## Introduction

- Cutaneous angiosarcoma is an aggressive, rare malignancy arising from the vascular epithelium.
- Incidence of sarcoma overall is 7.1 per 100,000 persons with cutaneous angiosarcoma making up 5% of all sarcomas.
- For patients with no cancer history, 83% of cutaneous angiosarcoma occur in the head and neck scalp, whereas 51% of cases occur on the trunk for patients with previous cancer diagnosis.
- Risk factors include past radiation exposure, chronic lymphedema, and exposure to foreign bodies such as Dacron and other graft materials.
- Cancers requiring radiation treatment may increase risk for cutaneous angiosarcoma development with breast cancer being the most common.
- The phenomenon of cutaneous angiosarcoma in areas affected by chronic lymphedema was first observed and described by Stewart and Treves in 1948 with postmastectomy patients.
- Stewart-Treves Syndrome is still described today as cutaneous angiosarcoma arising from tissues that have been chronically lymphedematous.
- MTVc gene amplification has been found in many patients with radiation-associated cutaneous angiosarcoma as well as other angiosarcomas.
- Figure 1 depicts the prognostic staging of soft tissue sarcomas that is used for cutaneous angiosarcoma.

### Table: Prognostic Staging of Soft Tissue Sarcoma

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<thead>
<tr>
<th>Category</th>
<th>N</th>
<th>M</th>
<th>Stage</th>
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<tbody>
<tr>
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<td>M0</td>
<td>IA</td>
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<tr>
<td>T2, T3, T4</td>
<td>NO</td>
<td>M0</td>
<td>IB</td>
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<tr>
<td>T5</td>
<td>NO</td>
<td>M0</td>
<td>IIIA</td>
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<tr>
<td>T1M0</td>
<td>NO</td>
<td>M0</td>
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<td>N</td>
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<td>M</td>
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**Staging:** 0 = No regional lymph nodes. 1+ = Regional lymph nodes involved. 5 = Distant metastasis.

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<tr>
<th><strong>T</strong> category</th>
<th><strong>N</strong> stage</th>
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<tbody>
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<tr>
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**Score of histologic grade refers to total differentiation, mitotic count and necrosis.

### Figure 1: Prognostic Staging of Soft Tissue Sarcoma

#### Case Description

**History**

- **Vital Signs:**
  - **BP:** 94/57 mmHg
  - **HR:** 88 beats per minute
  - **SpO2:** 97%
  - **Temp:** 37.0°C
  - **Wt:** 134 kg
- **General:** Acute distress noted, appeared to be uncomfortable and in pain on exam.

**PMH:**

- Chronic lymphoma of right lower extremity, relapsed cutaneous angiosarcoma, type 2 diabetes mellitus, chronic obstructive pulmonary disease, Barrett’s esophagus, hypertension, obstructive sleep apnea, gastroesophageal reflux disease, depression, sleep apnea, thymoma.

**PMHx:**

- Right above-knee amputation in 2020 after cutaneous angiosarcoma diagnosis in distal right lower extremity.

**Allergies:**

- No known drug allergies.

**FHx:**

- Leukemia and colon cancer in unknown family member at early age.
- No history of sarcoma or breast cancer.

**SHx:**

- Tobacco: quit 17 years ago, approx 20 pack-year history.
- No current ETOH or recreational drug use.

**Home Medications:**

- Ibrutinib 150mg twice daily
- Ferrous sulfate 325mg twice daily
- Pantoprazole 40mg once daily
- Propanolol 150mg twice daily
- Enalapril 2.5mg once daily
- Metoprolol 50mg twice daily
- Oxycodone 5mg 3 times daily
- Ativan 0.5mg once daily
- Allopurinol 300mg once daily
- Ativan 0.25mg once daily
- Oxycodone 7.5mg 3 times daily
- Allopurinol 300mg once daily
- Ativan 0.25mg twice daily
- Oxycodone 7.5mg 3 times daily

**Treatment/Outcome**

- **Plan:**
  - Rapid decline after hospitalization as described in Figure 2 with hypotension, sepsis, and findings similar to Figures 4 and 5.
  - Hypotension managed with IV midodrine 75mg twice daily as well as norepinephrine and vasopressin during rapid response.
  - Sepsis treated with IV vancomycin 750mg every 12 hours and IV levofloxacin 750mg every 24 hours.
  - Multidisciplinary consults obtained to aid in complex care.

**Conclusion**

- **Prognosis:**
  - Local recurrence of cutaneous angiosarcoma is approximately 57% with ulceration and bleeding at affected sites occurring frequently.
  - Five-year survival rate of cutaneous angiosarcoma is 12-33%, which is lower than other angiosarcomas.
  - Surgical treatment is typically indicated for cutaneous angiosarcoma followed by radiotherapy.
  - For cases that are inoperable or refractory to surgical resection, chemotherapy can be used.
  - Docetaxel, paclitaxel, and doxorubicin-based chemotherapy have shown success in some with cutaneous angiosarcoma.
  - Palliative embolization was suggested in this case, which could be used in a variety of cancers to mitigate negative outcomes with refractory, uncontrolled bleeding.

### Figure 2: Hospital Course

#### Days 1-2
- **Initial visit to ED for 24 hour observation.
- Second emergency department visit with BP of 94/57.
- Was provided 2 units of blood and stabilized before being admitted.

#### Days 3
- **Rapid response called for hypotension with systolic BP of 66, believed to be septic shock.
- Admitted to the ICU.
- Chemotherapy treatment for angiosarcoma initiated.
- Wound debridement due to active bleed in RLE.

#### Days 4
- **Hypotension:**
  - Man continued to improve.
  - Re-evaluated for septic shock.
  - Blood transfusions provided and Hct decreased.
  - Rapid response called again for hypotension.

#### Days 5-9
- **Levofloxacin and vancomycin provided throughout for UTI and possible cellulitis of RLE.
- Blood transfusions provided.

#### Days 10-11
- **Patient became comfort measures only and died on hospital day 11, secondary to septic shock.

### References