INTRODUCTION

- Sarcomas: a rare and heterogeneous group of malignant tumors of mesenchymal origin that comprise <1% of all adult malignancies

- Primary Cardiac Angiosarcomas: Autopsy prevalence of 0.0001-0.02%

- Physicians are 30-50x more likely to find metastatic tumors to the heart than to find a primary cardiac neoplasm itself
A previously healthy 50-year old male presents to emergency department (ED) with 5 week hx of worsening shortness of breath, intermittent chest pain, malaise, and fatigue.
<table>
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<tr>
<th>HISTORY OF PRESENT ILLNESS</th>
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<tr>
<td>▪ Patient HK is a 50-year-old male with a PMH significant for sinus tachycardia who presents to the emergency department primarily complaining of increasing shortness of breath and intermittent chest pain.</td>
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<td>▪ Fatigue + gradually increasing shortness of breath and intermittent retrosternal chest pain occurring 2-3x/week. Sharp in nature.</td>
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<td>▪ Worse with leaning forward. No other specific maneuvers exacerbate the pain. Did not occur specifically with inspiration or expiration.</td>
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<td>▪ Pain comes on quickly, lasts a second or two, then resolves.</td>
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<td>▪ Denies N/V, dizziness, or palpitations though he has noticed increased sweating both day and night, which he attributed to the particularly hot August weather.</td>
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<td>▪ Recent travel hx: within the last few weeks he had flown to upstate New York on a 6-hour flight and then went on a 5-mile hike while in New York without any problems.</td>
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<td>▪ Over the past week, he had traveled to a conference 3 hours away and complains that he could not walk more than a few feet before he needed to sit down and rest due to shortness of breath.</td>
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<td>▪ Denies joint swelling, lymph node swelling, pedal edema, leg pain, palpitations, orthopnea, paroxysmal nocturnal dyspnea, dizziness or wheezing. He endorsed poor appetite, weight loss, new-onset generalized pruritus, and a wet cough productive of white phlegm, no hemoptyis.</td>
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<td>PAST MEDICAL HISTORY</td>
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<td>▪ Sinus tachycardia</td>
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<td>▪ OSA; uses CPAP at home</td>
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<td>▪ GERD</td>
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<td>▪ Insomnia</td>
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<th>FAMILY HISTORY</th>
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<td>▪ Father: deceased, end-stage CKD + hx of DM</td>
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<td>▪ Mother: deceased, metastatic lung cancer</td>
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<td>▪ Brother: deceased age 53, colon cancer</td>
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<td>▪ Sister: still living, hx malignant melanoma</td>
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SOCIAL HISTORY

- Monogamous with wife x25 years
- Former police detective; investigator for the county DA
- Frequent contact with prisoners at various prisons, spends 6-8 hours with them at a time. Last known contact was a few weeks prior. He reports doing this multiple times per month
- Remote hx tobacco use: smoked 1/2ppd x5 years, quit smoking 30 years ago
- Alcohol: 1-2 drinks per month

ALLERGIES

- NKDA

MEDICATIONS

- Atenolol 25mg PO Q HS for sinus tachycardia
- Omeprazole 20mg PO Q HS for GERD
- Zolpidem 10mg PO Q HS PRN for insomnia
PHYSICAL EXAM

- **VITALS:** 114/78, 113, 20, 98.8F, 93% on room air
- **GENERAL:** Pleasant overweight gentleman, appeared to be in increasing pain as the exam proceeded
- **HEENT:** PERRLA, sclera non-icteric. Nares clear. Neck supple. No cervical or supraclavicular lymphadenopathy.
- **CARDIAC:** Tachycardic S1S2 no S3S4, no murmurs, rubs, gallops.
- **LUNGS:** CTAB, no rhonchi, wheezes, rales. Dullness to percussion bilaterally.
- **ABDOMEN:** Distended, but non-tender. No rebound or guarding. No hepatosplenomegaly. Negative peritoneal sign.
- **SKIN:** Mild area of erythema over upper chest, blanching, cool to touch.
- **MSK:** Chest wall non-tender to palpation. Extremities non-edematous.

LABORATORY AND DIAGNOSTIC DATA

- **EKG:** Sinus Tachycardia, no evidence of electrical alternans or ST changes
- Negative Troponin x2
- Negative chest x-ray
- **Hgb/Hct, platelets, sodium, potassium, chloride, bicarb, BUN, Cr, Glu, LFTs, TSH, and INR** were all normal within healthy limits
- **WBC:** 13.6
  ESR: 79, CRP: 163
- Elevated D-dimer > 2000
- Blood smear showed toxic granules
**IMAGING AND PROCEDURES**

- Given travel hx, underwent CT angiography of the chest with IV contrast
  - Results: large pericardial effusion with possible cardiac tamponade
  - CT-guided drainage of pericardial effusion: 350mL serosanguinous fluid drained with immediate relief to patient
  - Post-drainage TEE showed trivial effusion and mild LVH

**CYTOLOGY AND CULTURE**

- Basic studies of the pericardial effusion resulted in negative cytology to rule out underlying malignancy and negative culture for aerobes, anaerobes, and acid-fast bacilli
  - Tuberculin skin testing performed in the hospital was negative.
  - Mononucleosis screen was negative.
  - Prostate specific antigen was normal.
  - Recent colonoscopy within the past several months was also normal.
  - No skin lesions suggestive of melanoma.
- HK was ultimately discharged without a clear etiology...
SOMETHING WASN’T RIGHT...

