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Cardiac sarcoma presenting as tamponade

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CASE REPORT

Cardiac Sarcoma Presenting as Tamponade

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Abstract

Sarcomas are a broad group of neoplasm that originate from the mesenchymal layer and represent about 1% of malignancy in the adult population. We describe a 46-year-old woman with no prior medical history who presented with worsening dyspnea. Physical examination was significant for jugular venous distension and bilateral lower extremity edema. Chest Xray showed cardiomegaly. Echocardiography showed a normal ejection fraction of 65%, pericardial effusion with tamponade physiology and three epicardial masses. Patient had pericardiocentesis performed followed by pericardial window. Biopsy of pericardial mass showed morphologic and immunophenotypic findings supportive of diagnosis of malignant soft tissue sarcoma. Though this type of cancer is rare, malignancy should be included as one of the differential diagnoses of new-onset pericardial effusion in a young patient. Early diagnosis and referral to a specialized sarcoma center for treatment is recommended.

Keywords: Cardiac sarcoma, Sarcoma, Small round blue cell tumor, Cardiac tamponade, Pericardial effusion, Cardiomegaly, Leg swelling

1. Introduction

Sarcomas are a broad group of neoplasms that originate from the mesenchymal layer. They represent about 20% of malignancies seen in the pediatric population compared to 1% in the adult one.[1] Common locations include the bone, muscles, cartilage, joints, and nerves. Sarcomas arising from the heart are rare and have been associated with poor prognosis. Primary cardiac tumors have been reported in approximately 0.001−0.3% of autopsies.[2] About 90% are benign and 10% are malignant.[3] Of the malignant tumors, about 75% are sarcomas.[2] Primary cardiac malignant tumors usually occur in young patients with a median age of 44 years, and it is equally distributed between genders.[4] Based on histology, the most common types of cardiac sarcomas are angiosarcomas, undifferentiated sarcomas, and undifferentiated pleomorphic sarcomas.[5] These malignant tumors are rapidly progressive and produce early death through infiltration of the myocardium, obstruction of circulation, or distant metastases to lungs, lymph nodes, and liver. We present a 46-year-old female who presented with progressive dyspnea and leg edema. Cardiac tamponade was present and managed accordingly. Cardiac sarcoma was later diagnosed based on further imaging and biopsy. The patient was then transferred to a specialized sarcoma center for further management.

2. Case

A 46-year-old woman with no prior medical history apart from recent diagnosis for hypertension presented to the emergency department on account of progressive dyspnea on exertion for three months and bilateral leg edema. Significant physical examination findings included elevated jugular venous distension (JVD) and bilateral lower extremity edema. Laboratory findings showed normal brain natriuretic peptide (BNP) −15 pg/ml (normal range-

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<100 pg/ml), elevated alanine aminotransferase (ALT) - 180 U/L (normal range- 0–61 U/L), aspartate aminotransferase (AST) - 110 U/L (normal range - 5–34 U/L), normal thyroid stimulating hormone (TSH) - 3.1 uIU/ml (normal range- 0.35 - 4.94 uIU/ml). Acute hepatitis serologies were negative, HIV serologies/P24 antigen ELISA, and autoimmune markers including antinuclear antibodies (ANA) and rheumatoid factor (RF) were negative. Chest Xray (CXR) showed marked cardiomegaly suspicious for pericardial effusion (Fig. 1). Computed tomography (CT) scan of the chest, abdomen, and pelvis were remarkable for large pericardial effusion, trace ascites, and incidental finding of a 5.3 cm hepatic cyst. Transthoracic echocardiography (TTE) showed normal left ventricular function (ejection fraction – 65%) and large echo-free pericardial effusion up to 4 cm with associated right ventricular and right atrial diastolic collapse suggestive of tamponade physiology. The inferior vena cava was dilated with minimal respiratory variation. Additionally, 3 round masses were identified (largest 5.3 × 3 cm) and were attached to the epicardium within the pericardial base (Fig. 2). Pericardiocentesis was performed under fluoroscopy guidance which resulted in drainage of 1500 ml of bloody fluid and pericardial drain placement. Fluid analysis showed nucleated cell count of 357 × 10E6/L and red blood cell (RBC) –161,000 × 10E6/L. Cytology as well as routine culture, fungal, viral, acid-fast bacilli, and anaerobic culture were negative. Flow cytometry of the pericardial fluid showed no evidence of non-Hodgkin's lymphoma or high-grade myeloma neoplasm. Following removal of the drain, re-accumulation of fluid was seen on repeat TTE within 24 h. Mini thoracotomy with pericardial window and biopsy of a large encapsulated epicardial mass with necrotic center at the Right Atrium/Left Atrium groove were performed. Intraoperative findings showed large mass attached to the epicardium at the interatrial groove adherent to the right superior pulmonary vein, right atrial wall, and left atrial wall. Intraoperative frozen section was consistent with malignancy - small blue cell tumor, no gross tumor seeding, or evidence of tumor nodules in the lung or pleura. Biopsy showed morphologic and immunophenotypic findings supporting a diagnosis of a malignant small round blue cell tumor including monophasic synovial sarcoma, desmoplastic small round cell tumor, or Ewing sarcoma. Numerous stains including neuroendocrine, vascular, muscle, lymphoid, and other epithelial markers were negative. Flow cytometric analysis performed by Integrated Oncology (AFT21-011,370) showed 3% of total events to have scatter characteristics of lymphocytes, 89% of which were CD3-positive T-cells. The T-cells had a CD4-to-CD8 ratio of 0.6 and demonstrated retained expression of CD2, CD5, and CD7. There were insufficient B-cells for clonality studies. Fluorescence in situ hybridization (FISH)

