

Prenatal sonographic diagnosis of fetal cardiac rhabdomyoma: a case report*

Diagnóstico pré-natal de rabdomioma cardíaco fetal pela ultrassonografia: relato de caso

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Abstract Cardiac tumors constitute a rare condition, affecting 0.17–28/10,000 of the general population. Rhabdomyomas represent the most frequently found type of tumor in the prenatal period. The present report describes the case of a 31-week fetus presenting with a large intracardiac tumor with severe clinical repercussions. *Keywords:* Prenatal diagnosis; Ultrasonography; Fetal masses; Fetal tumors.

Resumo Os tumores cardíacos constituem condição rara, com incidência entre 0,17 e 28/10.000 na população geral. Os rabdomiomas são os tumores mais frequentes no período pré-natal. Este trabalho tem como objetivo relatar o caso de um feto com 31 semanas de gestação que apresentava tumoração intracardíaca de grandes dimensões, com graves repercussões clínicas.

Unitermos: Diagnóstico pré-natal; Ultrassonografia; Massas fetais; Tumores fetais.

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INTRODUCTION

Diagnostic imaging studies are frequently performed during the gestation to evaluate fetal diseases⁽¹⁾.

Cardiac tumors constitute a rare condition, affecting 0.17–28/10,000 of the general population, according to necropsy studies⁽²⁾. Recently, an increase has been observed in diagnoses of these tumors during the prenatal period, as result of the contribution of fetal echocardiography^(3,4). Depending on the intracardiac localization of

the tumor, the disease may progress with severe arrhythmias, congestive heart failure, hydrops, and not rarely, with fetal death⁽⁵⁾.

In the present study, the authors report a case of a fetus in the third gestational trimester presenting a bulky intracardiac mass diagnosed by means of prenatal echocardiography.

CASE REPORT

A 23-year-old, secundigravida, nulliparous patient at her 31st gestational week was referred to the institution to be submit-

ted to sonographic morphological study for cardiac malformation with arrhythmia diagnosed by routine sonographic study. Echographic findings demonstrated the presence of an intracardiac bulky mass in the right atrium, with the interatrial septum bulging. The mass presented a hyperechogenic, homogeneous aspect, and measured about 25 × 21 mm in its largest diameters (Figure 1). The fetus presented with persistent, marked tachyrrhythmia (on average, 263 beats/minute), with clear signs of congestive heart failure at 2D echography and ultrasound Doppler, and hydrops (Figures 2, 3 and 4).

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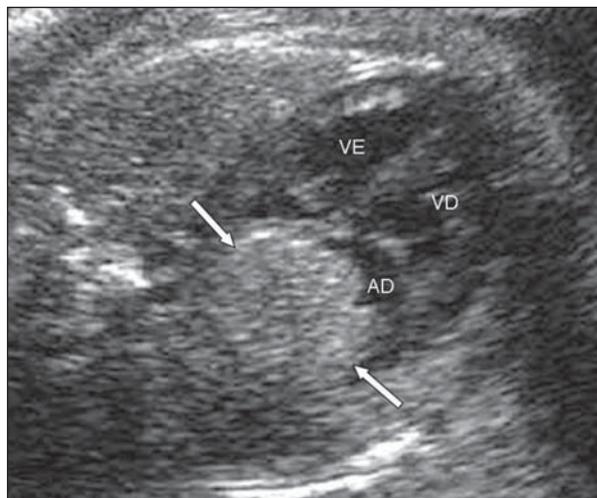


Figure 1. Fetal cardiac image - four-chamber view. A bulky, echogenic mass is observed in the right atrium, with interatrial septum bulging (arrows). AD, right atrium; VD, right ventricle; VE, left ventricle.

