

Right Arterial Leiomyosarcoma: A Very Rare Cardiac Tumor

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Introduction

Cardiac neoplastic involvement is not common. The incidence of primary cardiac tumor is 0.02% [1] and among all cardiac tumors, only 25% are malignant [2,3]. This unique case of Leiomyosarcoma presents a very rare presentation of primary cardiac tumor. Leiomyosarcoma is a type of malignant sarcoma, which is a neoplasm of smooth muscles because; they contain interlacing fascicles of spindle cell resembling smooth muscle. Approximately 20 different groups of sarcoma are recognized on the basis of the pattern of differentiation toward normal tissue with cardiac Leiomyosarcoma is the rarest of all forms of cardiac malignant tumors. Cardiac sarcoma has higher tendency in the female gender with an incidence of two times as that in males [4,5]. The most common sites are usually left atrium approximately 75% of all cases. Right atrium (16%) and the ventricles (7%) are considered the least occurrences sites [6].

Case Presentation

This was a 67-year-old African American male who was admitted to the hospital with increasing Shortness of Breath (SOB), Paroxysmal Nocturnal Dyspnea (PND), orthopnea and inability to lay flat. He stated that he had been sleeping in a reclining chair for a couple of years; however, for the last 7 weeks prior to admission, his condition had significantly deteriorated. Social history revealed that he smoke one pack of cigarettes per day (1pk/d) but quit 15 years ago. Family history revealed coronary artery disease (CAD), prostate cancer and diabetes mellitus. His past medical history indicated hypertension, diabetic mellitus, gastroesophageal reflux, hyperlipidemia, gastrointestinal bleeding, along with seizure disorder and chronic obesity (Table 1).

Past surgical history indicates prosectomy and no other operations. Physical examination: revealed a mesomorphic male with complaint of shortness of breath (SOB), chest tightness with edematous upper and lower extremities. Echocardiogram on admission indicated a large right atrial mass with moderate to severe pulmonary hypertension, moderately dilated Right Atrium (RA) along with hypertrophy of Right Ventricle (RV) and left ventricular. Subsequently, Patient underwent cardiac catheterization showing moderate coronary artery disease (CAD). Transesophageal Echocardiogram (TEE), as well as a CT scan of the chest confirmed a large right atrium (RA) mass. The mass appeared to be attached to the lateral wall with no gross attachment to the intra-arterial septum by TEE. No metastasis or other left ventricular abnormality was found. Following an informed consent, patient underwent exploration of the mediastinum and the heart via median sternotomy. A large poly-lobulated right atrium tumor was found measuring 8x6 cm (gross pathology images Figure 1a and 1b) starting at the proximal Superior Vena Cava (SVC) and extending to

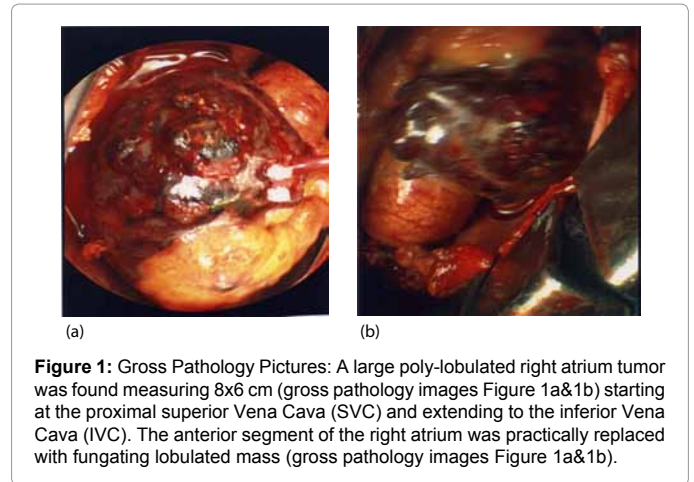


Figure 1: Gross Pathology Pictures: A large poly-lobulated right atrium tumor was found measuring 8x6 cm (gross pathology images Figure 1a&1b) starting at the proximal superior Vena Cava (SVC) and extending to the inferior Vena Cava (IVC). The anterior segment of the right atrium was practically replaced with fungating lobulated mass (gross pathology images Figure 1a&1b).

