Case report

A connective tissue pericardial hamartoma: A case report with literature review of cardiac hamartoma

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\textbf{ABSTRACT}

\textbf{Introduction:} Hamartoma is a tumor that can manifest anywhere in the body and results in the abnormal formation of tissue native to the anatomical location. It is usually a benign tumor that rarely arises from the heart or pericardium. Most of the cases are asymptomatic and discovered incidentally during the evaluation of other medical conditions.

\textbf{Presentation of case:} We present a case of a 65-year-old female with dyspnea who was diagnosed with a mediastinal mass. After removing the mass surgically, the mass was found to be heart-like containing four chambers located in the pericardium, and a pathology report of a connective tissue hamartoma.

\textbf{Conclusion:} Pericardial tumors including hamartoma should be considered in all patients with wide mediastinum and cardiopulmonary symptoms. We also present a review of the literature on the cardiac and pericardial hamartomas with a comparison of age, gender, location, symptoms, and management. Although cardiac hamartomas in general and epicardial hamartomas, in particular, are extremely rare, they should be considered in adults with cardiopulmonary symptoms and widened mediastinum.

1. Introduction

This case has been reported in line with the SCARE criteria \cite{1}. Pericardial tumors are scarce that can be either benign or malignant. The majority of pericardial tumors are secondary to invasion from nearby tumors and hematogenous or lymphatic spread \cite{2}. The most common primary malignant pericardial neoplasms are mesothelioma followed by angiosarcoma, lymphoma, and liposarcoma \cite{2}. On the other hand, the pericardial cyst is considered the most frequent benign pericardial tumor followed by lipoma \cite{2}. Whereas Hamartoma is mostly a benign mass, with a possibility of malignant transformation, that consists of disorganized tissue similar to its anatomical location of occurrence. Hamartoma results from the abnormal development of embryonic cells and can occur anywhere in the body but is mostly found in the hypothalamus, breast, lungs, colon, etc. \cite{3}. Hamartomas affect males more commonly than females with no apparent evidence of racial tendency, with an age range for occurrence in patients between 40 and 70 \cite{4}. Most hamartomas incidence is still unknown except for the pulmonary hamartoma with a 0.25 % incidence rate which represents about 8 % of lung tumors; usually, these cases are discovered while assessing the patient for other medical conditions as they are asymptomatic \cite{3,4}. Here we present a case of an elderly woman with a heart-like Intrapericardial hamartoma with a literature review of previous cardiac hamartoma.

2. Case report

A 65-year-old female was referred to the thoracic surgery department by a cardiologist due to second-degree dyspnea. Upon examination, her vital signs were as follows: heart rate 65 beats/min, blood pressure 150/100 mmHg, and respiratory rate 22 breaths/min. Further examination of the cardiovascular and respiratory systems was
unremarkable, her cardiac ejection fraction was within normal limits at 60%, and her electrocardiogram (ECG) showed normal sinus rhythm, left axis deviation, poor R-wave progression in the precordial leads, and an inverted T wave in V1 to V6, avf, and ||| (Fig. 1). Her previous medical history included controlled essential hypertension which is controlled with bisoprolol and furosemide, and her family history was unremarkable. Her laboratory tests including complete blood count, thyroid function tests, and kidney function tests were within normal limits. A chest X-ray revealed a widening mediastinum suggesting a mediastinal mass (Fig. 2). Computerized tomography (CT) scan revealed an oval opacity measuring 8 * 6.5 * 5 cm with regular borders and a density of 70 Hounsfield units situated on the lateral border of the superior vena cava next to its entrance into the right atrium causing an impression on the atrium with no signs of real histological infiltration. Based on the clinical manifestations and investigations, a decision to perform surgery was made by a Thoracic Surgery Consultant. During surgery and after opening the pericardium, an oval-shaped heart-like Intrapericardial mass containing four chambers was seen and excised (Figs. 3-4, Video 1). After dissection, a heart-like structure was seen, as it was composed of a muscular wall and four chambers (Fig. 5).
Histological study revealed regular fibroblasts, blood and lymphatic vessels, and lymphoid tissue. Focally, there was mature adipose tissue, blood, fibrin, inflammatory cells, foam macrophages, hemosiderin deposits, and regular mesothelial cells lining the mass. The result was suggestive of a connective tissue hamartoma with no signs of malignancy.

The post-operative period was uneventful except for an asymptomatic transitional atrial fibrillation. She has been on regular follow-up for three years without complications.
3. Discussion and conclusion

Primary cardiac tumors are extremely rare and affect all ages with a frequency of occurrence of approximately 0.02 % [5]. However, those originating initially from the pericardium are even less common with prevalence ranging from 0.001 to 0.007 % [2]. Benign pericardial tumors include pericardial cysts, lipomas, angiomas, lymphangiomas, fibromas, and teratomas. Hamartoma is considered one of the rarest primary benign cardiac tumors which consists of overgrowth of a mixture of adult tissue with the capability for malignant transformation.

