Mayaguez Medical Center
Family Medicine Residency Program
It was not Takotsubo

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INTRODUCTION

Soft tissue sarcomas are rare mesenchymal tumors, accounting for only 1% of all cancers. These tumors can develop anywhere in the body, with the most common locations being the extremities and abdomen. Mediastinal soft tissue sarcomas are especially rare, making up less than 10% of primary mediastinal tumors and less than 1% of all soft tissue sarcomas. Unlike synovial sarcomas found in the extremities, mediastinal tumors more frequently occur in male patients, with the average age of diagnosis being 39 years old.

The presenting symptoms can vary widely, but the most common ones include chest or shoulder pain, shortness of breath, cough, and pericardial effusion. On chest radiographs, the tumors may appear as well-circumscribed neoplasms with sharply defined borders or as ill-defined infiltrative lesions. Computed tomography may reveal large tumor masses with homogeneous or heterogeneous enhancement.

On immunohistochemistry, diffuse expression of bcl-2 is typically seen. In 60% of cases, these tumors stain for CD99. It's important to note that these markers are not specific to synovial sarcoma and are also expressed by a wide range of unrelated neoplasms. Overexpression of the Transducer-like enhancer of split 1 (TLE1) has been observed in synovial sarcomas. It is a relatively sensitive and specific immunohistochemical marker for these tumors, with sensitivity rates ranging from 82% to 100%.

Wide surgical excision is the primary treatment for addressing the tumor, and adjuvant radiation is used for larger and deeper lesions. The role of chemotherapy in treatment is still a topic of debate, and additional research is suggested. Factors such as increasing size, age, and tumor grade have been shown to negatively impact the likelihood of local disease recurrence and metastasis. Patients typically have a poor prognosis, with a five-year overall survival rate of only 14.8%.

CASE PRESENTATION

A 33-year-old male was sent to the emergency department after experiencing chest pain for the past 2 weeks following his father's death. He also presented with peripheral edema, a weight loss of 12 pounds, and a decreased appetite. The patient attributes his symptoms to "gastritis and stress" and denies experiencing fever, chills, night sweats, or palpitations.

Past medical history: intestinal obstruction at 2 year old
Allergies: bee stings (anaphylaxis)
Medications: None
Social: denies toxic habits (smoking, drinking, or drug use)
Family history: no mention of cancer

Physical examination:
- GEN: Alert, active, oriented x 3, no acute distress
- HEENT: PERRLA, EOMI, MOM, poor oral hygiene
- HEART: regular rhythm but tachycardic. S1/S2 heard, rub sound appreciated
- LUNGS: bi-basilar crackles bilaterally
- ABD: + BS in all quadrants, soft, depressible, non-tender. Well-healed RLQ surgical scar
- EXT: pitting edema +3 up to knee bilaterally with serous effusions
- NEURO: no focal deficit, CN II XII grossly intact

ADMISSION RESULTS

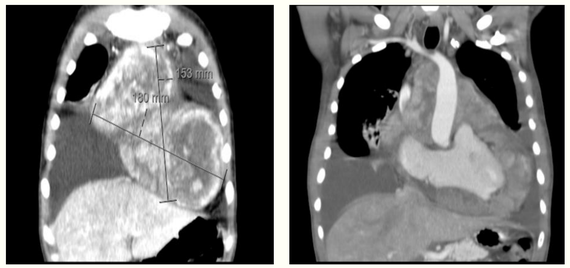
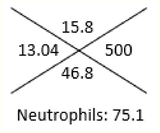
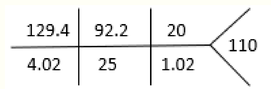


Fig 1. Admission Chest CT showing large infiltrative mass with varying degrees of enhancement

Admission Chest CT: large infiltrative mass in the pericardial space extending into the superior pericardial recess and mediastinum. The mass in the upper mediastinum measures approximately 15 x 10.5 cm, with a more inferior pericardial component measuring up to 15 cm in craniocaudal by 18 cm in transverse by 7.2 cm in anteroposterior. This mass is exerting pressure on both the right and left ventricles. The mass shows varying degrees of enhancement, with some areas appearing more enhanced and others less enhanced. Additionally, there are bilateral pulmonary artery-filling defects in the descending branches extending into the segmental branches.





