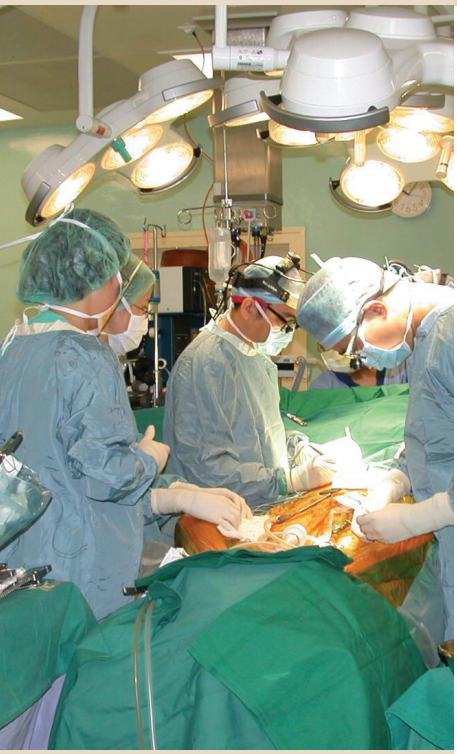
A Rare Case Report of Left Atrial Angiosarcoma



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Abstract

The most primary cardiac tumors are benign, and malignant tumors comprise about 25%. Primary cardiac angiosarcoma is the most common malignant tumor and aggressive tumor with a high incidence of metastatic spread. The diagnosis is often delayed because of the nonspecific clinical presentation. Cardiac angiosarcoma usually arises from the right atrium. We report an extremely rare case of primary angiosarcoma originating from the left atrium in a 57-year-old woman that had been referred with a one month history of exertional dyspenea. Transesophageal echocardiography showed a large mass on the left atrial that protrude through mitral valve. The tumor was resected successfully with curative intent. The patient underwent adjuvant chemotherapy. We have no problem up to now in six months follow-up.

Key words: cardiac tumors, primary cardiac angiosarcoma

Introduction

Primary cardiac sarcoma is a rare clinical entity, with an incidence of 0.0001% in collected autopsy series. Sarcomas are the main malignant primary heart tumor, with higher prevalence in men between the 3rd and 4th decades of life, located predominantly in the right atrium [1, 2]. As benign lesions, the clinical presentation of malignant cardiac tumors depends on location and not the histological type. No typical presentation of cardiac sarcoma exists because the common symptoms and signs are nonspecific. However, patients may complain of dyspnea, chest pain, and/or generalized fatigue [3,4]. Angiosarcoma is the most common sarcoma, with high incidence of metastasis, poor prognosis, and therapy without consensus . Echocardiography is the preferred diagnostic procedure for noninvasive imaging of cardiac tumors (2-dimensional and transesophageal echocardiography are complementary) [6]. The majority of the literature describes a uniformly dismal prognosis with a median survival of only 6 months for these aggressive tumors. Standard surgery, adjuvant chemotherapy, and radiotherapy have been consistently unsuccessful [13, 14]. Early heart transplantation and novel radiation therapy approaches may offer a survival benefit in nonmetastatic tumors, but up to 80% of the patients present with systemic metastasis at diagnosis [15]. Though several chemotherapeutic regimens have been tried, the role of chemotherapy is not well established and outcome data available is minimal.

CASE REPORT

A 57-year old woman was referred to the Department of Cardiovascular Surgery of our hospital to be submitted to an urgent surgery following an initial diagnosis of

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a left atrial tumor, probably myxoma. She was presented with one month history of progressive exertional dyspnea and respiratory distress, ralls and borderline hemodynamic on admission. Chest X-ray showed cardiomegaly, hilar adenopathy, pulmonary congestion and pleural effusion. The electrocardiogram (ECG) showed sinus tachycardia and nonspecific repolarization changes. The peroperative transthorasice echocardiogram showed hemodynamic compromise caused by very large LA mass that protrude through mitral valve.TEE showed a giant mass (90 ×60 mm) in left atrium that infiltrated interatrial septum and left atrium auricle and protruded into mitral valve (Figure .1, 2). From the morphological aspect, the left atrial mass was suggestive of a cardiac myxoma. Left ventricular function was normal and there were no valvular abnormalities.

