

Long-Term Follow-Up of an Asymptomatic Man with a Cardiac Fibroma

Acompanhamento a Longo Prazo de Paciente do Sexo Masculino com Fibroma Cardíaco Assintomático

Marina Costa Jonas,¹ Maria Fernanda Braggion-Santos,² Marcel Koenigkam-Santos,³ Paulo Henrique Manso,¹ Luiza Frison Dias, André Schmidt,² Fernando Amaral⁴

Pediatric1 and Adult4 Congenital Heart Disease Unit, Division of Cardiology2 Imaging Center3 – University Hospital, Ribeirão Preto Medical School, USP, Brazil.

Abstract

Here we describe a case of a 19-year-old asymptomatic man with a left ventricular fibroma on follow-up for 15 years with no treatment required.

Introduction

Cardiac fibroma, first described in 1855, is a benign tumor mainly affecting the left ventricular (LV) free wall that occasionally extends to the interventricular septum. Affected children may be afflicted by significant arrhythmias, creating a sudden death risk requiring prompt surgical treatment.¹ An asymptomatic presentation as well as first manifestation in adults is unusual and follow-up has not been well reported. Here we present the case of an untreated asymptomatic adult who was followed up for 15 years.

Case report

An enlarged heart was incidentally detected on the chest radiograph of a 4-year-old child with respiratory complaints (Figure 1A). Cardiac examination findings were normal. A 12-lead electrocardiogram (EKG) disclosed LV hypertrophy and inverted T waves in the anterior leads (Figure 1B). A 4.1 × 3.7 cm round mass was documented by two-dimensional echocardiography in the apical-lateral LV wall with unobstructed flow (Figure 2A). Twenty-four-hour Holter monitoring revealed one 7-beat episode of non-sustained ventricular tachycardia. On cardiac magnetic resonance (CMR), a 4.7 × 4.2 cm well demarcated mass was found in the lateral-inferior LV wall with late gadolinium enhancement consistent with cardiac fibroma (Figures 2B, 2C). Despite the brief tachycardia episode, surgery was not recommended. For 14 years, the uneventful follow-up was marked by irregular hospital visits every 1–2 years due to personal reasons including long distance to travel, but telephone consultations

were frequently performed. Currently, as an asymptomatic 19-year-old with normal cardiac examination findings, he leads a normal life including moderate physical exercise and is taking no medication.

Recent chest radiography (Figure 1C), EKG (Figure 1D), and echocardiography (Figure 2D) findings were similar to those obtained 15 years prior. Holter monitoring findings were normal and arrhythmias were not triggered by a recent exercise test. Sequential CMR showed small growth of the lesion (dimensions: 5.0 × 4.3 cm), which remained stable for the last 9 years (Figures 2E, 2F). Steady-state free precession (SSFP) cine images in systolic (Figures 3A, 3C) and diastolic (Figures 3B, 3D) phases on CMR showed no significant repercussions in left ventricular function at 5 and 19 years of age. The case has been recently discussed and no consensus was reached regarding surgery. According to patient and family desire, regular 6-month follow-ups were offered after a detailed explanation about the risks and benefits of such an approach.

Discussion

Cardiac fibroma, an unencapsulated tumor composed of fibroblasts and connective tissue, is rarely found in adults and usually diagnosed by echocardiography. After the detection of such a mass on echocardiography, the differential diagnosis includes other benign cardiac tumors (especially lipoma and hemangioma).² Computed tomography and CMR may define lesion extension and tissue characteristics, thereby improving diagnostic accuracy and treatment planning as in other tumors.³ In this scenario, CMR has an important role due to its unique capability to provide an advanced myocardial tissue characterization mainly through late gadolinium enhancement sequences, which allows the detection of fibrotic tissue, the main component of cardiac fibroma.⁴ Its clinical presentation in children may include syncope, dyspnea, chest pain, episodes of ventricular tachycardia, and, more rarely, cardiac arrest. In this setting, agreement exists regarding complete surgical resection, which, whenever accomplished, is curative with excellent early and late results.⁵ However, opinions diverge in cases of the asymptomatic child in whom the risk of sudden death is used to justify a preventable surgical policy, although fatal arrhythmia appears to occur more frequently in the first few years of life.

In 29 recently reported cases, eight patients experienced cardiac arrest in the first three years of life, seven within the first year.⁶ In this remarkable retrospective investigation, an interdigitating and entrapped myocardium was uniformly

Keywords

Adult, Fibroma, Cardiac Neoplasms, Sudden Death.

Mailing Address: Fernando Amaral •

Adult Congenital Heart Disease Unit, University Hospital, Ribeirão Preto Medical School USP. Rua Ten. Catão Roxo 3900, Vila Monte Alegre, 14015-010 Ribeirão Preto SP, Brazil.

E-mail: ftamaral@cardiol.br

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Case Report

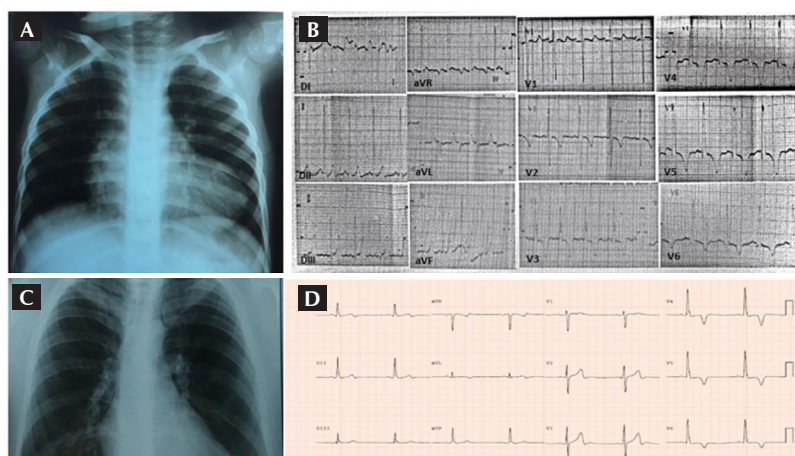


Figure 1 – Chest radiographs taken at 4 (A) and 19 (C) years of age. Electrocardiogram taken at 4 (B) and 19 (D) years of age.

