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Case Report

Mature cardiac myocytohamartoma: a case report and review of literature

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Abstract: Mature cardiac myocyte hamartoma is a kind of cardiac benign tumor which is extremely rare. Here, we reported the case of a 41-year-old male with cough and shortness of breath. The positron emission tomography computed tomography (PET-CT) showed one tumor in the right atrium, and then the patient had an operation to remove the tumor. Finally, we diagnosed this case as hamartoma of mature cardiac myocytes by histopathology and immunophenotype and also review the tumor which has been published in the literature.

Keywords: Mature cardiac myocytehamartoma, positron emission tomographycomputed tomography, disordered cardiomyocytes, adipose tissue, dilated vessels

Introduction

The primary cardiac tumors have been reported as of low incidence and generally as myxomas, rhabdomyomas, and fibromas during these years [1], while the case reports of mature cardiac myocyte hamartoma have been extremely rare. Hamartoma is a disease in which normal tissues combine and arrange incorrectly in organs, and the disordered changes of number, structure or maturity of these tissues will be constantly growing with the development of the progress of disease [2]. In addition, the mature cardiac myocyte hamartoma is major composed of disordered cardiomyocytes, adipose tissues, dilated vessels in histology [3]. In this article, we have reported a male patient diagnosed as the hamartomas of mature cardiac myocytes in the right atrium, and furthermore we also summarize the reported cases in the literature so far.

Case report

A forty-one-year old man came to our hospital because of a cough with intermittence or episode, and edema of both lower extremities and shortness of breath after daily activities. Importantly, these symptoms had been more and more serious in the last one month. The patient said that he had been prone to having a relapse of cough and dry cough without phlegm since he suffered from a cold without special treatments more than 30 years ago. He had shortness of breath after exercise and limited to generally physical activities three years ago, and then got edema in both lower limbs two years ago. However, he paid no attention to these symptoms as they would disappear after rest. In March 2018, the patient's condition became worse, so he went to Changsha Central Hospital for treatment. The color Doppler echocardiography showed that fluid dark areas could be detected in the pericardial cavity and a large number of pericardial effusions and positive tuberculosis bacilli were found. He was discharged after getting the anti-infection and anti-tuberculosis treatment and came to our hospital to seek further treatment. PET-CT indicated that a pericardial effusion. The outer edge from the right atrium by the superior vena cava had an abnormal increase of glucose metabolism (**Figure 1A**), and it was suspected as malignant mesothelioma. Besides, the patient had Chronic hepatitis without any cardiovascular disease or home history of angiocardiopathy.

The tumor was forwarded to the department of pathology postoperatively, and was performed a surgical resection. It was showed that there was a thickened right atrial myocardium and a

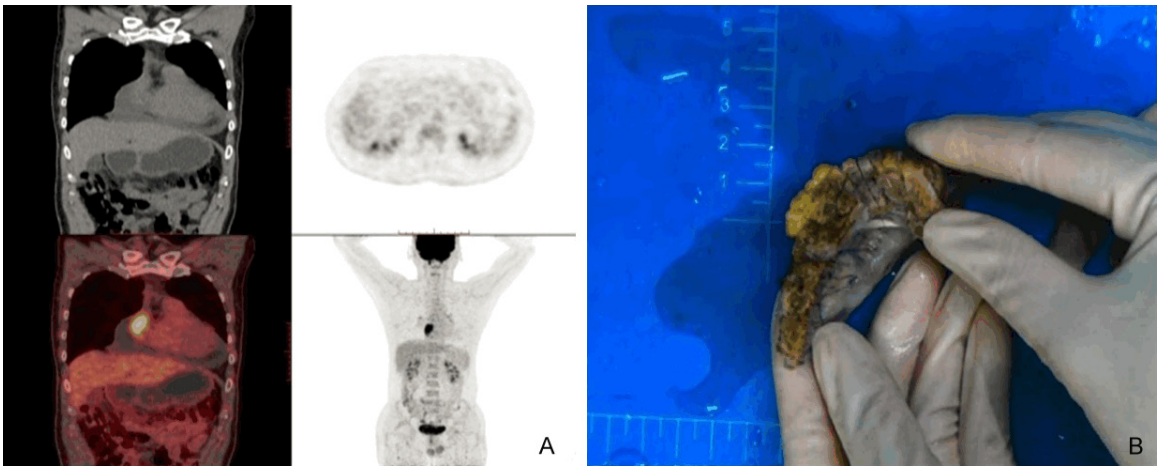

Figure 1. A. The result of PET-CT. B. The postoperative specimen.

