Journal of Clinical & Experimental Cardiology

Mustafa et al., J Clin Exp Cardiolog 2017, 8:1
DOI: 10.4172/2155-9880.1000497

Research Article Open Access

Rare Case of Hamartoma Mimicking Fibroma and Correlations with Literature

Eliana Migliorini Mustafa[1], Victor Rodrigues Ribeiro Ferreira[1], Sofia Braile Sabino[1], Maria Christiane Valéria Braga Braile-Sternieri[1], Rodrigo Serpa Sestito[1], João Carlos Leal[1], Bethina Canaroli Sbardellini[1], Giovanni Braile Sternieri[1], Lúcia Angélica Buffulin de Faria[1], Idiberto José Zotarelli Filho[1,2*] and Domingo Marcolino Braile[1]

[1]Domingo Braile Institute of Sao Jose do Rio Preto (SP), Brazil

[2]State 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) 8166-6537; E-mail: m.zotarelli@gmail.com

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

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 extracorporeal 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).

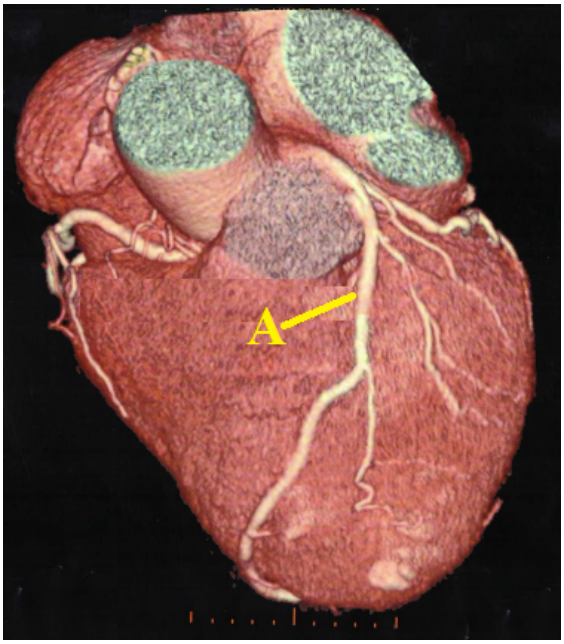

Figure 1: Computed tomography angiography (CTA) cardiac image showing "A" the Surface Myocardial Bridge.

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).

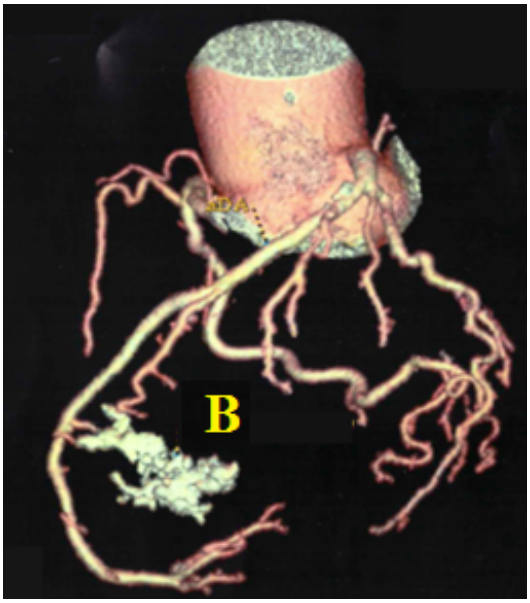

Figure 2: Computed tomography angiography (CTA) cardiac image showing "B" focal calcification.

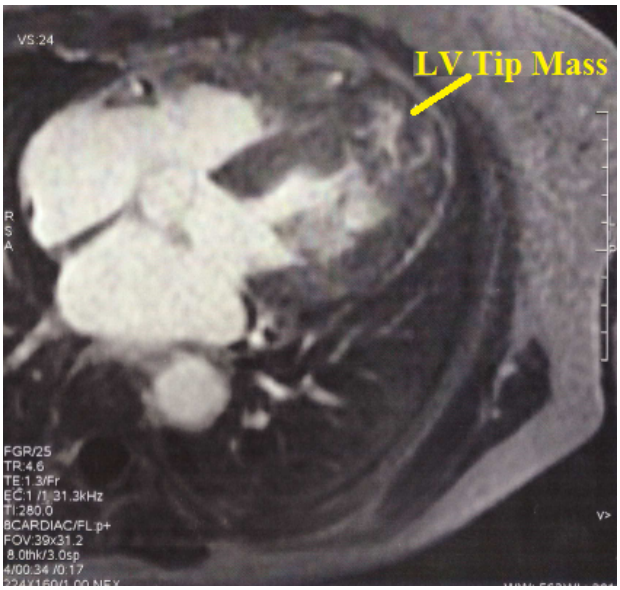

Figure 3: MRI image (2D) showing the Apical mass at the apex of the left ventricle (LV).

