Case Report
Middle East Journal of Cancer; July 2016; 7(3): 161-164

Primary Right Atrial Sarcoma Presenting with Cardiac Tamponade and Massive Pleural Effusion

Seied Hosein Ahmadi*, Shahram Momtahen*, Fereshteh Ensani, Afsaneh Alikhasi***, Sanambar Sadighi****, Farhad Samiee*****, Zahra Sedighi******, Mohammad Ali Mohagheghi*****♦**

*Department of Cardiac Surgery, Tehran Heart Center, Tehran University of Medical Sciences TUMS, Tehran, I. R. of Iran
**Department of Pathology, Cancer Institute, Imam Khomeini Hospital Center, TUMS, Tehran, I. R. of Iran
***Department of Radiology, Cancer Institute, Imam Khomeini Hospital Center, TUMS, Tehran, I. R. of Iran
**** Department of Hematology Oncology, Cancer Institute, Imam Khomeini Hospital Center, TUMS, Tehran, I. R. of Iran
*****Department of Radiotherapy, Cancer Institute, Imam Khomeini Hospital Center, TUMS, Tehran, I. R. of Iran
******Cancer Research Center, Cancer Institute, Imam Khomeini Hospital Center, TUMS, Tehran, I. R. of Iran

Abstract

Primary cardiac sarcomas are very rare and there is no consensus on management. Clinical presentation is usually late. Despite newer diagnostic technology, prognosis remains dismal. We report a case of right atrial sarcoma in a 28-year-old man who presented with acute cardiac tamponade. Emergency subxiphoid pericardial drainage stabilized the patient's critical condition. The lesion was advanced. Therefore, we only performed a suboptimal surgical resection. Despite planning for radiation, the patient's status deteriorated. Only palliative measures continued during the next four months before his death due to disseminated metastasis and progressive cardiopulmonary failure.

Keywords: Cardiac tumor, Surgical resection, Synovial sarcoma

♦**Corresponding Author:**
Mohammad Ali Mohagheghi, MD
Cancer Research Center, Cancer Institute, Imam Khomeini Hospital Center, Tehran University of Medical Sciences, End Keshavarz Blvd, Tehran, Iran
Tel: +98-21-66581542
Fax: +98-21-66581635
Email: mamohagheghi@gmail.com

Introduction

A total of 25% of cardiac tumors are malignant, among which 75% are sarcomas.[1] Unfavorable prognosis is due to extensive local invasion and distant metastases at presentation.[2] Pericardial effusion is prevalent at presentation in 29% of cases.[2]

Echocardiography is the diagnostic modality of choice. Transthoracic echocardiography depicts tumor characters and attachments, as well as cardiac chambers, septa and great vessels.

Received: November 15, 2015; Accepted: January 11, 2016

Its sensitivity is 93.3%.[3] For detailed assessment, metastatic work-up and completion of diagnosis, both magnetic resonance (MR) and CT scan are useful.[4]

Case Report

A 28 year-old man presented to the cardiac hospital with progressive chest pain, cough and dyspnea of two weeks duration. He experienced severe palpitations and breathlessness on walking. Physical examination at presentation revealed respiratory distress, cyanosis, tachycardia and hypotension. Chest X-ray and echocardiography confirmed subacute cardiac tamponade along with bilateral pleural effusion. The right atrium and right ventricle were collapsed, with normal left ventricular size and function. A non-mobile homogenous left atrial mass attached to the inter-atrial septum (IAS) was detected (Figure 1A).

Emergency sub-xiphoid drainage of 650 mL of serosanguinous fluid resulted in dramatic symptom relief (Figure 1B). The patient underwent a CT scan that confirmed a 6×3 cm right atrial mass with bilateral pleural effusion (Figure 2).

The patient refused surgical intervention and medical support continued during the next four weeks. Due to progressive symptoms and with the patient's informed consent, we scheduled surgical intervention. Complete surgical resection was not possible because of severe adhesions and excessive bleeding. A suboptimal resection of the cardiac tumor and repair of an atrial septal defect with an autologous graft was performed. The patient had an unremarkable post-operative course with good palliation of his symptoms. Histopathlogic (Figures 3 and 4) and immunochemical studies confirmed the diagnosis of synovial sarcoma. This case was discussed at the tumor board which recommended palliative chemoradiotherapy. However, the patient never underwent palliative chemoradiotherapy because of disease progression and poor general condition. Other palliative measures continued during the next four months, after which he died due to local recurrence and distant metastasis that led to cardiopulmonary failure.

Discussion

Primary synovial sarcoma is a rare cardiac malignancy that accounts for less than 1% of all primary cardiac tumors and 5% of cardiac sarcomas.[5,6] The male to female ratio is 2.5:1 and most tumors are located in the right side of the heart.[7]

Biopsy is essential for diagnosis and differentiation between benign and malignant cardiac tumors. Otherwise, a rapid clinical course, mediastinal spread, pericardial hemorrhagic effusion, combined intramural and intracavitary location, extension into the pulmonary and/or caval veins, and distant metastasis are clear sings of malignancy.[8] The current case exhibited many of these signs.