Fig. 4. An oval-shaped heart-like intrapericardial mass.
It was described initially in 1951 [6], and until March 2022, only 34 cases were mentioned in the literature (Table 1) [6–11]. Only two of them are located in the pericardium [6,9]. The first study reported a 38-year-old white woman diagnosed accidentally with pericardial hamartoma, lymphatic in type [6]. While the second one reported a 66-year-old male who presented with atrial fibrillation and was eventually diagnosed with fatty pericardial hamartoma [9]. In this study, we reported for the first time a 65-year-old female presented with dyspnea and was eventually diagnosed with a connective tissue pericardial hamartoma.
Table 1

Summary of cardiac hamartoma cases in literature.

| Year | Age | Sex | Location | Presentation | Management |
|------|-----|-----|----------|--------------|------------|
| 1951 | 38  | M   | Epicardium | Sensation of flopping in the left chest. | Surgical excision |
| 1988 | 68  | M   | LV apex   | Discovered during autopsy | – |
| 1989 | 66  | M   | Epicardium | Atrial fibrillation | Surgical excision |
| 1998 | 24  | M   | Interventricular Septum | Palpitation | Complete resection |
| 1998 | 9   | M   | Multiple; left ventricle free wall, posteromedial papillary muscle, atrial wall, right ventricle | Sudden death, discovered during autopsy | – |
| 1998 | 22  | M   | RV free wall | Asymptomatic | Complete resection |
| 1998 | 28  | M   | RA         | Syncope | Complete resection |
| 2001 | 33  | M   | LV apex   | SOB, light headedness, palpitation | Complete resection |
| 2004 | 76  | M   | RA         | Chest tightness on exertion | Complete resection |
| 2005 | 41  | F   | Interventricular Septum | SVT, palpitation, exertional angina | Partial resection |
| 2005 | 35  | M   | Posterolateral wall of LV | Palpitation, dyspnea | Partial resection |
| 2008 | 6   | M   | RA         | Respiratory distress, Discovered during autopsy | – |
| 2008 | 6   | F   | LV         | Respiratory distress, Discovered during autopsy | – |
| 2008 | 14  | M   | LV         | Respiratory distress, sinus tach | Resection |
| 2008 | 10  | M   | LV         | Asymptomatic | Complete resection |
| 2008 | 16  | F   | RV         | Asymptomatic | Biopsy |
| 2008 | 57  | M   | LV         | Sudden death, discovered during autopsy | – |
| 2008 | 74  | M   | LV         | Exertional dyspnea, discovered during autopsy | – |
| 2008 | 58  | M   | RA         | Chest pain, dyspnea | Partial resection |
| 2008 | 16  | F   | Posterior wall of RV | Weight loss | Biopsy |
| 2009 | 11  | M   | RA wall    | Palpitation, chest distress | Complete resection |
| 2009 | 19  | F   | LV         | V tach | Heart transplanted |
| 2009 | 56  | F   | Pulmonary infundibulum | Pericardial effusion | Complete resection |
| 2011 | 35  | F   | Left posterio-inferior ventricular wall | Chest tightness, dyspnea, palpitation | Partial resection |
| 2011 | 43  | F   | Inferior septum and inferior wall of LV | A fib, v tach, dyspnea | Biopsy, follow-up |
| 2013 | 41  | F   | RA         | Chest pain | Complete resection |
| 2017 | 14  | M   | LV         | Asymptomatic, autopsy pathobiology | – |
| 2017 | 39  | F   | LV         | Discovered during autopsy | – |
| 2018 | 44  | F   | LV apex   | Asymptomatic, incidental discovery | Complete resection |
| 2018 | 23  | F   | RA         | Paroxysmal nocturnal dyspnea, palpitations and edema of the lower limbs | – |
| 2019 | 41  | M   | Outer edge of RA | Exertional dyspnea, lower limbs edema | Complete resection |
| 2019 | 41  | M   | Right auricle | Progressive dyspnea after exercise | Complete resection |
| 2021 | 7   | M   | LA, connected to mitral valve | Incidental while investigating suspected Kawasaki disease | Complete resection |
| 2021 | 62  | F   | LV         | Chest pain, palpitation | Complete resection |

Furthermore, in this study, we included all the 34 cases of epicardial, myocardial, and endocardial hamartoma in one table (Table 1). We classified them according to the following: Year, Age, Sex, Location, Presentation, and Management (Table 1) [6–11]. The symptoms of cardiac hamartoma vary from one patient to another according to the size and location of the tumor. It might present with dyspnea, palpitations, syncope, chest pain, and other cardiopulmonary symptoms (Table 1). Furthermore, cardiac hamartoma can be asymptomatic and discovered incidentally in the contest of investigating another health issue, during a routine health check, or in some cases, it was discovered during an autopsy (Table 1). In the previously reported cases, cardiac hamartoma was more common in males (59 %) than females (41 %) (Table 1). The range of age was between 6 months and 76 years, and it was more common in the second and fifth decades (18.75 % each), followed by the first decade (15.6 %), third and fourth decades (12.5 % each), and the sixth, seventh, eighth were (9.4 %, 6.25 %, 6.25 %) respectively (Table 1). The most common place of origin was the left ventricle (LV) as half of the cases (50 %) originated from it, followed by eight cases (25 %) from the right atrium (RA), four cases (12.5 %) from the right ventricle (RV), two cases (6.25 %) from the interventricular septum, one case (3.1 %) from the left atrium (LA), and one case (3.1 %) were multiple tumors (Table 1). The optimum management of benign cardiac tumors in symptomatic patients is surgical resection; however, three cases were diagnosed with a biopsy and followed up accordingly, and one case was managed with a heart transplant.

The patient in our case presented with second-degree dyspnea; therefore, the optimal management was surgical resection. The outcome was satisfactory as there were no complications or residual symptoms after the follow-up over three years.

In conclusion, although cardiac hamartomas in general and epicardial hamartomas, in particular, are extremely rare, they should be considered in adults with cardiopulmonary symptoms and widened mediastinum.

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Ethical approval

The study was approved by ethics committee of Damascus University.

Consent

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Author contributions

HNA played a role in writing the first draft and revising the final...
draft. MK wrote the first draft and revised the final draft. AI participated in writing the first draft and revising the final draft. BD revised the final draft and followed the patient up.

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Declaration of competing interest

The authors declare that there is no conflict of interest.

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