An unusual heart

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In a female foetus of 31 weeks we diagnosed a mass of 15 x 17 mm, occupying most of the left ventricular cavity, which did not cause arrhythmia, cardiac failure, or obstruction.

After birth, we confirmed the presence of an intraventricular mass and performed 3D echocardiography to highlight the spatial relationship with cardiac structures.

The mass had an unusual heart shape and was visibly smaller (13 x 12 mm) compared with the prenatal aspect (Figure 1). There was no obstruction to left ventricular inflow or outflow and basal ECG was normal. The screening for tuberous sclerosis was negative. The baby remained asymptomatic during the hospitalization, but was readmitted 15 days later for the occurrence of runs of ventricular tachycardia, discovered by Holter monitoring, which we treated with beta-blockers.

During the foetal and neonatal period, rhabdomyomas represent the majority of cardiac tumours and are closely associated with tuberous sclerosis. Cardiac rhabdomyomas may be found in asymptomatic patients and may be incidentally discovered during echocardiography, or may cause cardiac dysfunction requiring medical and/or surgical intervention. On rare occasions, life-threatening conditions occur. These tumours generally regress after birth, and cardiac-related problems are rare after the perinatal period. Regression in utero is rare: in a meta-analysis, Chao AS in utero was 266 cases of cardiac rhabdomyomas diagnosed with antenatal ultrasonography and only one regressed.

This contribution confirms that the reduction of the mass size can start during foetal life and shows that the first arrhythmic event may occur several days after birth.

Panel A. Foetal echocardiography shows that the mass completely fills the left ventricular cavity.

Panel B. B-Mode postnatal echocardiography shows that the mass is adherent to the interventricular septum, has a heart shape form, and its size has diminished.

Panel C. 3D transthoracic echocardiography highlights the heart shape form of the mass.

Panel D. Holter monitoring shows a run of non-sustained ventricular tachycardia.

M, mass; RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; V, ventricular tachycardia.

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