

Cardiac Rhabdomyoma in an Adult

M.Serdar KÜÇÜKOĞLU, Hüsnü YÜKSEL, Sinan ÜNER, Halil TÜRKOĞLU, Ç. BAYINDIR*

Department of Cardiology and Department of Cardiac Surgery, Institute of Cardiology, University of Istanbul,

*Department of Pathology, Istanbul Medical Faculty, University of Istanbul

ERİŞKİNDE KALP RABDOMİYOMASI

Kardiyak rabdomiyoma bebeklik ve çocukluğu çağında en sık görülen primer kalp tümörüdür. Selim hamartom olarak kabul edilmesine karşın miyokard kontraktilite bozukluğu, sağ veya sol ventrikül giriş veya çıkış yolu obstrüksiyonu veya ritm ve ileti bozuklukları oluşturabilir.

Bu yazıda konjestif kalp yetersizliği klinik tablosu ile başvuran ekokardiografi ile perikard efüzyonu ve sol ventrikül içi kitle tespit edilip sağ atrium biopsisi ile rabdomiyoma tanısı konan 42 yaşında bir olguyu sunmaktayız.

Rhabdomyomas are the most common primary cardiac tumors of infancy and childhood; however, they are extremely rare in adults (1). Although considered as benign hamartomas, they may cause serious clinical symptoms due to inflow or outflow obstruction as well as myocardial depression or arrhythmias (1-4).

In this report we describe a case of cardiac rhabdomyoma in an adult presenting with congestive heart failure, pericardial effusion and left ventricular mass.

CASE REPORT

A 42-year-old female complaining of exertional dyspnea, fatigue and palpitation for the last 18 month was admitted to our Institute. On physical examination, she exhibited signs of right and left sided heart failure. A holosystolic murmur was heard at the apex. Cardiac enlargement with clear lung fields was detected by the chest x-ray. Electrocardiogram showed sinus rhythm with low voltage criteria in the precordial leads. Echocardiographic examination revealed moderate pericardial effusion, diffuse left ventricular hypokinesis, a mass in the apical portion of the left ventricular cavity, and regurgitant flow from both atrioventricular valves (Fig. 17). Coronary angiogram was normal. On ventriculography both ventricles contracted poorly with regurgitant flow from both atrioventricular valves and there was a filling defect in the apical portion of the left ventricular cavity (Fig. 2).

Since the patient's clinical condition deteriorated despite medical treatment, an explorative thoracotomy was done. Three hundred milliliters of pericardial effusion was evacuated and a biopsy specimen was taken from the right atrial appendage. The patient continued to deteriorate and died on the fortieth hospital day. The pathological report, which

arrived after her death, showed cardiac rhabdomyoma (Fig. 3).

DISCUSSION

Cardiac rhabdomyomas occur almost exclusively in infancy and childhood. In a series of 36 patients only one was over 15 years (1). In another series of 17 cardiac rhabdomyomas reported by Burke (5) none was over 8 years of age. Before 1987 only 6 adult cases had been published, all diagnosed at autopsy (6). We could find only 3 adult cases reported after 1987 (6,7,8). Our patient, too, was a 42-year-old adult, who had lived without any symptoms until the last 18 months.

The number and location of the rhabdomyomas are the most important factors establishing the clinicopathological correlation. The tumor may be either solitary or multiple, and its location may be intramural, intracavitary or both. In a large series, the tumor was multiple in 92 % and solitary in only 8% of the patients (1-2). Ventricles were involved in all cases, but only 30% had atrial involvement (1,2). In 50% of the cases the tumor was intracavitary (1).

Symptoms resulting from cardiac rhabdomyomas are largely determined by the size of the tumor and its location within the heart. Some cases may be asymptomatic and diagnosed incidentally by echocardiography of found at necropsy. Others may lead to pulmonary or systemic congestion or cyanosis. Intramural tumors may cause loss of functional myocardium resulting in low cardiac output and congestive heart failure (3). Rhythm and conduction disturbances such as ventricular preexcitation, supraventricular tachycardia, and atrioventricular block have also been reported (4).

