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


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

**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 interatrial 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. Histopathologic (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. The male to female ratio is 2.5:1 and most tumors are located in the right side of the heart. 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. The current case exhibited many of these signs.

Diagnostic evaluation of cardiac sarcoma

![Figure 1](image1.png) 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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{11}

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

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×).
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Conflict of Interest
No conflict of interest is declared.

References