JACOBS PUBLISHERS

OPEN ACCESS

Journal of Molecular Biomarkers and Clinical Trials

Case Report

Rapidly Progressing Left Atrial Primary Undifferentiated Pleomorphic Sarcoma

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Received: 05-16-2016

Accepted: 06-02-2016

Published: 06-10-2016

Abstract

Primary cardiac tumors are rare. Approximately, a quarter of those tumors are malignant and cardiac sarcomas account for 75% being the most common subtype of primary cardiac malignancies. A 61-year-old man was admitted to our hospital for progressive exertional cough, and dyspnea. Transthoracic echocardiography showed a large intra-cardiac mass originated from the roof of left atrium with a wide base. However, the histological diagnosis was a cardiac undifferentiated pleomorphic sarcoma. We present a case of primary undifferentiated pleomorphic sarcoma of the left atrium exhibiting very early recurrence after the first resections.

Keywords: Primary cardiac tumor; Sarcoma; Resection; Recurrence

Introduction

Primary cardiac tumors are extremely rare that most cardiac surgeons would not practice this disease sufficiently. In one autopsy report [1], the incidence is only 0.0001% to 0.0003%. Of these cardiac tumors, about a quarter is malignant with sarcoma being the most common subtype accounting for 75% of the malignant neoplasms [2]. More recently, in the largest published series [3], all subtypes of left atrial sarcomas incidence are 33% of the total primary cardiac sarcomas resected surgically. Recurrence is common. Herein, we reported the first recurrence of undifferentiated pleomorphic sarcoma within 34-day period post-surgery.

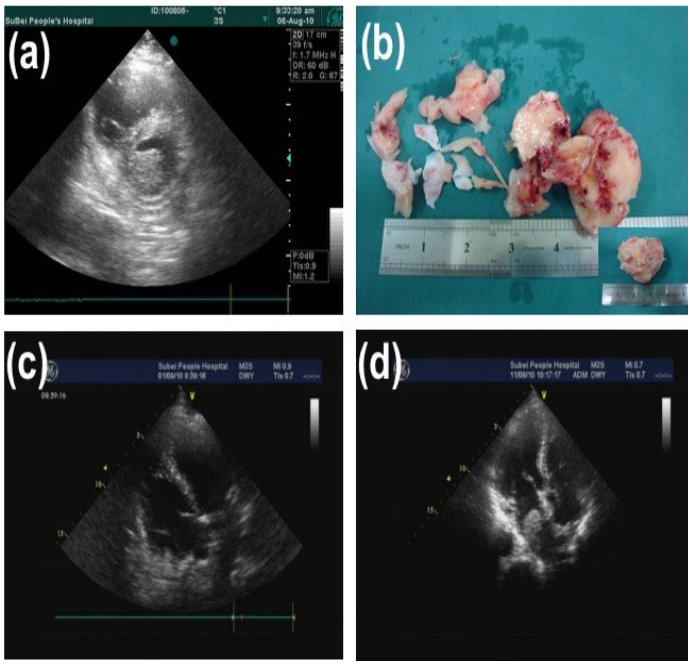
Case Presentation

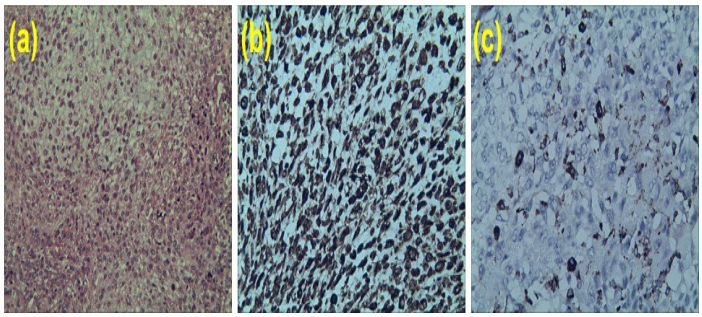
A 61-year-old male complained of dyspnea and chest pain on exertion with irrelevant medical history. On physical examination, his pulse was 85/min and his blood pressure was 150/80mmHg. On auscultation, no murmurs or other additional sounds. Chest x-ray, blood work, and coronary angiogram were normal. ECG displayed normal sinus rhythm with incomplete right bundle branch block. Transthoracic echocardiogram (TTE) revealed a large wide-based intra-cardiac mass originating from the left atrial roof, and normal bi-ventricular function (Figure 1a). Metastatic work up using computed tomography scanning showed no evidence for distant metastasis.

Due to deterioration of the patient's symptoms and progressive paroxysmal supraventricular tachycardia (SVT), he underwent emergent operation. Intra-operatively, transesophageal echocardiogram (TEE) confirmed the TTE findings. Median-sternotomy with mild hypothermic cardiopulmonary bypass (CPB) and trans-atrial approach were used. The tumor was identified and resected completely (Figure 1b). It was originated from the left atrial roof, close to right superior pulmonary vein and extended to the lower part of the left atrium. The mass was 4 × 3 × 5 cm in size and was white and gray in color. In one block, we resected a mass of 5 × 7 × 5 cm, resection margins reached normal tissue of left atrium as per frozen section, and the tumor infiltrated the atrium wall thickness entirely. The defect of left atrium was reconstructed with a bovine pericardium patch.