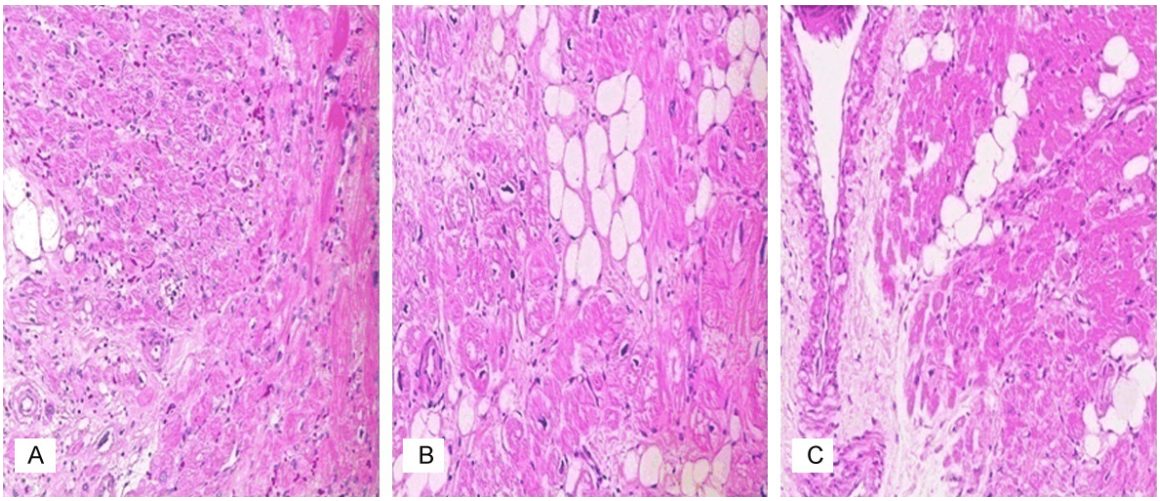

Figure 2. These pictures are the light microscopic results magnified 100 times. We can see the disarray of cardiac myocytes, fat cells and dilated vessels.

2×1×0.5 cm gray yellow lump area which is shown in **Figure 1B**. Microscopy exhibited the cardiac muscle arrangement disorder, some of the nuclei are large, irregular, hyperchromatic and with dozens of adipose cells between the cardiomyocytes. In addition, thickening and dilatation of vascular wall and a group of inflammatory cell infiltration we also perceived and revealed as **Figure 2**. Immunohistochemical results showed that Desmin, S-100, and Vimentin were diffusely positive, and MyoD1 was cytoplasmic positive (**Figure 3**). Incontrast, Ki-67, CD68, and NSE were negative. Based on all the above information, we finally diagnosed this case as mature cardiac myocyte hamartoma. Lastly, the patient was released from the hospital when his pericardial effusion and other symptoms disappeared.

Discussion

In this case, we should identify several kinds of disease. First, hypertrophic cardiomyopathy, which is a disorder of myocardial cell organization, has nuclear enlargements, interstitial fiber proliferation, and more importantly, it usually has a family history [4]. Secondly, consider cardiac rhabdomyoma, which is composed of hypertrophic cardiomyocytes whose cytoplasm is clear. Additionally, we can refer to the spider-like cells which is unique for tuberous sclerosis, and most of which are vacuolated and denatured and contained little myofilament [5].

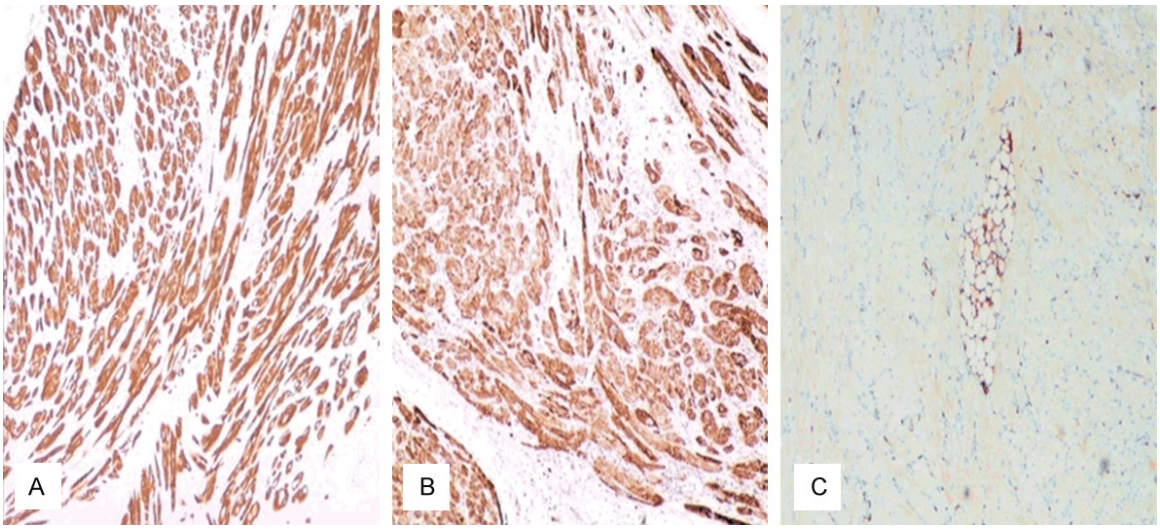

Figure 3. T Immunohistochemical results (A) a strong positivity of desmin. (B) A strong cytoplasmic positivity of myoD1, (C) Positivity of cell membrane with S-100. These images are magnified 100 times and we used DAB staining.