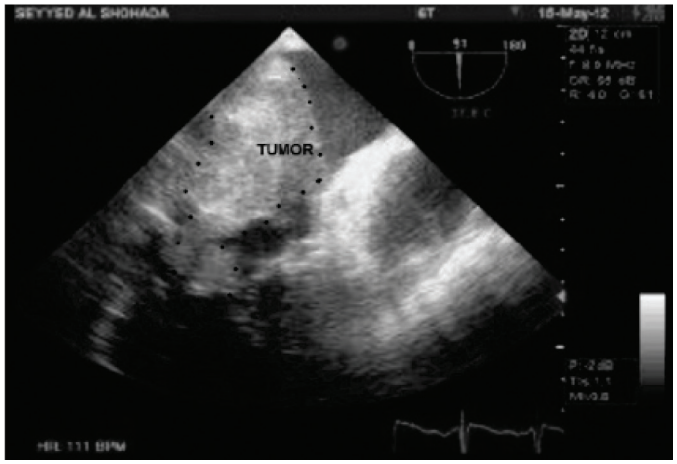

Figure 2- echocardiographic view of large left atrium tumor

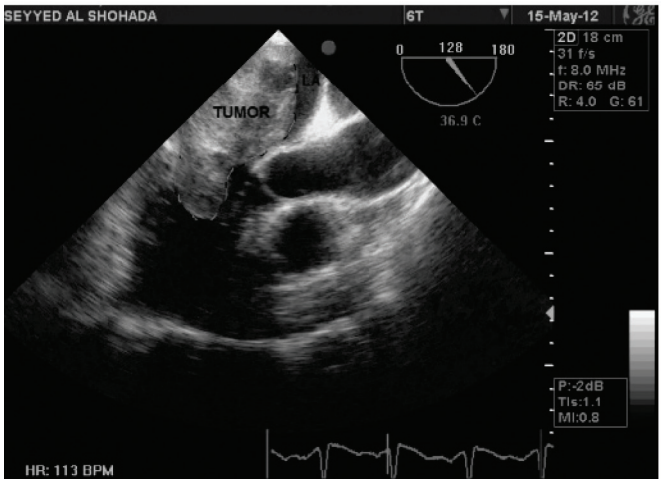

Figure 1-echocardiographic view of large left atrium tumor

The patient underwent surgical excision of the tumor. During the operation, the tumor was excised through a sternotomy from a biatrial approach.

The right and left atriotomy was performed. interatrial septum (fossa ovalis) was incised and stay suture with 2/0 ethibond was done. Initially the tumor seemed to be small, but progressive exploration and resection of tumor showed that the tumor was very large and adherent to left atrial wall. The tumor was protruded via mitral valve but not adherent to mitral leaflets. When tumor resection was completed, mitral valve tested with saline test and showed no significant mitral regurgitation. Defect lesion in posterior left atrial wall was repaired with 4/0 prolene and pericardial patch. interatrial septum defect was closed with pericardial patch . The tumor was resected successfully with curative intent. The resected tumor was lobulated and 85 × 75 × 45 in size

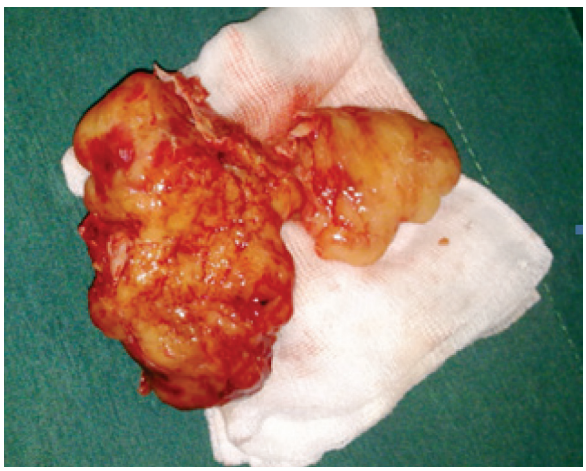

Figure 3 - Macroscopic photograph; resected large tumor of the left atrium

Histopathologic examination showed a hemorrhagic and necrotic malignant tumor (massive necrosis> 50%). The tumor contained solid areas and anastomosing vascular spaces lined by spindle-shaped cells with pleomorphic nuclei and numerous mitotic activities. Immunohistochemically, the tumor cells were positive for the endothelial markers factor VIII-related protein, CD31 and CD34. Diagnosis was high grade angiosarcoa. Postoperative recovery was uneventful and the patient was discharged on the eleventh postoperative day. After six months, the patient is fortunately symptom-free. Two months later CT-scan showed no lung and liver metastases without evidence of local tumor

recurrence. Chemotherapy was started (Adriamycin and doxorubicin). Six months later control echocardiography showed no mass or local recurrence.