- 4 weeks after diagnosis of acute pericarditis, HK returns to the emergency department...
- Extreme fatigue to the point of being bedridden
- Rapid continued weight loss: 20lbs in 4 weeks.
- Denies headache, upper respiratory symptoms, palpitations, abdominal pain, constipation or diarrhea, or documented fever.
- Vitals: BP 128/73, HR 98, RR 20, 99.7F.
- WBC: 14.5, H/H: 12.4/37.2
- Bedside Echo: small anterior effusion, no tamponade.

- Pericardial drainage didn’t go as expected....

WHAT IS GOING ON?
**MAGNETIC RESONANCE IMAGING**

*Figure-01.* MRI showed an enlarged heart with thickened irregular pericardium. There are multiple locules of complex pericardial fluid within intervening thick enhancing separations.

**PERICARDIAL WINDOW**

*Figure-02.* Pericardial window technique. A short vertical incision is made over the xiphoid, extending onto the midline of the abdomen. The retrosternal space is entered and the pericardium is sharply incised, allowing fluid to be aspirated or drained using a suction catheter.
THE DISEASE PROGRESSES...

Figure-03. CT scan showed rapidly progressive pericardial nodularity and likely complex effusion compared to the initial study from six weeks prior, with extrinsic compression of the cardiac chambers.

DISCUSSION: PRIMARY CARDIAC ANGIOSARCOMA

- Exceedingly rare tumors with an autopsy prevalence of 0.0001-0.02%\(^1\)
- Men younger than 65 years of age are predominantly affected with a male to female ratio of 2:1\(^2\)
- Most common sxs: dyspnea, chest pain, palpitations, fever, myalgia, sxs that mimic right-sided heart failure
- The insidious disease typically progresses silently until tumor burden induces hemodynamic instability or causes systemic deformity\(^3\)
PRIMARY CARDIAC ANGIOSARCOMA

- Pericardial effusion is the first manifestation of disease in almost all cases
- Literature on fifteen known cases of cardiac angiosarcoma reported that although pericardiocentesis frequently yielded bloody or necrotic tissues, cytology results were negative for malignancy in all, even with direct tumor invasion into the pericardium\textsuperscript{4, 5}.
- In many cases of the disease, pericardiocentesis fails and diagnosis is only made after exploratory thoracotomy\textsuperscript{6}.
- 80% of patients have metastatic disease by the time they are properly diagnosed\textsuperscript{7}.
- The lungs are the primary site of metastasis followed by spread to the liver, lymph nodes, bone, adrenals, spleen, and brain\textsuperscript{8}.

TREATMENT MODALITIES

- Up to this point, all treatments have been anecdotal:
  - Complete vs. Incomplete resection
  - Chemotherapy/Radiotherapy
  - Heart transplantation
OSTEOPATHIC CONSIDERATIONS

- Acupuncture, mediation, yoga, and osteopathic manipulative therapy can reduce pain, relieve chemotherapy-induced nausea, and alleviate peripheral neuropathy while also promoting immune function and an overall sense of wellbeing\(^9\).

- Osteopathic medicine as a form palliative therapy for cancer patients involves a gentle approach in which techniques such as myofascial release, muscle energy, and lymphatic pump are utilized to help control pain and tension, relieve cancer-related lymphedema, and improve the patient’s overall feeling of health\(^10\).

IN CONCLUSION

- Due to lack of research, there has been little to no progress establishing standardized diagnostic or treatment algorithms for this disease.

- 80% of patients already have distant metastasis at the time of diagnosis, yet our best tool to prolong patient lifespan is complete surgical resection which requires no metastasis in order to be successful.

- Until we are able to harness a more successful treatment modality, the best we can do for our patients is improve the time of diagnosis in an effort to catch the disease before metastasis has occurred. This is what will give our patient’s the best fighting chance.
ACKNOWLEDGMENTS

WE WOULD LIKE TO THANK BOTH DR. TRACY MIDDLETON AND DR. ANETTE GAWELKO FOR THEIR FEEDBACK OF OUR ORAL PRESENTATION SKILLS AND FOR THEIR CONTINUED GUIDANCE DURING PREPARATION OF THIS CASE. WE WOULD ALSO LIKE TO THANK DR. WILLIAM PEPPO FOR HIS FEEDBACK DURING THE EARLY STAGES OF WRITE-UP AND DR. KEVIN CONSIDINE OF CROWN ISLAND FAMILY PRACTICE FOR ALLOWING US TO USE THIS CASE.

REFERENCES


REFERENCES


Fast and Fatal:

A RARE CASE OF PRIMARY CARDIAC ANGIOSARCOMA

KALEY CAPITANO OMS-III
MIDWESTERN UNIVERSITY, ARIZONA COLLEGE OF OSTEOPATHIC MEDICINE