Diagnostic evaluation of cardiac sarcoma

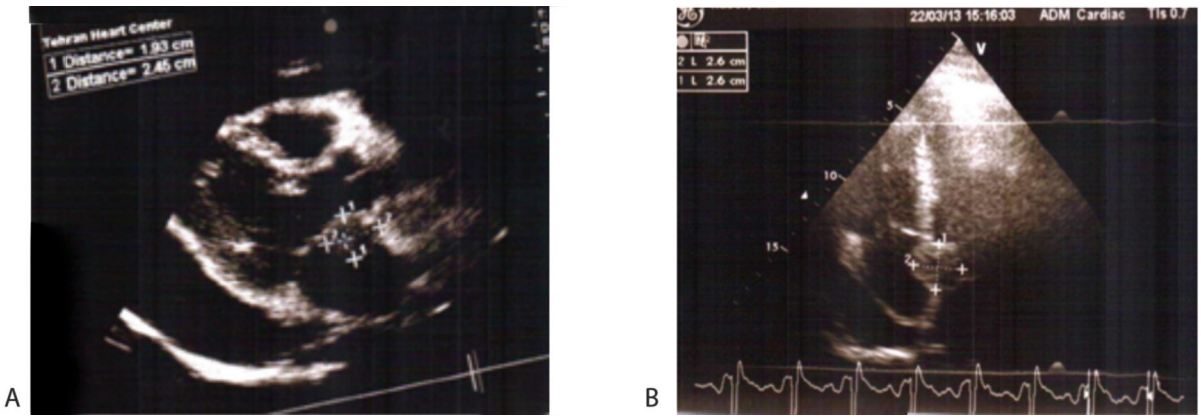

Figure 1. Echocardiogram that shows the presence of an atrial mass attached to the inter-atrial septum (IAS) before (A) and after (B) pericardiocentesis.

begins with chest-X-ray and continues with echocardiography as the investigation of choice. Cardiac MR and CT scan are used to confirm the diagnosis.[4] Transthoracic echocardiography (TTE), as the most effective, reliable, noninvasive, and widely available tool is used to diagnose most cases followed by presurgical transesophageal echocardiography (TEE). Transthoracic echocardiography is an essential part of the diagnostic evaluation of all suspected cardiac sarcomas; TEE is a very powerful diagnostic technique for better visualization of cardiac structures in order to differentiate malignant tumors from benign ones.[9]

Microscopic evaluation is important for definitive diagnosis and consists of architectural, cellular and microvascular patterns, stromal reaction, hemorrhage and necrosis. Histologically, the tumor cells are uniformly spindle-shaped, with an ovoid or oval nucleus. They proliferate in fascicular fashion with features of multiple mitoses. Details of diagnosis and differentiation need higher magnification and immunohistochemical studies.[10]

The presented patient's tumor was highly aggressive. Despite surgical resection and chemoradiation, prognosis is very poor with survival of less than 9 months.[6] Most often, treatment consists of palliative surgical resection followed by chemotherapy or chemoradiotherapy. Future developments such as better elucidation of chromosomal abnormalities and directed therapies will hopefully improve the outcome.[11]

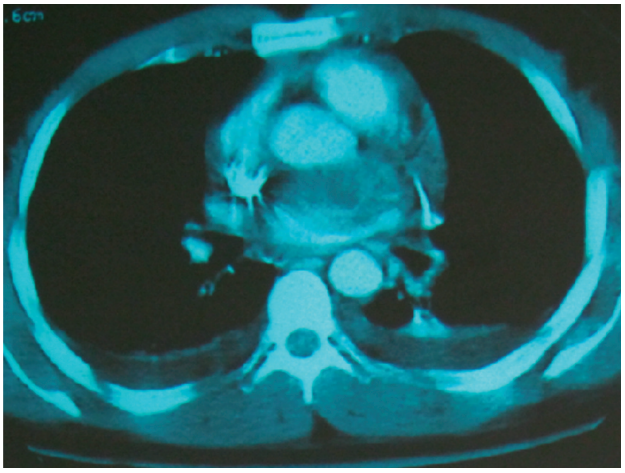

Figure 2. CT scan showing right atrial mass and bilateral pleural effusion.