Discussion

Primary tumors of the heart are extremely rare, occurring at a frequency of 0.02% in autopsy series and the majority of them are benign. In adults, approximately 75% of primary cardiac tumors are benign, with myxoma accounting for up to half of cases. The remaining 25% of primary cardiac tumors are malignant [1, 4]. Metastatic heart tumors are 20–40 times more frequent than primary cardiac tumors [1]. Sarcomas represent the commonest histology of malignant primary cardiac tumors, accounting for 20% of all cardiac neoplasms. Still, primary cardiac sarcoma is a rare clinical entity, with an incidence of 0.0001% in collected autopsy series [2]. The majority of cardiac sarcomas occur between the third and the fifth decades of life with a male preponderance (M: F ratio 2: 1) [1, 2]. The most common site of involvement is the right atrium, followed by left atrium, right ventricle, and left ventricle [3]. Ninety percent of the angiosarcomas are located in the right atrium. The lateral (free) wall of the right atrium is the most common site, the septum being spared in most cases [4]. The clinical signs and symptoms are often nonspecific. Dyspnea is the most common presenting symptom; additional symptoms include atypical chest pain, hemoptysis, orthopnea, and nonspecific symptoms such as nausea, emesis, fever, and anorexia [4]. More than half of these tumors have produced systemic metastases at the time of discovery, most commonly to the lungs; other sites include the liver, brain, and bone [4, 5]. Echocardiography has become the primary diagnostic technique because of its high degree of accuracy, non-invasiveness, and cost effectiveness. It is sufficient to provide enough information to diagnose an intracardiac lesion; however, it cannot be used accurately to differentiate between benign and malignant lesions [6]. Other modalities, such as MRI and CT are being used and are particularly useful to detect primary and secondary cardiac malignant tumors. They can detect tumors 0.5–1 cm in size. [6,7]. TEE has a higher resolution (1–3 mm) than MRI (5–10 mm), while MRI is better to identify tissue composition with ability to differentiate solid, liquid, hemorrhagic and fatty structure[1]. A less invasive procedure is the transvenous endocardial biopsy but it often false-negative. This procedure is not recommended due to the friability of the tumor and a predisposition to bleeding, with high morbidity [8]. Histopathology defines angiosarcoma as a malignant tumor whose cells display endothelial differentiation [9]. The diagnosis of cardiac angiosarcoma can be confirmed by additional immunohistochemical staining for endothelial markers, of which CD31 and factor VIII-related protein are most specific [10]. Most reported series of cardiac sarcomas describe patients with primary cardiac sarcomas and response to treatment and survival is anecdotal. Complete resection of cardiac sarcoma is difficult, in view of the location and extent of involvement. Surgical resection is indicated when no evidence of metastasis exists and when myocardial resection is reparative. Often tumors are so large at the time of the operation that complete resection cannot be done [11]. In general, recommendations for the treatment of nonmetastatic cardiac sarcoma include exploration for local control of the primary tumor, to relieve obstructive symptoms and to prolong disease-free survival [12]. Cardiac sarcomas generally have a dismal prognosis with a median survival of only 6 months [13]. Applying the general principles of treatment of soft tissue sarcomas occurring anywhere else in the body, the most critical element is complete surgical resection; however, the location itself is more difficult for obtaining an adequate margin of resection. The emphasis is on early detection and diagnosis. In one study, Patients with complete resection have a survival of 24 months compared with 10 months in those with incomplete resection [14]. Chemotherapy and irradiation were reported not to improve survival and their use is usually limited due to the poor physical condition of the patient [13]. Cardiac transplantation has been performed in a few patients, however, with a poor outcome. There is no evidence that cardiac transplantation improves the overall poor prognosis of these patients [15]. The prognosis of cardiac angiosarcoma is universally poor: survival ranges from six to nine months, regardless of the treatment chosen. Death results from infiltration of the myocardium, cardiac tamponade, obstruction of flow, and/or distant metastases [13].

In our case, we report an extremely rare case of primary angiosarcoma originating from the left atrium because Cardiac angiosarcoma usually arises from the right atrium. In a literature review through PUBMED from 1980 to 2008, more than 250 cases of cardiac angiosarcoma were found;

the tumors mostly arose from the right atrium with different extensions and clinical presentations. In the last two decades, only eight cases of this malignant tumor originating from LA were reported in the English literature [16].

Review of the literature reveals that this case of cardiac angiosarcoma is unusual by virtue of its occurrence in a female. The mean age of presentation for all primary cardiac angiosarcoma is 40 years with the male to female ratio of 2:1. From the analysis of the nine reported cases with left atrial angiosarcoma, seven of them are female; the mean age on presentation is 60.2 years [16]. Clinical presentation of left-sided cardiac angiosarcoma often mimics left atrial myxoma. During surgery sometimes it is needed to mitral valve repair or replacement because of mitral valve involvement [17].