Our case was diagnosed as a cardiac rhabdomyoma with atrial involvement on the basis of the pathological examination of the specimen taken from the right atrial appendage wall. Atria have been reported to be involved in 30% of cardiac multiple rhabdomyomas (1). Considering 100% involvement of the ventricles

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Correspondance address: Doç. Dr. M. Serdar Küçüköğlü İ.Ü. Kardiyoloji Enstitüsü, Haseki-İstanbul



Figure 1: Two-dimensional echocardiogram in four chamber view demonstrating the mass in the opical portin of the left ventricular cavity (arrows): LA: Left atrium, RA: Right atrium LV: Left ventricle RV: Right ventricle PE: Pericardial effusion.

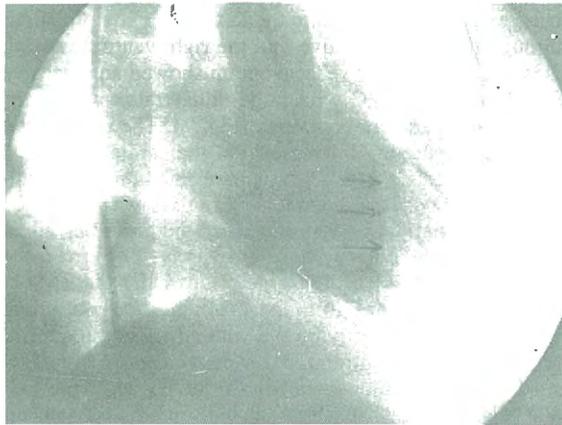


Figure 2: Left ventriculography showing a filling defect in the anterolateral, apical, and inferoapical walls (arrows).

and absence of solitary atrial rhabdomyomas in former series, we assume our patient also had multiple rhabdomyomas involving both the atrium and the ventricle, although the left ventricular mass could not be examined pathologically. Low cardiac output and congestive heart failure may contribute as evidence to ventricular involvement.

Besides congestive heart failure our patient had pericardial effusion which is rare in adult patients with cardiac rhabdomyomas. According to our knowledge, there is only one report of pericardial may either be due to pericardial involvement or congestive heart failure.

In conclusion, although extremely rare in adults, due to their potential mortality, cardiac rhabdomyomas should be considered in patients presentig with con-

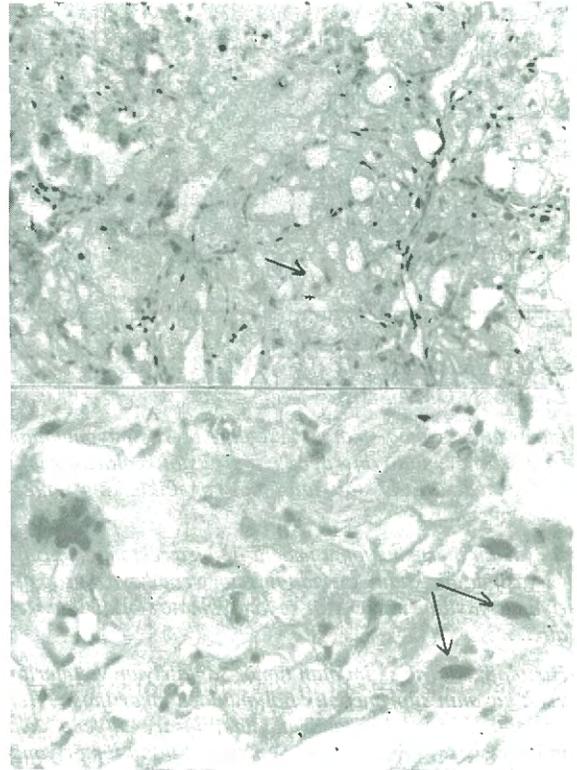


Figure 3: Cross-sections of biopsy material taken from right atrial appendage: Pleomorphic, round or polygonal gaint tumor cells are viewed. The cells contain acidophilic thin granulated or translucent vacuolated cytoplasm, and peripherally situated nuclei. (a) x200, (b) x500.

gestive heart failure and pericardial effusion with an intracardiac mass.

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