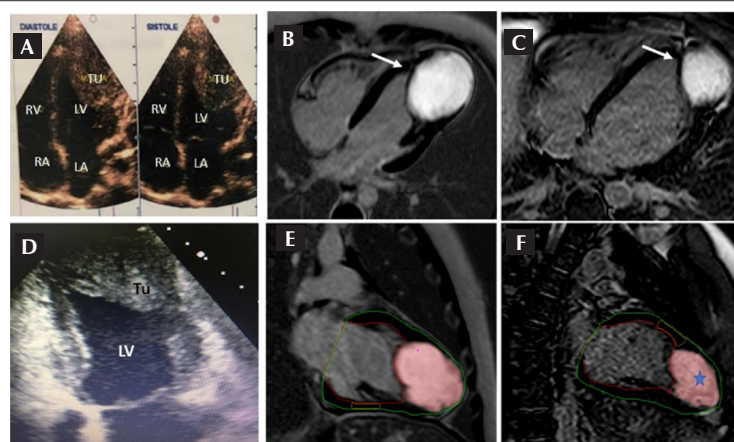


Figure 2 – Echocardiogram at 4 (A) and 19 (D) years of age (LV, left ventricle; Tu, tumor). Cardiac magnetic resonance imaging late gadolinium enhancement at 4 (B) and (E) and 19 (C) and (F) years of age. (B) and (C) Four-chamber view characteristic hyperintense signal (arrow); (E) and (F) Two-chamber view quantitative analysis of myocardial fibrosis using the full-width half-maximum method showing the hyper-enhanced tumor (star) without areas of fibrosis mixed with normal myocardium (gray zone).

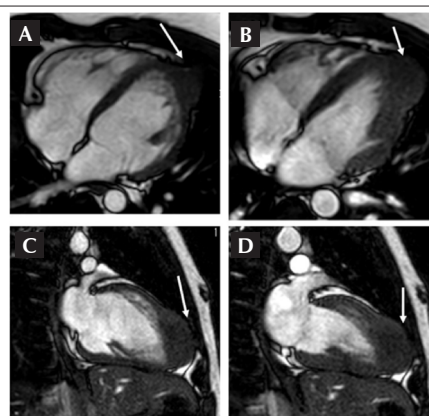


Figure 3 – Relationship of cardiac fibroma with left ventricular contractility. Steady-state free precession cine magnetic resonance images in systolic (A and C) and diastolic (B and D) phases. (A) and (B) Four-chamber view. (C) and (D) Two-chamber view. White arrows: cardiac fibroma – note the isotense signal of the tumor related to myocardium in these sequences.

present to various extents within the tumor in at least 80% of cases and presented three interesting features: they were more extensive at a younger age at the time of surgery, were of moderate or marked extension in the majority of patients with cardiac arrest, and tended to gradually disappear with age. This peculiar histologic aspect is similar to the one found in peri-infarct zones and is very likely the arrhythmia substrate since complete surgical resection eliminated the arrhythmia in these cases.

Management of the adult with a cardiac fibroma is debatable. Although surgery is not without risk and ventricular reconstruction might be difficult, 38 patients over 16 years of age were reported by 2011; surgery was performed in 34 of them, including four who were asymptomatic.⁷ The case presented here, as well as other non-operated patients without⁸ or with routine⁹ follow-up, should not be used to recommend a conservative approach for the asymptomatic adult. The consensually obtained recommendation against surgery when the patient was 4 years old was not uniform when he was 19 years old, including a detailed debate in an international panel. The possible risks and benefits of the clinical or surgical management that emerged from these discussions were presented to the patient and his family, who opted for clinical management considering the patient's well-being and strong religious belief. It is interesting to note that the recently described myocardial tongues/entrapment characteristics⁶, which tend to disappear with age, suggest that the asymptomatic adult probably has a low risk of

arrhythmia; thus, surgery can be avoided. The CMR tumor signal homogeneity demonstrated here, suggestive of a low mixture of normal and pathological tissue, is very likely responsible for the electrical stability noted in our patient. Could this gray zone¹⁰ absence be considered an expression of scarce interdigitating/entrapped myocardium and be used to decide its appropriate management? Which degree of this myocardium abnormality would identify a high-risk patient? There are currently no answers for these questions which could eventually be achieved using a specific registry of a good number of patients adequately followed up, however difficult. In the meantime, common sense and shared decision-making with the patient and family seems the best policy.

Author's statement: Permission was granted by the patient for the publication of this case report.

Authors' Contributions

Research conception and design: Jonas MC, Amaral FA; data collection: Jonas MC, Dias LF; data analysis and interpretation: Jonas MC, Braggion-Santos MF, Koenigkam-Santos M; manuscript writing: all authors; critical review of the manuscript: Manso PH, Schmidt A, Amaral F.

Conflict of interest

The authors have declared that they have no conflict of interest.

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