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.

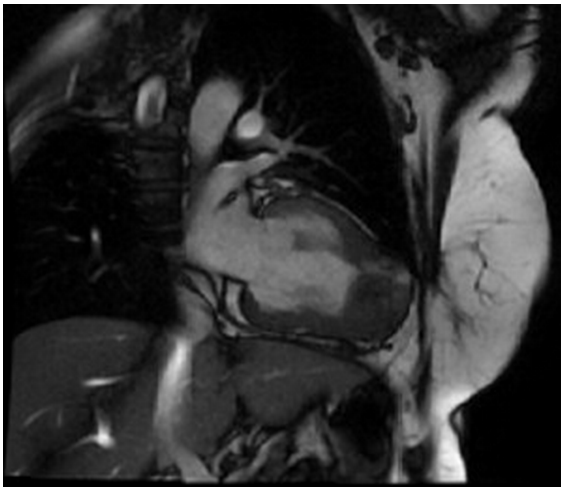

Figure 4: MRI (2D) pre-operative cardiac.

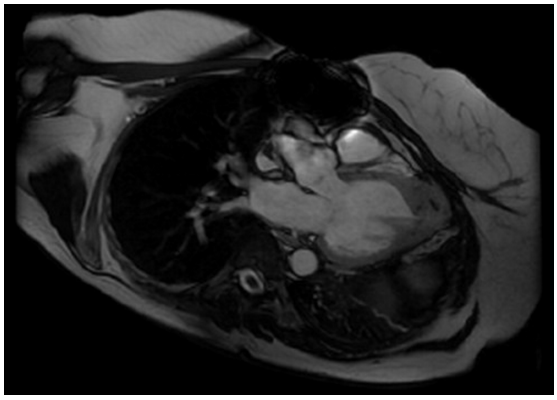

Figure 5: MRI (2D) post-operative cardiac.

Discussion

The present paper describes a case report of very rare apical cardiac HA, with akinesia of the apical region with presence of intramyocardial fibro-calcified mass, presenting intense local coronary vascularization, with dilated epicardial coronary artery at the tip with 5.0 mm, where intramyocardial vessels that irrigate the tumor of the apical region. The MRI result showed an increase in myocardial thickness in the apical region and in the anteroposterior-medial segment of the LV, around 2.2 cm, with irregular appearance and presence of extensive areas of calcification and microaneurysms. This directed strongly to the diagnosis of apical HA. In addition, one characteristic of cardiac HA is the presence of disordered hypertrophied cardiomyocytes and mixed with more vascularized tissue, fibrous and adipose tissue in various proportions [16].

Since minor surgical or biopsy specimens represent the only tissue available for evaluation removed from the ventricular septum, the distinction of HA from hypertrophic cardiomyopathy (HCM) may be impossible microscopically [1,7,15]. Therefore, the two can be differentiated only in light of clinical findings and imaging studies [1]. 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 hyperintense 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].

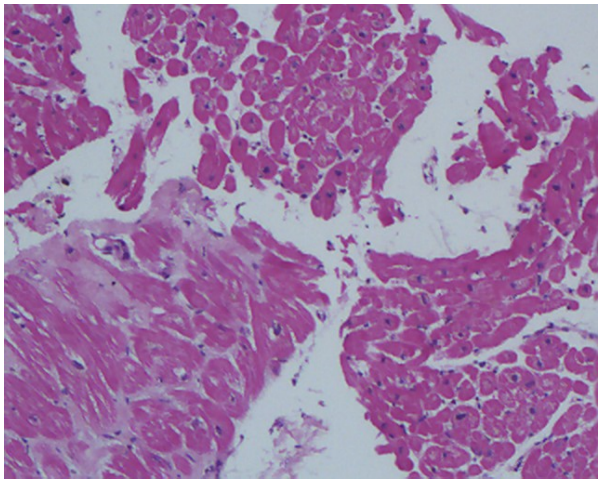

Figure 6: Lesion. (*HE). Increase by 40X. Microscopically, Hamartoma presented dense collagen, adipose tissue, hypertrophic and with disarticulated cardiomyocytes. It is also suggested irregular junction of connexin 43. *HE= Hematoxylin-eosin.

