11,000 (0.08%) postmortem examinations[7]. Malignant, primary cardiac tumors account for approximately 25 percent of all primary cardiac tumors reported in the literature. In this rare entity, rhabdomyosarcoma is the second-most common neoplasm representing five percent of all heart tumors, but only 2.2 percent in children under 15 years of age[1]. Over a 62-year period, 16 children with primary cardiac tumors were seen at the Hospital for Sick Children in Toronto, and only one of these was rhabdomyosarcoma[3]. Putnam et al.[2] reported 21 primary cardiac sarcoma cases that underwent surgical resection in a 25-year period; two of these patients were children with rhabdomyosarcoma.

Cardiac rhabdomyosarcoma may arise in any cardiac chamber and with nearly equal frequency on the left and right sides of the heart[1,8]. In autopsy series, multiple cardiac sides were found to be involved in 60 percent of the patients, as in our case[9]. The pericardium was also involved in 50 percent of patients, usually by direct extension of the tumor[8].

Clinically the diagnosis is difficult to establish, not only due to its rarity but also because of its varied presentation[1]. The clinical manifestations vary considerably depending upon the histologic type, size and location of the tumor and the degree of involvement of the myocardium[7].

Pericardial effusion seen in primary or secondary malignant heart tumors may cause cardiac tamponade[7]. Pericardial effusion is rarely the first finding in extracardiac tumors[10]. The massive pericardial effusion of our case led us to consider malignant tumor. On the other hand, when pericardial effusion is minimal the echocardiographic diagnosis of cardiac tumor is suspicious, but surgery or necropsy reveals the diagnosis[11].

Rhythm disturbances, ST and T changes, and voltage supression, as in our case, may also be present.

Primary cardiac tumors with non-specific clinical manifestations, and telemetry findings often remain undetected during life, and the diagnosis is made at autopsy[3,5,11]. Cardiac catheterization and angiography, echocardiography, nuclear imaging, computerized tomography and magnetic resonance imaging (MRI) are diagnostic tools in cardiac tumors. Methods such as catheterization and angiography are not preferred because of the risk o rhythm disturbance and embolisation[6].

Echocardiography has become the definitive imaging method for the diagnosis of cardiac tumors in children; in most cases it has eliminated the need for further invasive studies in establishing the diagnosis[7]. Recently, cardiac-gated MRI has been used for the evaluation of cardiac tumors[12]. We could not use MRI for our patient because of her fast demise. In our patient, two-dimensional echocardiography revealed a noncapsulated solid tumor, protruding irregularly, especially toward the left ventricular cavity from the apex. These echocardiographic findings led us to a diagnosis of malignant cardiac tumor. Despite the absence of a full autopsy, microscopic findings such as infiltration of the myocardium and involvement of the pericardium by direct extension of tumor cells suggested primary cardiac rhabdomyosarcoma.

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