Idiopathyc Dilation of the Right Atrial Appendage: Prenatal Diagnosis and Postnatal Evolution

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Case Report

We present the case of a pregnant woman without incidences in the previous controls, referred to our unit in the 21st week of gestation due to diagnostic suspicion in her area center of tricuspid dysplasia / Ebstein’s disease with significant tricuspid insufficiency and dilation of the right atrium. In the fetal echocardiographic examination in our center we could see a tricuspid valve with normal insertion and without insufficiency, a slight dilation of the right atrium and a dilatation of the right atrial appendage, without other fetal or cardiac pathology that can be assessed (Figure 1). In evolutionary controls at week 28 and 32 of gestation, no significant changes were observed, with an observed atrial conduction and non-conduction extrasystole of non-elevated frequency, with no tachycardia episodes. At birth, he was admitted in Neonatology of our center for observation, study and monitoring. The echocardiography showed dilation of the right atrial appendage (Figure 2) described in the prenatal study without other cardiac defects, a complete cerebral and abdominal ultrasound echo study that is normal. In successive ECGs, lead-induced and non-driven atrial extrasystole was seen frequently (Figure 2), without repercussion, and after 72h, atrial tachycardia was observed frequently, so treatment with Propranolol orally (2.5 mg/kg/day) was started, control of tachycardia in the following 24 hours, so that Amiodarone orally (5 mg/kg /day) was added to the treatment, which results in control of the arrhythmia.

Discussion

Dilation of the atrial appendage is a very rare entity, characterized by dilatation of the appendage in the absence of another cardiac anomaly. They are classified into two groups: congenital and acquired (mainly secondary to trauma). Those that affect the left atrial appendage were the first ones described in 1938 by Semans and Taussing [1]. They are usually manifested mainly in the third and fourth decade of life as tachyarrhythmias of difficult pharmacological control and thromboembolic phenomena. Those that affect the right appendage are even rarer, with very few cases published, most of them in adults. Its diagnosis is fundamental due to the possible complications discussed (pulmonary and systemic thromboembolism and arrhythmias). There are very few published cases, just a few pediatrics, and very rarely with a previous prenatal diagnosis. The age of presentation varies from the fetal period to the adult. The clinical presentation varies within the pediatric population, with most of the patients being asymptomatic with a casual diagnosis [2]. There is also clinical variability in the affected fetuses, from absence of symptoms to fetal hydrops due to severe tricuspid regurgitation. It is essential due to the possible complications that can present: tachyarrhythmias of difficult control [3], thromboembolic phenomena, respiratory distress, sudden death or palpitations. The natural evolution is still uncertain, which makes a consensus in the management of this condition difficult, so it must be individualized according to the symptoms and echocardiographic evolution. Some authors propose an expectant attitude in asymptomatic patients, while others suggest anti-platelet treatment, oral anticoagulant or surgical excision, depending on the evolution to avoid complications. For the postnatal diagnosis, techniques such as transthoracic and transesophageal echocardiography, computed tomography and magnetic resonance imaging can be used, although transthoracic echocardiography image is usually sufficient for the definitive diagnosis in pediatrics [4].

Figure 1. Prenatal study at 21 SG cut of 4 cameras in which there is significant dilatation of the right atrial appendage (RAA) and slight right atrial (RA), being the tricusid valve (TV) normal.

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Figure 2. Postnatal echocardiographic study showing persistence of right atrial appendage dilatation (RAA) with normal tricuspid valve. Serial extrasystoles and nonsustained atrial tachycardia are seen in serial ECGs.

References