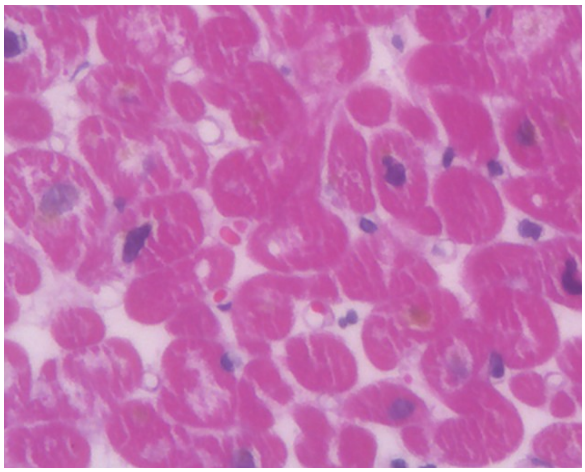

Figure 7: Lesion (*HE). Increase by 100X. Microscopically, Hamartoma presented dense collagen, adipose tissue, hypertrophic and with disarticulated cardiomyocytes. It is also suggested irregular junction of connexin 43. *HE= Hematoxylin-eosin.

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

1. Karigyo CJT, Silva FBF (2014) Tumores cardíacos: uma breve revisão da literatura. Rev Med Res 16: 27-34.
2. Tanimura A, Kato M, Morimatsu M (1988) Cardiac hamartoma. A case report. Acta Pathol Jpn 38: 1481-1484.
3. Burke AP, Ribe JK, Bajaj AK, Edwards WD, Farb A, et al. (1998) Hamartoma of mature cardiac myocytes. Hum Pathol 29: 904-909.
4. Amano J, Nakayama J, Yoshimura Y, Ikeda U (2013) Clinical classification of cardiovascular tumors and tumor-like lesions, and its incidences. Gen Thorac Cardiovasc Surg 61: 435-447.
5. De Martino A, Blasi S, Lorenzini D, Fornaro M, Basolo F, et al. (2016). Lipomatous hamartoma-like lesion of a bicuspid aortic valve: an incidental surgical finding. Cardiovasc Pathol 25: 500-502.
6. Fealey ME, Edwards WD, Miller DV, Menon SC, Dearani JA (2008) Hamartomas of mature cardiac myocytes: report of 7 new cases and review of literature. Hum Pathol 39: 1064-1071.
7. Torii Y, Yamada H, Matsukuma S, Nishio S, Kusunose K, et al. (2016). Left Ventricular Lipomatous Hamartoma Mimicking a Calcified Amorphous Tumor. Circulation 23: e408-e110.
8. Zhang F, Yin N, Yin B, Xu S, Yang Y (2009) Giant right atrial cystic hamartoma: a case report and literature review. BMJ Case Rep.
9. Patini R, Staderini E, Gallenzi P (2016) Multidisciplinary surgical management of Cowden syndrome: Report of a case. J Clin Exp Dent 8: e472e474.
10. Giuseppe MR, Tarelli G, Balzarini L, Torta D, Monti L (2013) Hamartoma of mature cardiac myocytes: a cardiac tumour with preserved Contractility. European Society of Cardiology.
11. Raikhlin A, Felipe ST, Elsie TN (2011) Cardiac hamartoma:Magnetic resonance and computed tomographic imaging. European Journal of Radiology Extra 78: 85-88.
12. Fazio G, Grassedonio E, Cracolici E, Novo G, Sutera L (2009) Cardiovascular magnetic resonance characterization of a hamartoma in an asyntomatic child. International Journal of Cardiology 132: 102-104.
13. Bradshaw SH, Hendry P, Boodhwani M, Dennie C, Veinot JP (2011) Left ventricular mesenchymal hamartoma, a new hamartoma of the heart. Cardiovasc Pathol 20: 307-314.
14. Gómez FJT, Olivera FJT, Gómez AT (2008) Hamartoma of mature cardiac myocytes with limphangiomatous areas. Rev Esp Patol 41: 286-288.
15. Gilman G, Wright RS, Glockner JF, Starrett RS, Hansen WH, et al. (2005) Ventricular Septal Hamartoma Mimicking Hypertrophic Cardiomyopathy in a 41-year-old Woman Presenting with Paroxysmal Supraventricular Tachycardia. Journal of the American Society of Echocardiography 18: 3.
16. Chu PH, Yeh HI, Jung SM, Chien LY, Cheng NF, et al. (2004) Irregular connexin43 expressed in a rare cardiac hamartoma containing adipose tissue in the crista terminalis. Virchows Arch 444: 383-386.

17. Lam KY, Dickens P, Chan AC (1993) Tumors of the heart. A 20-year experience with a review of 12,485 consecutive autopsies. Arch Pathol Lab Med 117: 1027-1031.