Acknowledgement

The authors wish to acknowledge the contributions of the following distinguished colleagues Seyed Khalil Foroouzan Nia MD, Abbas Salehiomran MD, Abbas Ghiasi MD, Mahdi Najafi MD, Soheil Mansourian MD, Saeid Davoudi MD, Mahmoud Shirzad MD, Mohammadreza Rezaei MD, Kyomars Abbasi MD, and Hassan Soltaninia MD. We would like to express our appreciation to Ms. Maryam Sotudeh Anvari, Associate Professor of Clinical

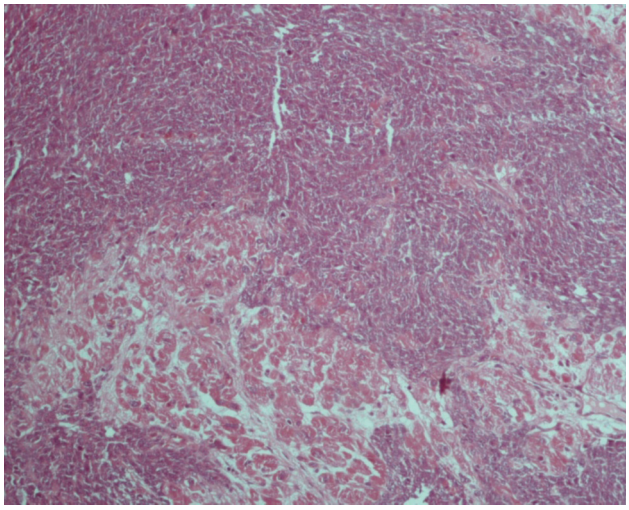

Figure 3. Spindle cell sarcoma with a more prominent storiform pattern, focal pale cytoplasm, mild nuclear atypia, and myocardial involvement by the tumor cells (H & E, 10×).

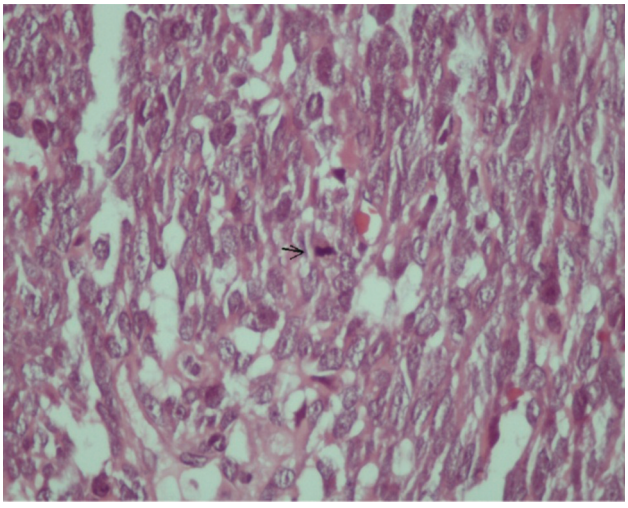

Figure 4. High-power view that shows moderate pleomorphism in plump spindled cells and mitotic figures (arrow) (H & E, 40×).

and Surgical Pathology, for her technical assistance with preparing the pathology slides.

Conflict of Interest

No conflict of interest is declared.

References

1. Devbhandari MP, Meraj S, Jones MT, Kadir I, Bridgewater B. Primary cardiac sarcoma: reports of two cases and a review of current literature. *J Cardiothorac Surg.* 2007;2:34.
2. Shanmugam G. Primary cardiac sarcoma. *Eur J Cardiothorac Surg.* 2006;29(6):925-32.
3. Meng Q, Lai H, Lima J, Tong W, Qian Y, Lai S. Echocardiographic and pathologic characteristics of primary cardiac tumors: a study of 149 cases. *Int J Cardiol.* 2002;84(1):69-75.
4. Benassi F, Maiorana A, Melandri F, Stefanelli G. A case of primary cardiac angiosarcoma: extensive right atrial wall reconstruction with autologous pericardium. *J Card Surg.* 2010;25(3):282-4.
5. Miller DV, Deb A, Edwards WD, Zehr KJ, Oliveira AM. Primary synovial sarcoma of the mitral valve. *Cardiovasc Pathol.* 2005;14(6):331-3.
6. Talukder M, Joyce L, Marks R, Kaplan K. Primary cardiac synovial sarcoma. *Interact Cardio Vasc Thorac Surg.* 2010;11(4): 490-2.
7. de Zwaan C, Bekkers SC, van Garsse LA, Jansen RL, van Suylen RJ. Primary monophasic mediastinal, cardiac and pericardial synovial sarcoma: a young man in distress. *Neth Heart J.* 2007;15(6):226-8.
8. Devbhandari MP, Meraj S, Jones MT, Kadir I, Bridgewater B. Primary cardiac sarcoma: Reports of two cases and a review of current literature. *J Cardiothorac Surg.* 2007;2:34.
9. Hsieh PL, Lee D, Chiou KR, Kung MH, Lin SL, Liu CP, et al. Echocardiographic features of primary cardiac sarcoma. *Echocardiography.* 2002;19(3):215-20.
10. Orlandi A, Ferlosio A, Roselli M, Chiariello L, Spagnoli LG. Cardiac sarcomas: an update. *J Thorac Oncol.* 2010;5(9):1483-9.
11. Zhang L, Qian J, Li Z, Jing H. Primary synovial sarcoma of the heart. *Cardiol J.* 2011;18(2):128-33.