the Inferior Vena Cava (IVC). The anterior segment of the right atrium was practically replaced with fungating lobulated mass (gross pathology images Figure 1a and 1b). Intraoperative wedge biopsy of this mass was obtained. Since the report of the frozen section was inconclusive, the chest was closed awaiting final pathology report. The final biopsy report was not conclusive for Leiomyosarcoma or Hemangiosarcoma. Subsequently, patient underwent re-exploration and an 8 x 6 cm hard mass, including the anterior right atrium, was removed. The mass covered the entire lateral portion of the right atrium but not including the tricuspid valve. A pericardial patch was measured to fit the resected section of the right atrium. Then the patch was anastomosed to the base of the resected right atrium from superior vena cava to inferior vena cava along with reconstructing and reforming a new anterior right atrium wall. Patient also had an A-V sequential pacemaker inserted. He did well after his extensive surgery and subsequently, was sent home. Patient was referred to the cancer center and a 6-month post-operative workup with CT scan and PET scan reported liver metastasis only. Patient underwent successful chemotherapy and was tumor free during his one-year follow up.

Histopathology Presentation and Diagnosis

Leiomyosarcoma is malignant tumor with smooth muscle differentiation. It is an uncommon type of cardiac malignancy which adds up approximately 8% - 9% of all sarcomas [7]. At gross examination, Leiomyosarcoma most commonly appears as gelatinous,

Past Medical History

- Hypertension (HTN)
- Diabetes Mellitus (DM)
- Gastroesophageal reflux (GERD)
- Obesity
- Hyperlipidemia
- Seizure disorders
- GI bleed

Table 1: Past Medical History of 67-year-old African American male.

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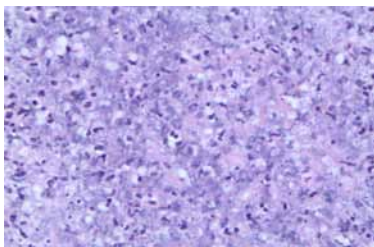
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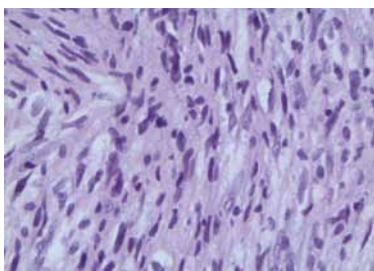
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sessile masses, other occasion may present in multiple approximately 30% of cases [7]. Cardiac sarcomas presentation are of broad range of cell differentiation [8], but Leiomyosarcoma are characterized by prominent cellular atypia, abundant mitosis and areas of coagulative cell necrosis; which is referred to as Stanford Criteria [9].

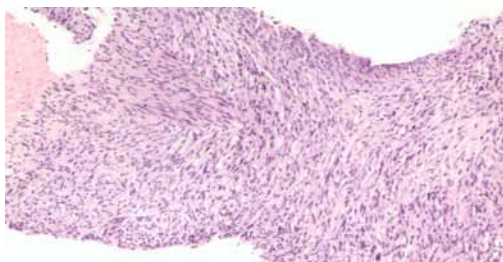
Histologically Leiomyosarcoma consist of malignant smooth muscle cell contain bundles of thin filament with dense bodies and pinocytic vesicles [10]. Initial histopathology examination demonstrated hypercellular specimen with cigar-shaped spindle cells commonly arranged in a herringbone pattern along with marked nuclear atypia and frequent mitosis (Slides 1 and 2). Adjacent tumors necrosis presented in (Slide 3) is indicative of malignant nature of Leiomyosarcoma. Individual cell are surrounded by basal lamina. Immunohistologically, they stain with antibodies to smooth muscle actin and desmin [10]. Immunohistologically the constituent cells were positive with smooth muscle actin and desmin. Cells of smooth muscle origin stain brown with smooth muscle actin stain. This stain is a specific stain for smooth muscle, thus giving the current specimen a diagnosis of Leiomyosarcoma (Slide 4). The Vimentin is one of the five major intermediate filaments found in cells. Cells of mesenchymal



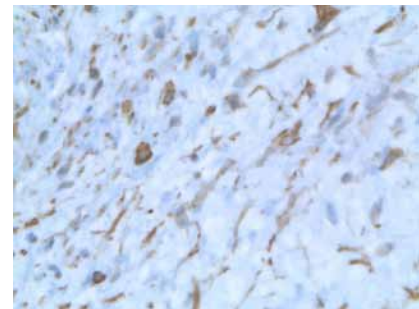
Slide 1: Hypercellular specimen with cigar-shaped spindle cells arranged in a herringbone pattern in a background of increased vascluarity.