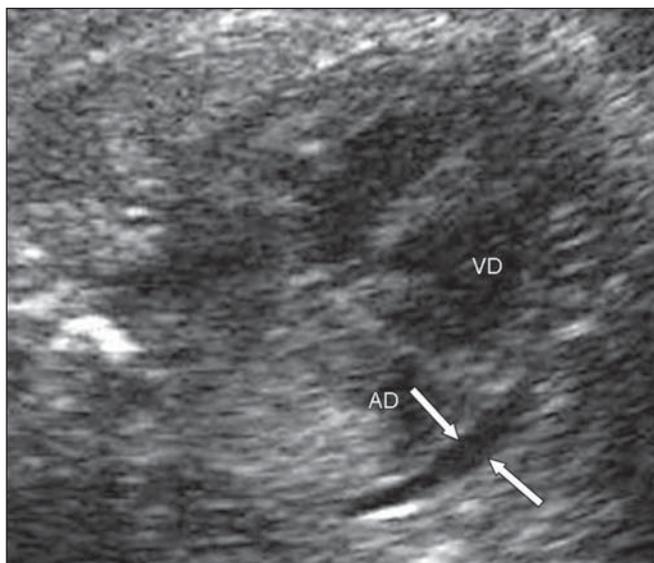


Figure 2



Figure 3

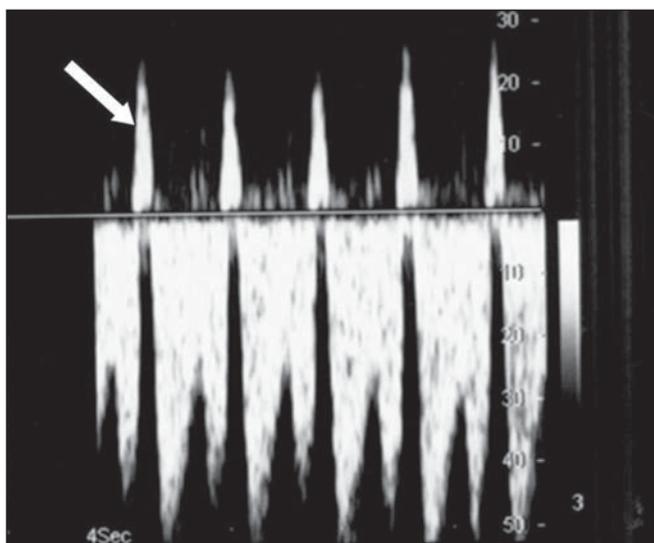


Figure 4

Figure 2. Heart failure. Image demonstrating pericardial effusion (arrows). AD, right atrium; VD, right ventricle.

Figure 3. Fetal hydrops. Fetal abdomen image demonstrating high-volume ascites (*). Additionally, placentomegaly is observed (**).

Figure 4. Heart failure. Venous duct demonstrating reverse A wave during atrial contraction (arrow)

In spite of the established therapy, the fetus progressed to death in ten days. Necropsy confirmed the prenatal diagnostic suspicion of cardiac rhabdomyoma.

DISCUSSION

Cardiac tumors, although constituting a rare condition, have presented an increased incidence of up to 0.2% in reference centers for pediatric cardiopathy⁽⁶⁾. Varied hemodynamic repercussions may be observed. According to their localization, size and number, these tumors may lead to blood flow obstruction and alteration of the ventricular ejection fraction, causing arrhythmia, congestive heart failure, pericar-

dial effusion, cardiomegaly, hydrops and fetal death^(5,7).

As regards histological types, cardiac rhabdomyomas or hamartomas represent the most frequently found histological lineage in the prenatal period^(6,7). The frequent association of this entity with tuberous sclerosis, a disease with autosomal dominant transmission, emphasizes the relevance of the diagnostic screening both in the fetus and its parents. In fact, tuberous sclerosis is observed in more than 50% of the fetuses with primary cardiac tumors⁽⁷⁾. In the present case, sonographic evaluation of the fetal central nervous system did not detect any suspicious intracranial involvement, and the parents are currently under-

going investigation for tuberous sclerosis. Fibromas, hemangiomas and teratomas are less frequently found in the fetal heart^(8,9). Fibromas usually present as a single lesion, frequently involving the left ventricular wall or the interventricular septum⁽⁶⁾. Teratomas originate in the pericardium and present as a single, heterogeneous, encapsulated and large sized mass, frequently reaching the base of the heart at right⁽⁸⁾. On the other hand, cardiac hemangiomas are rare in the intrauterine life and in the childhood⁽⁷⁾. Although cardiac myxomas are prevalent in the adult age group, only one study in the literature reports this histological type of tumor diagnosed in intrauterine life⁽¹⁰⁾.

Usually, at ultrasonography, cardiac rhabdomyomas are detected by the end of the second gestational trimester either as a single or multiple, well delimited, solid, homogeneous, hyperrefringent mass with varied dimensions^(2,6,7). Frequently, these lesions affect the left ventricle (60%) and, at a lower ratio, the right ventricle (26%)⁽⁶⁾. Cardiac dysrhythmias are observed in 16% to 47% of cases⁽⁵⁾. Therapy with digitalis and other antiarrhythmic drugs orally administered to mother or by fetal intravenous route (cordocentesis), and therapeutic preterm delivery should be considered in the presence of cardiac function involvement^(6,7). Intrauterine surgical resection of these tumors has been suggested for management of immature fetuses with hydrops, but a consensus is still to be reached in relation to the indication of this therapeutic alternative⁽¹¹⁾.

Most of times, however, cardiac rhabdomyomas are typically well tolerated, with risk for fetal death estimated in 4% to 6% of cases⁽⁷⁾. There are studies in the literature reporting spontaneous tumor regression, particularly in the third gestational trimester⁽⁷⁾. In a retrospective study, 19 cases of fetal cardiac tumors have been

evaluated, with partial or complete regression of these masses in eight patients, and with 10 patients presenting later development of tuberous sclerosis⁽⁷⁾. In fact, a perinatal benign course, with progressive regression and complete resolution of the tumor can be observed in more than 80% of cases in the childhood^(6,7).

CONCLUSION

The prenatal diagnosis of fetal cardiac tumors can be achieved by means of a detailed echographic study, with rare cases of false-positive diagnosis⁽¹²⁾. The early detection of these tumors allows a specialized prenatal follow-up and, if indicated, the institution of appropriate therapeutics, thus contributing for reducing the perinatal morbimortality.

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