CASE REPORT

Cardiac fibroma as a cause of ventricular arrhythmias in childhood

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Cardiac tumors are a rare pathology although fibromas are reported to be the second most frequent in the literature. Not much is described about their presentation and management. We present a case of a preschool age child with sudden onset of difficult-to-control ventricular tachycardia events, finding a cardiac fibroma as the cause of the event. Surgical excision was required as a treatment.

Key words: **Arrhythmia; Cardiac tumor; Fibroma; Surgery.**

Los tumors cardiacos constituyen una patología rara, aunque los fibromas han sido reportados como el segundo tumor más frecuente en la literatura. No se ha descrito mucha información acerca de su presentación y manejo. Presentamos aquí un caso de un niño en edad preescolar con aparición súbita de episodios de taquicardia vent4icular de dificil control. La excisión quirúrgica fue necesaria como tratamiento.

***Palabras clave*: Arritmia; Tumor cardiaco; Fibroma; Cirugía.**

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Cardiac tumors are a rare pathology, with an incidence of 0.001 to 0.03% [1]. Cardiac fibroma is a tumor of connective tissue derived from fibroblasts, infrequent. No specific age group has been described for this kind of tumor; however, it can be observed predominantly in the neonatal and childhood stages. It is the second most common primary cardiac tumor in pediatrics [2]. Due to its infiltrative nature, conduction tissue invasiveness can cause arrhythmias or sudden death [3]. We present herein the case of a cardiac fibroma located at the posterior wall of the left ventricle, which was diagnosed at the age of 5 years. Clinical presentation was under the way of ventricular arrhythmias. The case required surgical excision as treatment of this pathological condition.

CLINICAL CASE

A 5-year-old child was referred to our institution for sudden onset of monomorphic ventricular tachycardia (**Fig.1A**). Transthoracic echocardiogram showed a tumor in the posterior wall of the left ventricle (**Fig. 1B**). Magnetic resonance imaging (MRI) reported a tumor with well-defined and regular margins in the anterolateral wall located on the basal and middle third of the left ventricle. Esteemed measures were 13mm by 22mm, probably related to diagnosis as fibroma (**Fig. 1C**).

The surgical approach was through median sternotomy, with bicaval and aortic cannulation protocol. Aortic cross-clamping and retrograde cardioplegia were used as well as moderate hypothermia. Reference sutures using monofilament suture in four quadrants were placed along blunt dissection of the tumor. The complete tumor was obtained, sizing 30mm by 20mm by 10mm (**Fig. 2**). Total aortic cross-clamping was of 25 minutes, and cardiopulmonary bypass of 45 minutes. Cardiopulmonary bypass weaning was uneventful, maintaining sinus rhythm.

Anatomic-pathological report from intraoperative scraping of the tumor showed spindle-shaped cells with a mesenchymal appearance, regular nuclei with fine and homogeneous chromatin. Frozen sections of the mass revealed spindle cell neoplasia in fibrous stroma. Findings were compatible with cardiac fibroma (**Fig. 3A**). Macroscopically, an ovoid, delimited, encapsulated tumor mass of 30mm by 20mm by 10mm was identified. The external surface was whitish, homogeneous, with fibrous appearance and firm consistency (**Fig. 3B**).

Postoperative course was free of complications. In-hospital discharge was at the third postoperative day. No cardiac rhythm alterations were observed at the electrocardiogram (**Fig. 3C**).

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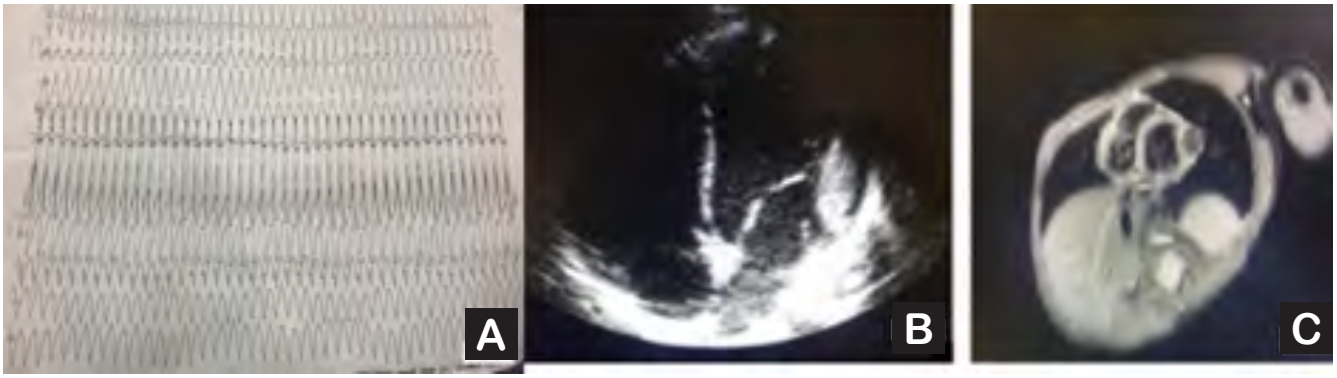

Figure 1. A: Electrocardiogram with monoform ventricular tachycardia. B: Echocardiogram with tumor in the posterior wall. C: Magnetic resonance imaging of the tumor with well-defined borders in the anterolateral wall of the left ventricle.

COMMENT

The incidence of primary cardiac tumors varies from 0.001-0.003% [4]. Cardiac fibroma is rare, appearing at any age; however, 75% appear in the first two years of life [2]. So, the age of our patient was unusual. It is about a single intramural tumor. Left ventricle and interventricular septum are the most affected zones [3]. Symptoms are related to size and location; nonetheless, most of them are asymptomatic. Compared to rhabdomyoma, in this variety there is no spontaneous regression, and they tend to cause complications with high mortality if surgical resection is not performed [5]. One of the main characteristics of this neoplasm is that arrhythmias are difficult to manage or can even debut with sudden death. In our case, no symptoms were observed, but just one month prior to removal, with the presence of palpitations secondary to ventricular tachycardia.