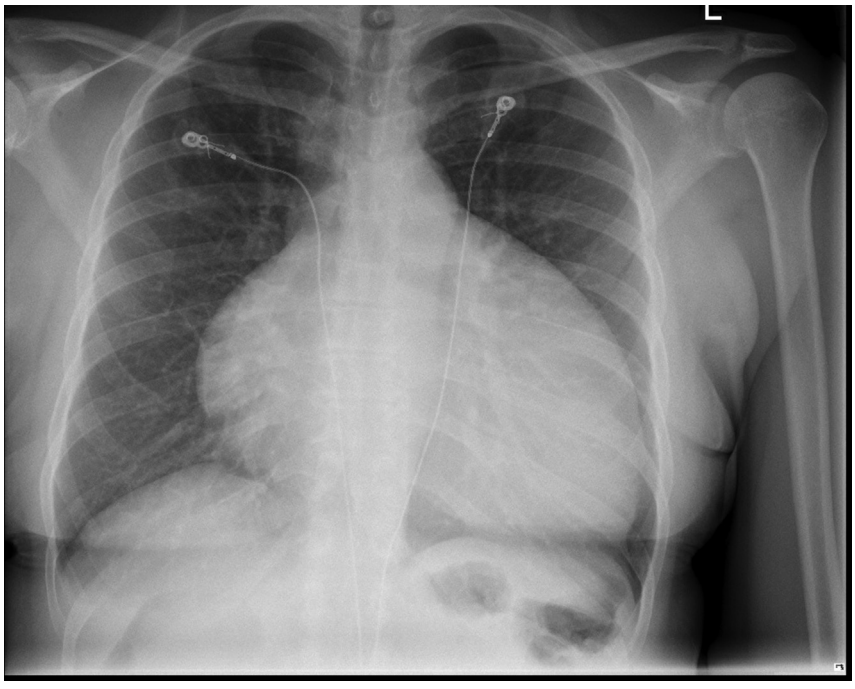

Fig. 1. Chest Xray showing markedly enlarged heart.

