Rare Case of Hamartoma Mimicking Fibroma and Correlations with Literature

Elíana Migliorini Mustafa¹, Victor Rodrigues Ribeiro Ferreira¹, Sofia Braile Sabino¹, Maria Christiane Valéria Braga Braile-Sternieri¹, Rodrigo Serpa Sestito¹, João Carlos Leal¹, Bethina Canaroli Sbardellini¹, Giovanni Braile Sternieri¹, Lucía Angélica Buffulin de Faría¹, Idiberto José Zotarelli Filho¹,² and Domingo Marcolino Braile¹

¹Domingo Braile Institute of Sao Jose do Rio Preto (SP), Brazil
²State University of Sao Paulo, UNESP, Brazil

Corresponding author: Prof. Dr. Idiberto José Zotarelli Filho, Domingo Braile Institute of Sao Jose do Rio Preto (SP), Brazil, Tel: + 55 (17) 3203-4039, +55(17) 8186-6537; E-mail: m.zotarelli@gmail.com

Received date: December 19, 2016; Accepted date: January 29, 2017; Published date: January 31, 2017

Copyright: © 2017 Filho IJZ, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Background: The incidence of primary cardiac tumors ranges from 0.02 to 0.05% in autopsy studies. Hamartomas from mature cardiac myocytes do not present a predilection for age at detection and present a higher prevalence in the left ventricle with the advent of extracorporeal circulation and the advancement of image diagnostic medicine such as echocardiography, computed tomography and magnetic resonance imaging, diagnosis and surgical treatment became more feasible.

Objective: To present the case report of a very rare HA with diagnosis by clinical correlation, imaging equipment and microscopy, in order to differentiate it from fibroma.

Case: A 63-year-old female patient with a history of chest pain and dyspnea on exertion with progressive worsening six months ago. The echocardiogram presented apical akinesia with an intramyocardial fibro-calcified mass, presenting an intense local coronary vascularization, with a dilated epicardial coronary artery at the tip with 5.0 mm, where the intramyocardial vessels that irrigate the tumor of the apical region originate. A superficial myocardial bridge was visualized by MRI in the middle segment of aDA. The patient underwent endomyocardial biopsy with a suggestive result of Hamartoma of mature cardiac myocytes.

Conclusion: Despite the diverse range of cardiac neoplasms, heart tumors are very rare, even more so as the apical Hamartoma and confused with fibroma presented in the present work.

Keywords: Cardiac tumors; Image devices; Microscopic analysis; Hamartoma; Fibroma; Vascularization; Calcification

Introduction

The incidence of primary cardiac tumors ranges from 0.02 to 0.05% in autopsy studies. In secondary tumors an incidence of 1.0% is observed [1]. Benign tumors represent about 75.0% of the cases, with 50.0% of them being myxomas, the remainder being lipomas, papillary fibroelastomas and rhabdomyomas. About 25.0% of the cardiac tumors exhibit malignant characteristics or behave in such a way, 95.0% of them consisting of sarcomas and 5.0% of lymphomas [1].

In this context, in the 1950s, Hamartoma (HA) was considered a rare finding. The HA of mature heart muscle was first described in 1988 by Tanimura et al. [2]. Three additional cases were reported in 1998 by Burke et al. [3]. Lesions are characterized by disorganized myocytes that also hypertrophy and by variable interstitial fibrosis and adiposity. Myocyte derangement includes 3 patterns: (1) Random, (2) Fishbone and (3) Whorl or Pinwheel, with a higher prevalence in the left ventricle [3].

With the advent of extracorporal circulation and the advancement of diagnostic medicine such as echocardiography (ECHO), computed tomography (CT) and magnetic resonance imaging (MRI), diagnosis and surgical treatment became more feasible [4-6]. However, few are the literary findings on the incidence and prevalence of HA, that is, after a systematic review of the present study, only 74 scientific papers on HA was found of 245 of the total on cardiac tumors.

To identify them, there are microscopic HA characteristics of mature cardiac myocytes. However, they are also characteristic of hypertrophic cardiomyopathy [7]. However, Burke et al [3] have suggested that the whorl / pinwheel form of disorder, which is commonly observed in hypertrophic cardiomyopathy, is not characteristic of HA in mature cardiac myocytes. The differentiation between HA and hypertrophic cardiomyopathy may be simple in patients with non-hypertrophied hearts.

HAs from mature cardiac myocytes do not present a predilection for age at detection and present a higher prevalence in the left ventricle (VE) [8]. In addition, the development of nonspecific abnormalities in the ECG and other clinical manifestations may be related mainly to the size of the HA and its higher prevalence in the LV. Therefore, echocardiography, electrocardiography and magnetic resonance imaging have been useful for visualizing location, size and calcification [8]. In addition, vascularization is variable and angiography can be difficult to interpret. Thus, the only way to establish a more probable diagnosis of HA is by imaging and by microscopy [9].

Therefore, the only way to establish an HA diagnosis is by histological examination. However, clinical history and imaging findings play a key role in distinguishing other entities in differential
diagnosis. Thus, the objective of the present study was to present the case report of a very rare HA with diagnosis by clinical correlation, imaging equipment and microscopy, in order to differentiate it from fibroma.

Case Report

A 63-year-old female patient with a history of chest pain and dyspnea on exertion with progressive worsening six months ago. The echocardiography presented apical akinesia with an intramyocardial fibro-calcified mass, presenting an intense local coronary vascularization, with a dilated epicardial coronary artery at the tip with 5.0 mm, where the intramyocardial vessels that irrigate the tumor of the apical region originate. A superficial myocardial bridge was visualized by CTA in the middle segment of aDA (Figure 1).

