Fetal Rhabdomyoma: Prenatal Diagnosis And Postnatal Outcome

Fetal Rabdomiyoma: Prenatal Tanı Ve Postnatal Prognoz

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Abstract

Fetal cardiac rhabdomyoma is a very rare condition. We report a case with cardiac mass discovered in utero by prenatal ultrasonography at 38 weeks of gestational age. A giant echogenic mass at the two third of the interventricular septum was observed. The patient was followed up for three years with echocardiography, physical examination and cranial tomography whatever of tuberosclerosis.
Key Words: Cardiac tumor, Prenatal Echocardiography, Rhabdomyoma.

Özet

Fetal kardiyak rabdomiyoma nadir bir hastalıktır. Prenatal ultrasonografi ile 38. gestasyonel haftasında kalpte kitle saptanan bir vakayı sunmaktayız. İnterventriküler septumun 2/3'ünü kaplayan, rabdomiyoma tanısı konulan dev bir ekojenik kitle vardı. Hasta, tüberoskleroz açısından ekokardiyografi, fizik muayene ve kraniyal tomografi ile üç yıldır izlenmektedir.
Anahtar Kelimeler: Kardiyak tümor, Prenatal Echocardiography, Rabdomiyoma.

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INTRODUCTION

Cardiac tumors are rare at all ages and are even less common in children (1,2). The most common primary tumor of the heart in children is the rhabdomyoma, followed by teratomas and fibromas (1,3). Rhabdomyomas may be associated with tuberous sclerosis complex manifested (3,4).

CASE REPORT

A fetal echocardiographic examination was performed on a 36-year-old woman at 38 weeks of gestational age because of intrauterine cardiac mass at the first obstetric ultrasonography. Sonographic evaluation of the fetus at the time of referral showed that there was no cardiomegaly at the cross sectional examinations of the fetal thorax.

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There was a large homogeneous solid, hyperechoic mass with measuring 22.8x20 mm (4.9 cm^3) in the heart (Fig. 1). The mass was primary located at two third of the inferior interventricular septum and extended in rightward, leftward and superior direction. There was no evidence of a heart failure, arrhythmias, inflow or outflow obstruction.

Cesarean section was performed yielding a 3300 g male infant with an Apgar score of nine at both first and fifth minutes. His heart sounds were normal without any murmurs. An examination of the infant's skin revealed no abnormalities. There was no family history for evidence of tuberous sclerosis. Postnatal echocardiography revealed an echodens mass involving two third of the interventricular septum at the left ventricle.

The infant's cardiovascular and respiratory status remained stable, he was subsequently discharged home. At the end of first month, echocardiogram revealed that the mass had enlarged to 27x26 mm (6.8 cm^3) and mitral E/A ratio was two with impaired diastolic functions.

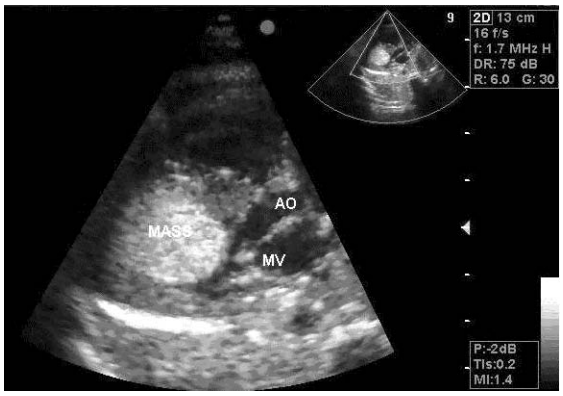

Figure 1. Fetal echocardiography of a 38-week fetus revealed a round echogenic mass in the lower portion of the left ventricle protruding to the mitral valve. (AO: aorta, MV: mitral valve).

The size of the tumor was decreased to 20.5x15.9 mm and diastolic functions were normalized at the end of third month without any therapy (Fig. 2). Physical examination, electrocardiogram, chest radiography, renal ultrasonography and cranial tomography results were normal at the end of three years.

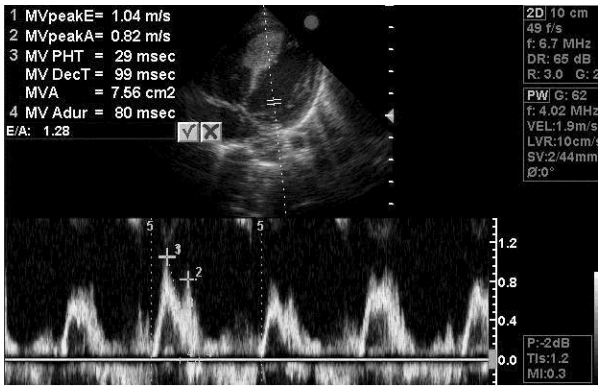

Figure 2. The size of the tumor decreased with normal diastolic functions at the end of third month.

DISCUSSION

Rhabdomyomas are the most common cardiac tumors in infants and children. They may be detected in utero (4,5). The clinical picture is variable and the natural history remains uncertain. Cardiac rhabdomyomas are benign tumors composed of altered myocytes with large vacuoles and considerable quantities of glycogen (1,2). They are usually multiple, spherical in shape and range from one mm to ten cm in diameter, grow slowly in utero but tend to regress spontaneously after birth. Their most frequent localization is the ventricles, especially interventricular septum.

Their association with tuberous sclerosis (1-5). Is well known. But our patient's neurological examination and related radiological examination were normal.

The differential diagnosis is limited to the other cardiac tumors. However, rhabdomyomas are more echogenic and firmly attached to myocardial walls and their size regresses after birth (1,2,4). Treatment for these tumors is indicated only if they result in critical blood flow obstruction and arrhythmias. Echocardiography provides guidance for therapeutic maneuvers, but also is beneficial for follow-up observation of the patients and assessment of the therapy. There was no need for surgical resection because our patient had not symptoms related to obstruction or compression of cardiac cavities and arrhythmias.

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