Thirdly, the lipoma of the heart, which is full of fat cells and no disordered cardiomyocytes and dilated vessels. Fourthly, granular cell tumor, which has no striations and vacuoles in cytoplasm, and the NSE is usually positive by Immunohistochemistry [6].

Furthermore, we reviewed articles about the mature cardiac myocyte hamartoma published from 1998 to 2017 (**Table 1**). Among these 20 cases, we have found that the patient's age of this disease ranges from 6 months to 76 years old, and the average age is 30.5 year old, and the males' average age is older than females' and their proportion is probably 2:1 (The male patients were 13 while the female patients were 7 which is shown in the **Table 1**). The clinical symptoms are varied, for instance, it can include palpitations, dyspnea, and episode of syncope or free-symptom. We believe that it may have some connections with the location of the tumor. In these cases, most masses were located in the ventricle, and only four mass were in the right atrium. The size of the tumor ranged from 0.1 cm to 9 cm and the tumor can have no clear boundaries which is easily misdiagnosed as a malignant tumor on imaging [18].

The pathogenesis of mature cardiomyocyte hamartoma is unclear nowadays, but there are some opinions that it may result from congenital embryonic dysplasia [14]. The features of the pathology of congenital embryonic dysplasia are myocyte disarray, fat cells between the myocytes, and the blood vessel expansion. In addition, there are three kinds of the disorder of the myocyte: haphazard, herringbone, and pinwheels. The treatment of hamartomas of mature cardiac myocytes usually is surgical resection, the prognosis of which is grossly satisfactory as the symptoms of the patient can completely disappear. However, when the tumor leads to cardiac outflow tract obstruction or severe arrhythmia, it can also endanger the conditions of patients [1]. Also, there are some diagnoses of mature cardiomyocyte hamartoma in autopsy, but we are not sure that whether the cause of death is related to the disease [7, 16, 17].

In summary, the mature cardiac myocyte hamartomais is extremely rare. It is diagnosed by histopathology according to the images of derangement of myocardial cells, fat tissue, expanded blood vessels, few fibrous tissues and a small amount of inflammatory cell infiltration in the microscope. The patient can have a good prognosis after an operation removing the lump.

Disclosure of conflict of interest

None.

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Table 1. Reported literature of mature cardiac myocytohamartoma

Published year	Author	Patient's age	Sex	Clinical symptoms	Tumor location	Single or multiple	Tumor size	How tumor was found
1998	Burke AP et al. [7]	22	Male	None	Right ventricle	Single	5 cm	Echocardiogram
1998	Burke AP et al. [7]	28	Male	Wpw syndrome and an episode of syncope	Right atrium	Single	Not clearly	Echocardiogram
1998	Burke AP et al. [7]	9	Male	Sudden death	Right atrium	Multiple	1 mm-2 cm	Autopsy
2001	Dinh MH et al. [8]	33	Male	Generalized tachycardia	Left ventricle	Single	4.5×3.1×4.4 cm	Echocardiogram
2004	Chu PH et al. [9]	76	Male	History of hypertension	Crista terminalis	Multiple	5×5×5-10×5×5 mm	In surgery
2005	Martínez QM et al. [10]	33	Male	Palpitations and dyspnea	Left ventricle	Single	4.5×5.5 cm	Echocardiogram
2008	Movahedi N et al. [11]	58	Male	Progressive dyspnea	Right atrium	Single	1.5×1×0.5 cm	In surgery
2008	Fealey ME et al. [12]	6 months	Male	None	Not clearly	Not clearly	Not clearly	Not clearly
2008	Fealey ME et al. [12]	6 months	Female	None	Not clearly	Not clearly	Not clearly	Not clearly
2008	Fealey ME et al. [12]	14 months	Male	None	Not clearly	Not clearly	Not clearly	Not clearly
2008	Fealey ME et al. [12]	10	Male	None	Left ventricle	Not clearly	5×3 cm	Echocardiogram
2008	Fealey ME et al. [12]	16	Female	None	Right ventricle	Not clearly	8×9 cm	Echocardiogram
2008	Fealey ME et al. [12]	57	Male	Sudden death	Not clearly	Not clearly	Not clearly	Not clearly
2008	Fealey ME et al. [12]	74	Male	Exertional dyspnea	Not clearly	Not clearly	Not clearly	Not clearly
2009	Hsu PS et al. [1]	19	Female	Intermittent palpitations and dizziness	Left ventricle	Single	4×7 cm	Echocardiogram
2009	Galeone A et al. [13]	56	Female	None	Pulmonary infundibulum	Single	9×9×4 cm	Chest CT
2011	Dell'Amore A et al. [14]	35	Female	Palpitations and dyspnea	Left ventricle	Single	4.2×3.3×2.7 cm	Echocardiogram
2013	Raffa GM et al. [15]	41	Female	Chest pain	Right atrium	Single	2.5×1.3 cm	Echocardiogram
2017	Ayoub C et al. [16]	14	Male	None	Left ventricle	Single	9×5×6 cm	Echocardiogram
2017	Hadravská Š et al. [17]	39	Female	Ruptured aneurysm and severe pneumonia	Left ventricle	Single	4.5×3×3 cm	Autopsy

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