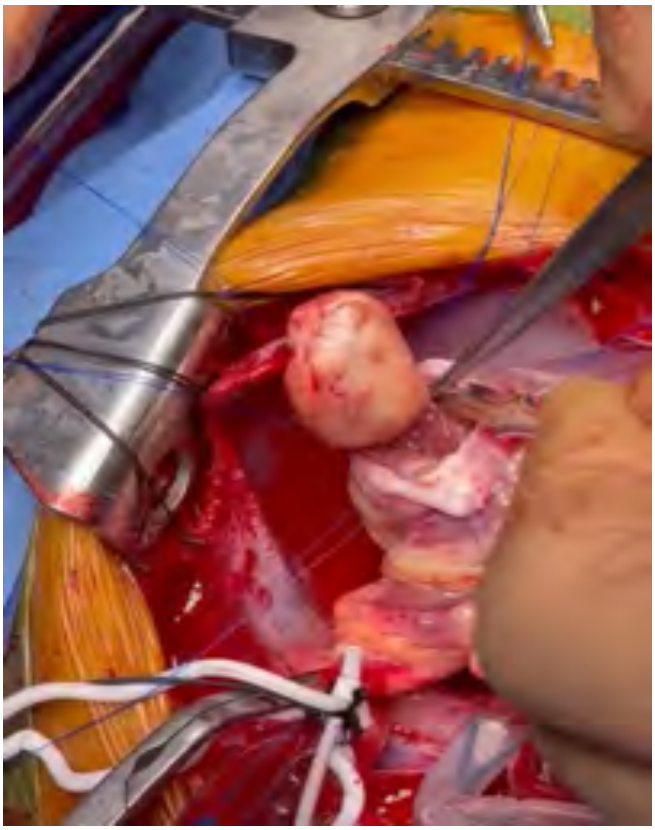

Figure 2. In situ image of the tumor. A) initial dissection. B) excision of the posterior wall.

Macroscopically, the fibromas are round, well circumscribed, located within the ventricular myocardium, protruding towards the cavity, being simple and measuring from 2cm to 10cm [4], with some central calcifications [5].

With the advancement of non-invasive imaging methods for the detection of cardiac tumors, the approach to their diagnosis and treatment has changed [5]. The most specific non-invasive diagnostic technique is MRI, which provides valuable information about the size, location, and extension of the tumor [5]. In our case, MRI provided important information that was essential for surgical planning.

There are few long-term retrospective studies addressing the entire spectrum of cardiac tumors in children. In 2012, a multicenter study of the European Association of Cardiac Surgeons was carried out, which reported the early and late results of surgery, confirming that the surgical resection is indicated with symptoms and hemodynamic impairment [6]. In general, surgical indications are related to the hemodynamic or mechanical effect, arrhythmias or ventricular dysfunction, rather than to the pathological condition of benignity or tumor size [5]. Tumor resection has low mortality [4]. In these cases, complete resection is preferred over partial or limited one [5]. In our case, it was decided to perform tumor resection due to the main condition as cardiac arrhythmia.

In conclusion, the diagnosis of cardiac tumors is challenging. The clinical scenario depends primarily on the location and tumor rather than on histopathological basis.

Cardiac tumors are a rare entity, most of them asymptomatic; however, rhythm disturbances are not uncommon and can be dangerous. Cardiac fibroma, although the literature refers to it as the second most frequent pediatric tumor and the more resectable, in our environment there are few articles published on the experience regarding to

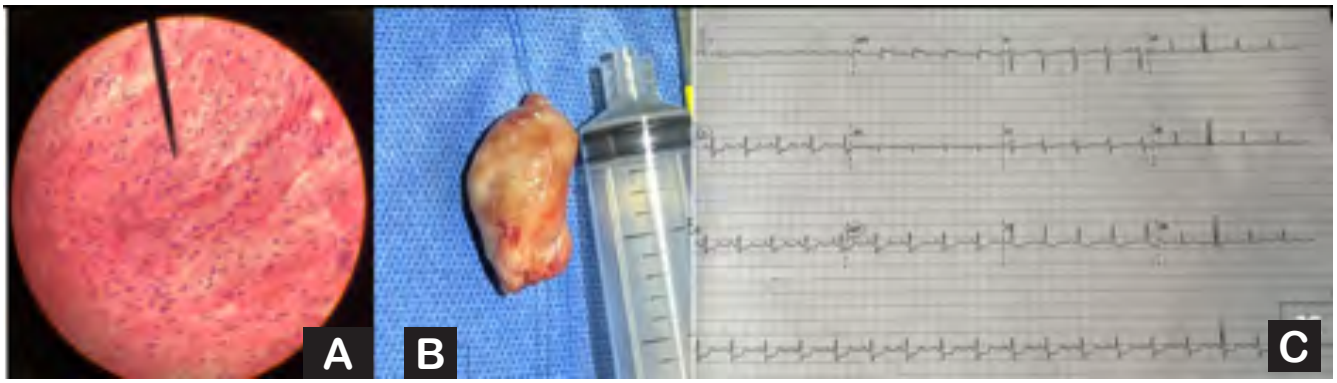

Figure 3. A: Neoplasm frozen sections compatible with cardiac fibroma. B: In situ image of the tumor. C: Postoperative electrocardiogram.

this subject. This case described here was the first one at our institution.

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