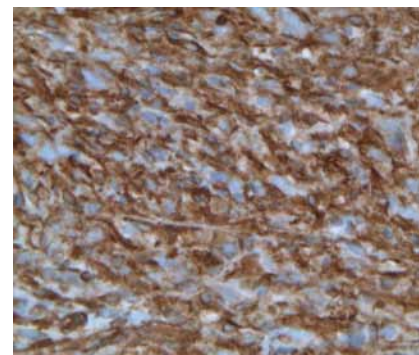
Slide 2: High-power view of the spindle cells. Cells are hyperchromatic with marked nuclear atypia and frequent mitosis are present.



Slide 3: Leiomyosarcoma with necrosis: adjacent tumors necrosis, indicative of malignant nature of Leiomyosarcoma.



Slide 4: Smooth muscle actin stain: Cells of smooth muscle origin stain brown with smooth muscle actin stain. This stain is a specific stain for smooth muscle, thus giving the current specimen a diagnosis of: Leiomyosarcoma.



Slide 5: Vimentin stain: cells of mesenchymal origin stain brown with Vimentin. This is a sensitive stain for any mesenchymal-derived soft tissue tumors.

origin stain brown with Vimentin (Slide 5). This is a sensitive stain for any mesenchymal-derived soft tissue tumors.

1. Leiomyosarcoma low power: Hypercellular specimen with cigar-shaped spindle cells arranged in a herringbone pattern in a background of increased vascluarity.
2. Leiomyosarcoma high power: High-power view of the spindle cells. Cells are hyperchromatic with marked nuclear atypia and frequent mitosis is present.
3. Leiomyosarcoma with necrosis, high power: Adjacent tumors necrosis, indicative of malignant nature of Leiomyosarcoma.
4. Smooth muscle actin stain: Cells of smooth muscle origin stain brown with smooth muscle actin stain. This stain is a specific stain for smooth muscle, thus giving the current specimen a diagnosis of: Leiomyosarcoma
5. Vimentin stain: Cells of mesenchymal origin stain brown with Vimentin. This is a sensitive stain for any mesenchymal-derived soft tissue tumors.

Conclusions

Cardiac tumors are usually manifesting itself in either primary or secondary presentation. They frequently can arise from myocardium or the pericardium. Other incidences such as by direct extensions of primary tumors or metastasis from adjacent structures are also conceivable. The first diagnosis of a cardiac tumor by clinical examination and electrocardiography was in 1934. Prior to this date,

cardiac tumors were only identified post-mortem and Intra-cardiac cavitory tumor was first demonstrated by angiography in 1951. In general, the origins of primary cardiac tumors are either mesothelial or epithelial which include following: myxoma, fibroma, sarcoma, thymoma, lymphoma, angioma hemangioma, hemangiopericytoma, angiomylipathy, angiomyo hamartoma, lymphangiomas, and mycosis fungoid [10-20].

Echocardiography is initially used to evaluate intra-cardiac tumors. Transesophageal echocardiography (TEE) is used to differentiate LA masses from thrombi. MRI and MRA (magnetic resonance angiography) and Computed Topography (CT) are preferred to better asses contiguous extra-cardiac involvement on the metastatic disease and more complex cases [11,21].

Primary cardiac tumors are rare findings with an incidence of 0.02% and have dismal prognosis. Primary right atrium leiomyosarcoma is amongst the rarest of all malignant cardiac tumors. The average duration from onset of symptoms until final diagnosis is shorter for malignant cardiac sarcoma (approximately 4 months) than benign myxoma with 15 months [20,22].

Treatment for primary cardiac leiomyosarcoma requires rapid action and comprehensive approach. Essential surgical resection followed by adjuvant radiation therapy and/or chemotherapy are fundamental of treatment [5,23,24]. Final short term outcome is promising but long term prognosis for these tumors is not. The prognosis for these tumors is very poor and most patients die within one year of diagnosis [3,6] therefore, immediate and complete surgical resection of tumor along with adjuvant therapy provides the best hope for palliation and longer survival outcome [5,23]. Heart transplantation for patients with primary malignant tumors has not offered convincing results [6,24,25].

The patient in our report has been followed for duration of two years who represent the longest surviving patient with cardiac leiomyosarcoma.

In summary, we have presented a rare case of right atrial leiomyosarcoma with successful radical surgical resection followed by adjuvant treatment as described.

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