In conclusion, Cardiac tumors have nonspecific signs and symptoms.Most clinical presentation reflects hemodynamic impaired. Left-sided cardiac angiosarcoma often mimics left atrial myxoma. Primary left-sided cardiac angiosarcoma is a rare disease; unlike right-sided angiosarcoma, it usually affects females, and it usually presents late in their lifetime. Because of the rarity of cardiac angiosarcoma, there are no generally applicable guidelines for treatment. Surgery, adjuvant chemotherapy, and radiation have been used. Despite aggressive treatment, the prognosis is poor, and death usually occurs within one year after the diagnosis.

References:

1. L. Antonuzzo, V. Rotella, F. Mazzoni,et al." Primary Cardiac Angiosarcoma: A Fatal Disease". Case Reports in MedicineVolume 2009 (2009), Article ID 591512, 4 pagesdoi:10.1155/2009/591512.
2. Dhatri Kodali, and Kala Seetharaman. "Primary Cardiac Angiosarcoma". Sarcoma, 2006; 2006: 39130.
3. Monique Esteves Cardoso,Leonardo Secchin Canale, Rosana Grandelle Ramos,et al ." Cardiac Angiosarcoma". Case Reports in Cardiology , Volume 2011 (2011), Article ID 340681, 3 pagesdoi:10.1155/2011/340681.
4. Wobbe Bouma, Chris PH Lexis, Tineke P Willems,et al. "Successful surgical excision of primary right atrial angiosarcoma". J Cardiothorac Surg. 2011; 6: 47. Published online 2011 April 9. doi: 10.1186/1749-8090-6-47PMCID: PMC3090321.
5. R. R. Brandt, R. Arnold, R. M. Bohle, et al." Cardiac angiosarcoma: case report and review of the literature". Zeitschrift für Kardiologie , Volume 94, Number 12 (2005), 824-828, DOI: 10.1007/s00392-005-0296-0.
6. Mbakise Pula Matebele, Paul Peters, Julie Mundy and Pallav Shah." Cardiac tumors in adults: surgical management and follow-up of 19 patients in an Australian tertiary hospital". Interact CardioVasc Thorac Surg (2010) 10 (6): 892-895. doi: 10.1510/icvts.2009.230029.
7. Sandra Fussen , Bart W.L. De Boeck , Michael J. Zellweger et al. " Cardiovascular magnetic resonance imaging for diagnosis and clinical management of suspected cardiac masses and tumours". Eur Heart J (2011) 32 (12): 1551-1560. doi: 10.1093/eurheartj/ehr104.
8. R. Nitta, Y. Sakomura, K. Tanimoto, et al. "Primary cardiac angiosarcoma of the right atrium undiagnosed by transvenous endocardial tumor biopsy".Internal Medicine, vol. 37, no. 12, pp. 1023–1026, 1998.
9. Burke A, Virmani R. In: Atlas of tumor pathology. Tumors of the heart and great vessels. Rosai J, Sobin LH, editor. Washington DC: Armed Forces Institute of Pathology; 1996. Primary cardiac sarcomas; pp. 127–170. 3rd series, fascicle 16.
10. Nayar S, Nayar PG, Cherian K. "Angiosarcoma presenting as syncope". Asian Cardiovasc Thorac Ann. 2008;16:154–156. [PubMed]
11. Movsas B, Teruya-Feldstein J, Smith J, et al." Primary cardiac sarcoma: a novel treatment approach". Chest. 1998;114(2):648–652. Selected Reports. [PubMed]
12. Baay P, Karwande SV, Kushner JP, et al." Successful treatment of a cardiac angiosarcoma with combined modality therapy". Journal of Heart and Lung Transplantation , 1994;13(5):923–925. [PubMed]
13. Herrmann MA, Shankerman RA, Edwards WD,et al. " Primary cardiac angiosarcoma: a clinicopathologic study of six cases". Journal of Thoracic and Cardiovascular Surgery, 1992;103(4):655–664. [PubMed]
14. Putnam JB, Sweeney MS, Colon R,et al." Primary cardiac sarcomas". Annals of Thoracic Surgery , 1991;51(6):906–910. [PubMed]
15. Gowdamarajan A, Michler RE." Therapy for primary cardiac tumors: is there a role for heart transplantation?" Curr Opin Cardiol. 2000;15:121–125. doi: 10.1097/00001573-200003000-00010. [PubMed] [Cross Ref]
16. Cally K.L. Ho, F.R.C.S., Elaine Wang, D.A.B.Path,W.K. Au, F.R.C.S., and L.C. Cheng, F.R.C.S "Primary Cardiac Angiosarcoma of Left Atrium" 10.1111/j.1540-8191.2009.00872.x(J Card Surg 2009;24:524-525.
17. karldenning,M.D. , Guntherlehmann, M.D. ,thomasrichter, M.D."An Angiosarcoma in the Left Atrium" N Engl J Med 2000; 342:443-444 February 10, 2000.