Bilirubin: elevated total bilirubin at 3.6, direct bilirubin at 1.9 and indirect bilirubin at 1.7
Lactate dehydrogenase: elevated at 132, BNP elevated at 640.9

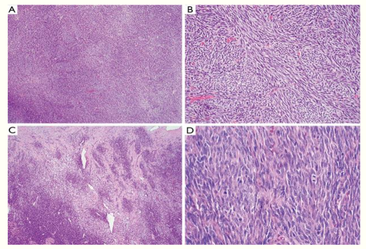


Fig 2. Histopathology of monophasic synovial sarcoma. (A) Low power view of a neoplasm composed of sheets of monomorphic spindle cells; (B) tumor cells arranged in fascicles; (C) alternating hypo and hypercellular areas; (D) mitotic activity increased.

Pathology: Monophasic fibrous synovial sarcoma
- BCL2: strong diffuse positive
- Caldesmom: negative
- CD34: negative
- CD 99: strong diffuse positive
- CK AE1/AE3: negative
- EMA: negative
- Ki 67: increased proliferation (10%)
- Myogenin: negative
- S-100: negative
- TLE-1: strong diffuse positive
- VImentin: strong diffuse positive

CASE PRESENTATION CONTINUUM

During admission, the patient was diagnosed with bilateral pulmonary emboli and bilateral pleural effusions. The pulmonary embolism was treated with anticoagulation. A thoracentesis was performed to remove more than 1,200 mL of dark yellow pleural fluid. Pathology results for the pleural fluid were negative for malignancy.

The patient underwent an incisional biopsy for a mediastinal mass the day after admission. During the biopsy, he developed cardiogenic shock, likely due to the compression effect of the mass. Cardiogenic shock was treated with vasopressors until the blood pressure stabilized to a mean arterial pressure above 65 without the need for vasopressors. Chest tubes were placed bilaterally during the biopsy due to recurrent effusion. Pneumothorax developed, which can be a complication of the procedure but oxygen supplementation was sufficient to improve the pneumothorax.

Although not yet conclusive, the effect of the chemotherapies on synovial sarcoma, being a young patient with no relevant past medical history, prompted the placement of a medport for the possibility of radiation and chemotherapy following discharge.

CONCLUSION

It is imperative to conduct a thorough history and physical examination in cases involving chest pain. Prompt referral and treatment are essential for addressing any alarming symptoms. The patient exhibited chest pain, and elevated heart rate for a duration of 2 weeks. He also experiences decreased appetite, a weight loss of 12 pounds, and peripheral edema over 6 months.

Although sarcomas in the mediastinum are uncommon, they can manifest a diverse range of clinical symptoms based on the affected area and structures. In this instance, the specific type of sarcoma, monophasic fibrous synovial sarcoma, affected the pulmonary artery and its branches, thereby increasing the likelihood of pulmonary emboli. Additionally, the sarcoma exerted pressure on the pericardium, elevating the risk of cardiogenic shock.

The diagnosis of monophasic fibrous synovial sarcoma necessitates immunochemistry testing, which may yield positive results for general markers such as BCL2, CD99, and vimentin. Notably, the most specific marker, TLE-1, was assessed and demonstrated positivity in this case.

Clinical studies recommend tumor resection as the optimal approach, followed by radiation therapy. The role of chemotherapy in treatment is still under investigation. After diagnosis, the patient was scheduled to commence treatment with radiation and chemotherapy. However, surgical resection had to be postponed due to the development of cardiogenic shock during the preceding surgery.

LIMITATIONS

This is a case report that limits the generalization of findings.

Radiation and chemotherapy were not administered during hospitalization due to delays related to the medical insurance process.

As this case report was conducted retrospectively, the patient or family members were not contacted for follow-up. It is important to acknowledge that potential complications may have emerged after the patient's discharge.

REFERENCES