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PAEDIATRIC CARDIOLOGY - CASE PRESENTATIONS

Sudden cardiac arrest as first symptom of a benign cardiac tumor growth

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Abstract Primary cardiac tumors are rare, especially in the pediatric age. Most of them are benign in the sense they are not invasive. However, benign tumors maintain the potential for serious illness related to significant hemodynamic compromise or life-threatening dysrhythmias. We present the case of an infant with an initial diagnosis of cardiac rhabdomyoma who suffered ventricular arrhythmia and cardiac arrest. He suffered irreversible severe neurologic sequelae, due to his prolonged cardiopulmonary arrest and was finally diagnosed of cardiac fibroma. Good arrhythmia control was obtained after an extensive partial surgical resection of the tumor. This case highlights the importance of arrhythmia burden in this condition. A correct diagnosis based essentially in different imaging modalities and closer clinical and rhythm follow up could have avoided this ominous event.

PALABRAS CLAVE
Tumores;
Pediatría;
Parada cardíaca

Muerte súbita como primer síntoma de crecimiento de tumor cardíaco benigno

Resumen Los tumores cardíacos primarios son raros, especialmente en la edad pediátrica. La mayoría de ellos son benignos, en el sentido de que no son invasivos. Sin embargo, los tumores benignos tienen el potencial para producir enfermedades graves que pueden causar compromiso hemodinámico significativo o arritmias potencialmente letales. Se presenta el caso de un niño con un diagnóstico inicial de rabdomioma cardíaco, quien sufrió una arritmia ventricular y una parada cardíaca. Tuvo secuelas neurológicas severas irreversibles debido al tiempo prolongado en parada cardiorrespiratoria y se le diagnosticó finalmente fibroma cardíaco. Se obtuvo un

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buen control de las arritmias después de una extensa resección parcial del tumor. Este caso pretende subrayar la importancia del riesgo de aparición de arritmias en esta situación. El diagnóstico correcto basado fundamentalmente en el uso de distintas modalidades de imagen y el seguimiento clínico y arritmológico, podrían haber evitado este desenlace fatal.

Primary cardiac tumors are rare, especially in the pediatric age. The incidence in this age group is approximately 0.17% in the echocardiographic database review at Boston Children's Hospital,[1] which is similar to the incidence of 0.2% reported at the Hospital for Sick Children, Toronto.[2]

Most primary pediatric cardiac tumors are benign in the sense they are not invasive. Fewer than 10% of these primary tumors are malignant.[3] However, benign tumors maintain the potential for serious illness related to significant hemodynamic compromise or life-threatening dysrhythmias.

Is presented a case of a patient diagnosed of a benign primary cardiac tumor who developed a serious complication and poor outcome in follow up.

Case

A two year-old infant with prenatal diagnosis of cardiac mass, who collapsed at home while playing. He was being followed since birth in a cardiology outpatient clinic with the diagnosis of benign cardiac rhabdomyoma. He suffered a cardiopulmonary arrest and was taken to the emergency department where advanced resuscitation maneuvers were practiced for 40 minutes. First documented cardiac rhythm was ventricular fibrillation. After initial stabilization and treatment, he was transferred to our centre for multidisciplinary treatment.

Chest x-ray demonstrated an increased cardiothoracic index (0.70) with pulmonary congestion. 12-lead electrocardiogram showed unremarkable sinus rhythm with an average heart rate of 130 bpm, normal PR and QRS intervals, and QRS axis in 0°. There were symmetrical and deep T-wave inversions in the lateral precordial leads. Transthoracic echocardiography demonstrated a homogeneous intramural left ventricular mass in the free wall and apex, which involved the anterior papillary muscle, with no valvular incompetence nor mechanical left ventricular outflow or inflow obstruction. The image suggested a fibroma as the first diagnostic possibility (fig. 1). Cardiac magnetic resonance images revealed a $4.5 \times 4.8 \times 4.1$ cm, firm, well-demarcated intramural mass related to the anterior papillary muscle and the subendocardial free wall region of the middle and apical segments of the left ventricle. Late contrast-enhanced cardiac magnetic resonance images were acquired after gadolinium injection, showing a homogeneous and intense bright mass, suggesting the nature of fibroma (fig. 2). A slight global left ventricular systolic dysfunction was observed, with an ejection fraction of 55%.

The patient had a severe irreversible neurological dysfunction due to a severe hypoxic ischemic encephalopathy secondary to the prolonged cardiac arrest. Given this situation, he was unable to swallow and a percutaneous gastrostomy was placed.

During admission, he presented episodes of non-sustained ventricular tachycardia as well as multiple episodes of different monomorphic sustained ventricular tachycardia. Ventricular tachycardia morphology suggested its origin from left ventricular apex, coinciding with cardiac tumor's location. Pharmacologic treatment was initiated with amiodarone and propranolol with poor control of the arrhythmia. The decision was taken to perform a complete surgical resection of the tumor (fig. 3). Finally, only an extensive partial tumor resection was undertaken due to technical

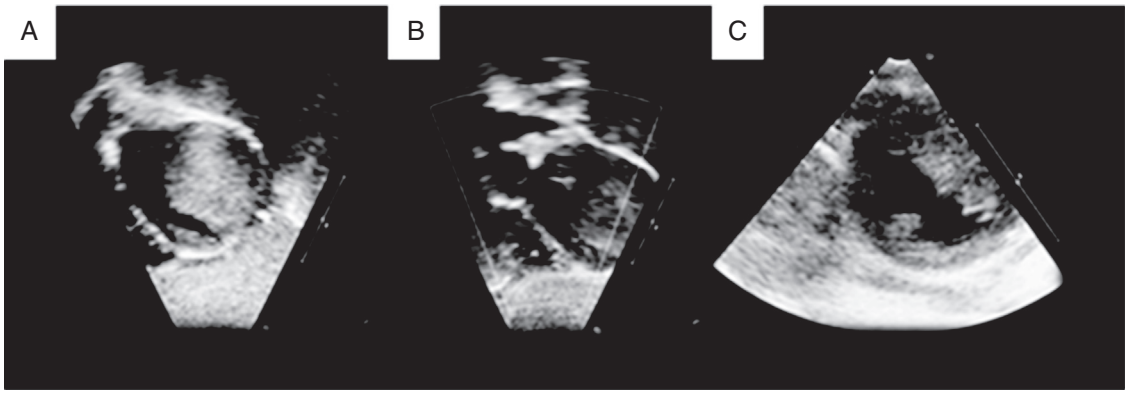

Figure 1 Two-dimensional echocardiogram in a 2-year-old child with a large left ventricular fibroma. A. Subcostal sagittal sweeps showing homogenous intramural mass extending from the left ventricular free wall into the cavity. B. Subcostal coronal sweeps revealing no left ventricular outflow obstruction. C. Parasternal short-axis shows tumor involving the anterior papillary muscle.

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