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Prenatally diagnosed cardiac rhabdomyomas: characteristics and outcome

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Objective

The aim of this study is to determine the sonographic characteristics of cardiac rhabdomyomas in prenatally diagnosed cases and to characterize their evolution and postnatal management.

Methods

This is a retrospective descriptive study. It includes cases diagnosed prenatally in our University hospital between 2003 and 2018. Exclusion criteria were postnatal diagnosis, twins and cases with intra-uterine fetal demise. The information is collected from fetal and postnatal echocardiography as well as birth records and newborns' records. In viable fetuses, postnatal echocardiography and patient outcome data are also obtained. This study was approved by our ethics committee.

Results

Eight cases of rhabdomyoma diagnosed antenatally met the inclusion criteria. The mean gestational age at diagnosis of rhabdomyoma is 31w. The earliest diagnosis is at 26w while the latest is 35w + 5d. Five of eight patients were diagnosed after 32 weeks (62.5%) The mean age of the mother is 29.5 years. Gender of the babies is male in 6 / 8 (75%) and female in 2/8 (25%). Only one patient (fetus) has a familial Tuberous sclerosis (12.5%). Four fetuses have multiple rhabdomyomas (50%) and the remaining 4 have single rhabdomyomas (50%). A total of 17 tumors were found in the 8 patients. Seven are located in the interventricular septum (41%), 5 in the left ventricle (29.5%), 3 in the right ventricle (17.5%), one in the atrioventricular septum (5.8%) and the last at the level of the right atria (5.8%). At diagnosis the average size is 19.5 +/- 8.8 mm. The smallest diameter is 4 mm and the largest is 48 mm. During follow-up, in all the 5 cases that had a control ultrasound there was a size increase with a mean of 1.68 mm +/- 0.55. In two patients of eight (25%) there was a valvular insufficiency: one patient has mitral insufficiency (MI) grade III, and another has tricuspid insufficiency (IT) grade I. The patient with mitral insufficiency had a large 48 mm rhabdomyoma with a mass effect on the left atrium, left ventricle, aorta, coronary and pulmonary vein (12.5%), pericardial effusion (12.5%), dilation of the right atrium (OD) and right ventricle (RV) (12.5%), heart failure, ventricular hypokinesia, counterflow aortic irrigation. The baby later developed fetal anasarque with polyhydramnios. No patient had an associated cardiac malformation. All patients delivered at term between 36 and 40 weeks of age (87.5%) including five cesareans (62.5%), and three vaginal deliveries (37.5%). One patient had a preterm birth at 33 weeks due to polyhydramnios (12.5%). The Apgar at 1 minute is 9 in three patients, 8 in one patient and 7 in two patients; one baby died (he had a mass effect and a hypokinesia of the 2 ventricles) and another required reanimation with intubation. Three patients required resuscitation at birth (37.5%). The first required an urgent surgery but unfortunately died during the operation (12.5%), the second died despite resuscitation (12.5%) (he had a large heart mass with heart failure, abdominal ascites and cardiac effusion). The third improved after a three-day resuscitation (12.5%). So in total six babies survived (75%); three of them were later diagnosed with Tuberous sclerosis (50%), and two were lost to follow-up. Two of the four patients with a single tumor antenatally were found to have several tumors postnatally.

Conclusion

Cardiac rhabdomyomas are often asymptomatic, benign tumors that have multiple presentation. Neonatal death occurred however in 12% of the cases. Association with tuberous sclerosis syndrome could impact the long term neurological status.