

An Unfortunate Case of Poorly Differentiated Spindle Cell Sarcoma Involving the Heart

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Introduction

Soft tissue sarcomas are malignant tumors of connective tissue that arise from mesenchymal tissue. They can arise at any body site and are extremely rare, representing less than 1% of all newly diagnosed malignancies. A small subset of these rare tumors originate as primary cardiac malignancies.

Here we describe an unusual case in which a patient's rapidly deteriorating clinical course is explained by one such tumor, only elucidated after his unfortunate demise.

Case Description

History & Physical Exam

- 70 year old male presented with failure to thrive, weakness, decreased appetite, and weight loss.
- Past medical history of diastolic heart failure, atrial fibrillation, recurrent pericardial effusions s/p pericardial drain, and pulmonary sarcoidosis.
- Pericardiocentesis 1 month prior to presentation was negative for malignant cells.
- Vitals normal except for low blood pressure 90/71.

Labs & Imaging

- Labs demonstrated elevated lactate, troponin, creatinine, and liver enzymes which up-trended.
- Echocardiogram revealed severe left ventricular hypertrophy with increased wall thickness, severely reduced right ventricular systolic function, and a complex pericardial exudate without conclusive evidence of tamponade.
- CT chest (Figure 1) demonstrated a circumferential pericardial effusion with areas of loculation and multiple epicardial nodules.

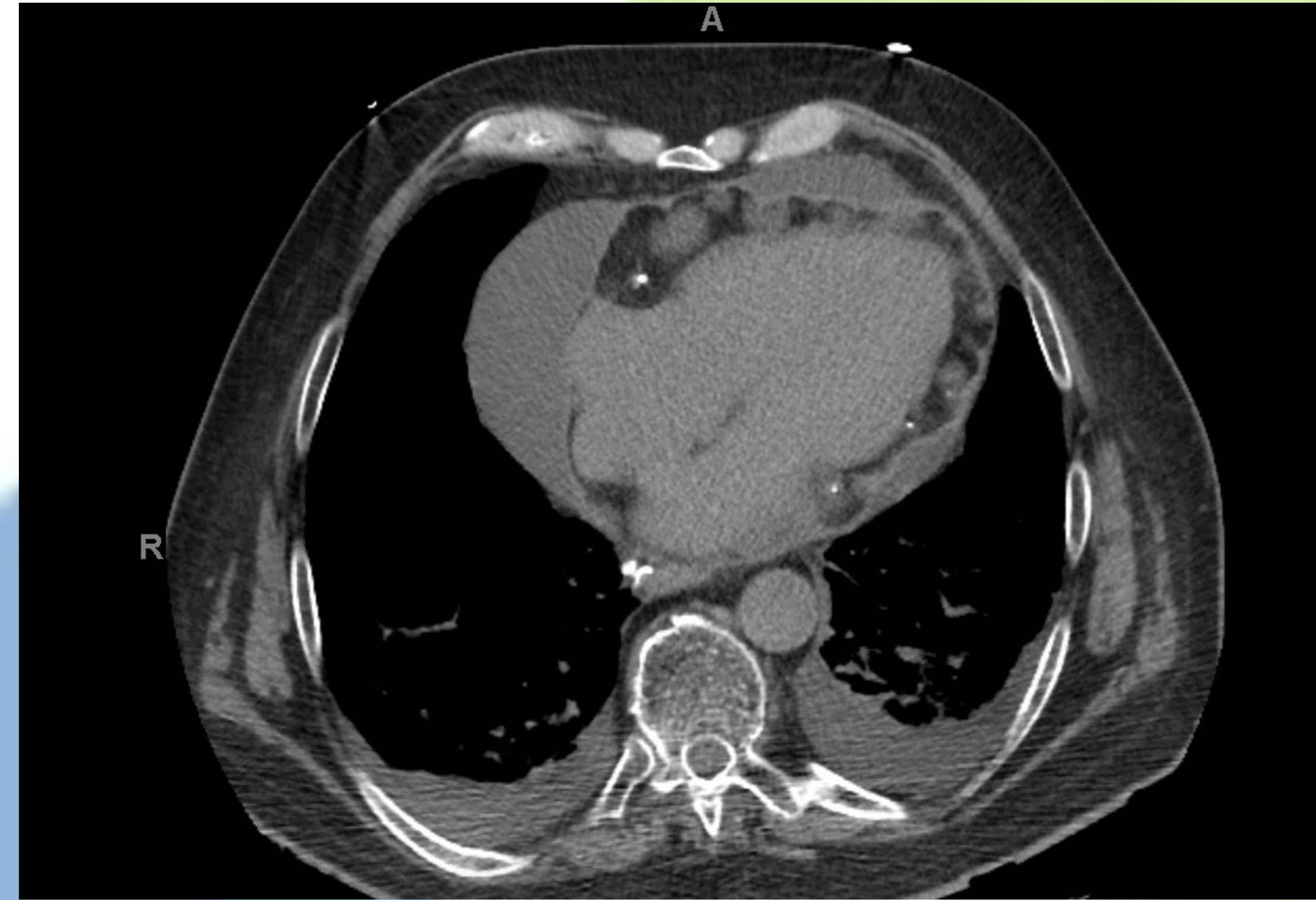


Figure 1: CT Chest demonstrating loculated pericardial effusion with pericardial nodules.

Clinical Course

- There was concern for a low-flow state and cardiac tamponade, but echocardiography was inconclusive for a definitive diagnosis of tamponade.
- Due to worsening labs and hypotension, patient was taken for surgical evacuation as shock was felt to be driven by pericardial constriction.
- Intraoperative transesophageal echocardiogram demonstrated biventricular failure with focal areas of tamponade physiology caused by constrictive pericardial thickening which was restricting the heart's ability to fill.
- Emergency sternotomy revealed a thick fibrin material that had fibrosed and coated the epicardium along the entire anterior heart from the great vessels to the diaphragm.
- Given the extent of infiltrative disease, the decision was made to close the patient up and transition to comfort measures as there were no medical or surgical therapies that could have led to his survival. The patient died shortly thereafter.
- Biopsy results of the fibrin material returned with poorly differentiated malignant spindle cell neoplasm, sarcoma vs sarcomatoid carcinoma.

Discussion

Malignant involvement of the pericardium is detected in 1-20% of cancer cases in autopsy studies. However, primary cardiac tumors are exceedingly rare, with metastatic involvement of the heart over 20 times more common.

Patients generally present due to symptoms related to compression secondary to tumor enlargement. Tumors readily proliferate and infiltrate the myocardium which usually causes death due to obstruction of blood flow through the heart. Fibrosarcomas and undifferentiated sarcomas are composed of spindle cells which can more readily infiltrate myocardium.

Our patient had severe left ventricular hypertrophy which may have represented this extensive myocardial infiltration. The involvement of the pericardium led to focal areas of tamponade, making echocardiographic diagnosis extremely difficult.

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