In the CTA (Figures 1 and 2) and magnetic resonance imaging (MRI) (figure 3) study, myocardial thickness increased in the apical region and in the anteroposterior-medial LV segment around 2.2 cm, with an irregular appearance and extensive areas of calcification (figure 2) and microaneurysms. This mass (Figure 3) also shows a cleavage plane with the structures adjacent to the myocardium and heterogeneous signal. The myocardial perfusion had a partial defect in the calcified areas, presence of areas of fibrosis and apical necrosis and in the papillary muscle.

Coronary angiotomography showed important obstructive coronary artery disease due to atherosclerotic disease in the circumflex artery, the first diagonal and right coronary branch. The patient underwent endomyocardial biopsy with a suggestive result of Hamartoma of mature cardiac myocytes. Subsequently submitted to the withdrawal of the tumor mass in apical region associated to the myocardial revascularization, presenting good postoperative evolution (Figures 4 and 5, respectively).

Microscopic analyzes of the anatomopathological showed that Hamartoma presented dense collagen, adipose tissue, hypertrophic and with disarticulated cardiomyocytes. It is also suggested irregular junction of connexin 43. Thus, these events correlated with those of imaging and clinical, evidence strongly the diagnosis of HA.
Cardiac rhabdomyomas may also have a similar appearance to HAs of mature heart muscle [1]. However, they do not represent a calcified lesion and the myocytes are diffusely vacuolated and contain a lot of glycogen, whereas the myocytes in HA are hypertrophied and only exhibit focal areas of vacuolization [1-3].

In this context, several imaging devices such as echocardiography, electrocardiography and magnetic resonance imaging have been useful to visualize the location, size and calcification [11,12]. In addition, vascularization is variable, angiography can be difficult to interpret. Currently, the best ways to establish a more likely diagnosis of HA are through clinical analysis, imaging, and microscopy [11].

Strategically, the HA of mature cardiomyocytes must be differentiated from several other more common entities. Thus, hypertrophic cardiomyopathy (HCM) can mimic a cardiac tumor and share similar histological findings with an HA, especially when smaller surgical or biopsy specimens are available. However, mass HCM is distinguished from HA by lack of vascularization, more diffuse myocardial involvement and septal tract characteristic [5,6]. In addition, HCM presents varying degrees of contractility, whereas a mass of HA does not present normal contraction [2]. Yet, HAs can also be mistaken for fibroids. However, fibroids are poorly vascularized, late in the increase, and contain calcifications [2,5]. In addition, some cardiac hemangiomas may contain fat, fibrous tissue, calcifications; however, cardiomyocytes may appear more hypointense than HAs in MRI [2,5]. However, once the diagnosis is established, it becomes amenable to surgical resection [10].

Despite the low cost and availability of the echocardiogram, the visualization of the extracardiac extension is suboptimal because of the limited field of vision [10]. Therefore, MRI is the entity of choice for assessing heart mass, offering superior contrast resolution, hemodynamic information, tissue characterization, and small amounts of fat mixed with normal myocardium may also be best demonstrated [10,11]. However, it presents the inability to demonstrate calcifications as well as tissue characterization of small masses, due to their limited spatial resolution [10-12]. Thus, computed tomography complements MRI by definitive characterization of calcifications.

Based on the work of Fealey et al. [6], identified the left ventricle as the most common site of HA incidence (62.0%), followed by the septum (15.0%), right ventricular free wall (15.0%) and right atrium. Still, in adults, all HA were solitary and contained interstices of fat and also had small calcifications. Although some clinical manifestations are similar, the presentation of a cardiac HA is nonspecific, and varies from asymptomatic cases to presentations with ventricular, supraventricular and death tachycardia [6].

In general, cardiac tumors can produce common effects and symptoms, such as obstructions, since atrial tumors can impede blood flow through the atrioventricular valves and mimic valvular stenoses [4]. Already in ventricular cavities they can cause obstructions to the exit routes, attending with symptoms of the type thoracic pain, dyspnea or syncope [4,9,17]. Another problem is embolisations, small and multiple emboli can originate from the tumors themselves or by thrombi of their surroundings, mimicking vasculitis or endocarditis, and the larger ones may evolve with occlusive events such as stroke [1,14,17]. They can also cause arrhythmias due to infiltration of tumor tissue in the myocardium and in the conduction pathways, as well as the presence of tumor mass can cause alterations in the cardiac rhythm, such as atrioventricular blocks, ventricular tachycardias or even sudden death [1].
Exhaustive tumor sampling will be required to characterize histology and make an accurate diagnosis, and metastases are more common than primary neoplasms [5,13]. The morphological characteristics and the clinical location can identify the five types of these lesions that are Rhabdomyoma, Histiocytoid Heart Disease, Lipomatous Hamartoma in the Atrial Septal, Lipomatous Hamartoma in the Heart Valves and Hamartoma in Cardiomyocytes [5,7]. There is still little description in the literature of the histological characteristics of the lesions. Regarding the treatment, surgical excision is the best indicated, since the general condition of the patient does not bring clinical results [7].

Conclusion

Despite the diverse range of cardiac neoplasms, heart tumors are very rare, even more so as the apical Hamartoma and confused with fibroma presented in the present work. Benign tumors make up the majority of the cases, and both conservative and surgical treatments show favorable results.

Acknowledgements

The work was supported by Domingo Braile Institute - São José do Rio Preto - SP, Brazil.

Conflict of Interest

The authors declare no conflict of interest.

References