On coming off CPB, the patient resumed normal sinus rhythm, and weaned-off ventilation 12 hours post-operatively. He did well with good hemodynamics till postoperative day (POD) # 4 when he experienced SVT ended up by repeat VF cardiac arrest in spite of giving multiple anti-arrhythmic therapy. Inotropes were initiated and DDD permanent pacemaker was inserted successfully followed by inotropes withdrawal. On POD # 24, due to persistent SVT, TTE was done and revealed normal structure of the heart chambers and there was no evidence of tumor recurrence (Figure 1 c). Later, on POD # 34, because the patient starts a short of breath again, TEE was done and showed a new mass with a size of 2 × 3 × 3cm (Figure 1 d) originating from the left atrial septum side. The patient declined taking him back for re-resection using auto-transplantation and he expired on POD # 39 due to irreversible circulatory collapse.

On microscopy, with Hematoxylin and Eosin (Figure 2 a) and immunohistochemical staining (Figure 2 b and 2 c), the mass revealed criteria well-matched with a histological diagnosis of undifferentiated pleomorphic sarcoma. The free margin of the tumor was confirmed with the convention histopathology indicating that none of the tumor tissues was left behind.





Discussion

Usually, primary malignant neoplasms of the heart develop from the right side of the heart, particularly in the right atrium. For undifferentiated pleomorphic sarcoma, it usually arises in the left atrium [4] and inclines to be located on the left atrial posterior wall or on the left side on the interatrial septum [5]. Surprisingly, histopathological diagnosis of this subtype of tumor is mainly achieved by exclusion after complete failure of different immunohistochemical stains to have a proof of specific differentiation of other kinds of sarcomas. Therefore, we are anticipating a decline of incidence of undifferentiated pleomorphic sarcoma over time where the immunohistochemical stains will be available for every histopathology institution to diagnose more differentiated tumors on the expense of undifferentiated sarcomas.

Generally, primary sarcomas of the heart behave aggressively and usually have a deadly outcome [3]. Such potential fatal prognosis is because this tumor grows up intracavitary impeding blood flow and causing rhythm disturbances as in our case. Moreover, this tumor could develop systemic embolization resulting in a stroke formation or organ infarction. Recently, a case of amaurosis fugax has been reported due to embolization resulted from left atrial undifferentiated pleomorphic sarcoma [6]. In most of the cases, left atrial sarcoma causes dyspnea and arrhythmias due to encumbering blood flow [7]. Therefore, it presents late and carries poor prognosis. Local tumor recurrence remains the most frequent cause of mortality [8]. Hence, frequent TTE follow up for recurrence was encouraged post-surgery [7]. Recently, multiple recurrences after repeat resection with [9] and without concomitant metastasis [10] associated with poor outcomes have been published. Having clear surgical margins intraoperatively is not guarantee for complete cure due to high probability of missed microscopic disease to adjacent structures. However, the surgeon has to be wise in having large free margin and keeping behind a sufficient myocardium for performing heart [11]. Surprisingly, orthotopic cardiac transplantation did not prolong the survival significantly [12].

Our case is a new presentation of very early recurrence of such disease after the first resection in spite of having a free margin on the definitive conventional histopathology results. Based on the fact that tumor-free margin was achieved, in addition to excision of the tumor in one chunk, we think our case is likely to be an early recurrence of the tumor rather than being a growing tumor, which was not found out during surgery or a secondary dissemination of the tumor during our surgical intervention. Other investigators reported recurrence within a month after the second resection [10] and none of the recurrences reported after the first resection was early like our case.

In our case, we think that once a left atrial sarcoma has progressed to the point that symptoms start to occur, it has often

spread to other adjacent parts of the heart like pericardium, making treatment difficult and challenging. Because of insufficient data for chemotherapy and radiotherapy due to rarity of the tumor, the benefit of such adjuvant therapy is still controversial. However, we believe in complete surgical resection as the primary goal in addition to multimodal therapy.

We had the plan of multimodality therapy, starting with complete surgical excision as the primary therapy, followed by chemotherapy and radiation adjunctively according to a big series with long follow up of cardiac sarcomas [13]. Our patient has been seen by the Oncology and Cardiology service for evaluation and to arrange for out-patient chemotherapy/radiation therapy post-surgery. Unfortunately, due to hemodynamic instability of our patient, he did not achieve this multimodality therapy. We did not offer our patient orthotopic cardiac transplantation to avoid subsequent immunosuppression therapy that might lead to developing of new tumors.

Conclusion

To the best of our knowledge, this is the first case report of primary undifferentiated pleomorphic atrial sarcoma that recurred within 34 days of the first complete resection in absence of concomitant distant metastasis. We recommend more frequent monitoring for recurrence to improve the prognosis.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Conflicts of interest statement

None declared.

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