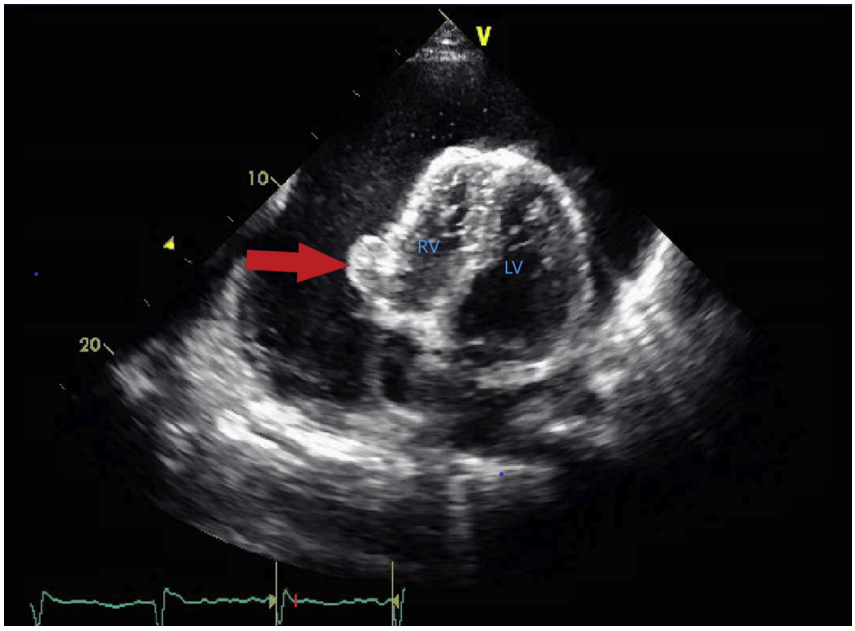

Fig. 2. Echocardiography finding showing epicardial mass.

studies were negative for SS18 (SYT) (18q11) gene arrangement and EWSR1 (22q12) gene arrangement. Patient was then transferred to a sarcoma center for further management where chemotherapy was initiated. One year later, patient has completed two rounds of chemotherapy and now on immunotherapy. Restaging scans with cardiac magnetic resonance imaging (MRI) showed slightly decreased cardiac masses and CT imaging were negative for distant metastases.

3. Discussion

Cardiac sarcomas though rare can be aggressive. Therefore, early diagnosis and treatment is important for overall survival. Patients usually present with similar symptoms suggestive of an underlying heart disease. The common manifestations that have been reported include dyspnea (48%), chest pain (22%), heart failure (13%), and pericarditis (5%).[6]

Initial workup for common causes of dyspnea includes a CXR which in our patient showed a globular heart concerning for pericardial effusion, then confirmed by TTE. Pericardial effusion and cardiac tamponade are common findings that have been previously reported in patients diagnosed with cardiac sarcoma.[7,8] TTE is the first diagnostic imaging to evaluate the size, site of origin, mobility, and hemodynamic effects of the tumor.[3] In patients with cardiac tamponade, the initial procedure of choice is pericardiocentesis. In patients with recurrent effusion, a surgical pericardial window is usually needed.[9] A contrast TTE can also be useful to differentiate vascular tumors from thrombi. Most malignant tumors are heavily vascularized and show significant contrast enhancement while benign tumors i.e., myxomas show less contrast enhancement. Thrombi show complete absence of contrast opacification being avascular structures.[10] CT can provide useful information on site of origin, anatomical relationships, surrounding structures involvement and extracardiac localization.[11]

Cardiac MRI remains the best available noninvasive diagnostic tool to provide information about morphology, dimensions, location, extension, perfusion, and tissue characterization of the mass.[11] Positron Emission Tomography (PET) scan using 18F-fluoro-D-glucose (FDG) in combination with CT (or MRI) can also be used to assess cardiac masses, differentiating benign and malignant tumors by the extent of FDG uptake by tumors.[12] Further CT abdomen and pelvis is recommended given the high likelihood of extra-peritoneal metastasis to the abdomen and retroperitoneum especially in round cell/myxoid liposarcomas.

Management of patients with cardiac sarcomas may vary according to the stage at diagnosis. Primary cardiac sarcomas are best classified by the site of origin. which is further directly linked to the surgical approach used for resection.[13] In early stage, surgical removal is suitable although feasible in only in less than half of patients as most have evidence of metastasis at the time of presentation or most tumors recur rapidly, limiting survival to

approximately one year.[14,15] Cytoreductive chemotherapy with or without radiotherapy are other therapeutic options.[16] First line chemotherapy would include Adriamycin, whereas second line gemcitabine plus docetaxel would be used. Nevertheless, the impact of chemotherapies on the natural history of cardiac sarcomas is limited, and hence the importance of a multidisciplinary evaluation to define the best therapeutic approach based on the experience of the group.[13]

In conclusion, primary cardiac neoplasms should be included in the differential diagnosis of new onset pericardial effusion in a younger patient. Though the prognosis of primary cardiac sarcoma is poor, early diagnosis, surgical resection, and chemotherapy may prolong survival.

Conflicts of interest

All authors have no conflict of interest.

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