Case Report

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Stewart-Treves Syndrome: An Interesting Case of Angiosarcoma after Radical Mastectomy

Cosmin Florescu[1], MD, MA, MPH and Christine Ahn[2], MD
[1]Department of Emergency Medicine, Eisenhower Medical Center, Rancho Mirage, California, USA
[2]Department of Emergency Medicine, Stony Brook University Hospital, Stony Brook, New York, USA

Abstract

Introduction: This article discusses Stewart Treves Syndrome, a rare form of angiosarcoma that occurs with chronic lymphedema after radical mastectomy with lymph node dissection. STS presents with purplish cutaneous nodules or reddish blue macules that enlarge. The mortality rate is high and survival is low.

Case Report: A 68 year old female with history of breast cancer status post radical mastectomy with lymph node dissection, radiation and chemotherapy six years prior was evaluated in the Emergency Department for bleeding from her left upper arm lesion. The patient required resuscitation and was subsequently admitted to the ICU. The following day, she suffered a stroke and, after discussion with her family, she was placed on comfort care measures. She died three days after admission.

Conclusion: Emergency physicians should be aware of this rare, but highly lethal, malignant syndrome. A patient with history of breast cancer and radiation therapy who presents with an enlarging red-purple plaque should be admitted or referred to oncology or plastic surgery for immediate biopsy and discussion with family and palliative care specialists regarding goals of care.

Key words: Stewart Treves Syndrome, angiosarcoma, lymphagiosarcoma, chronic lymphedema

Introduction

This article discusses Stewart Treves Syndrome (STS), a rare disease that presents with purplish cutaneous nodules or reddish blue macules that enlarge. The mortality rate is high and survival is low. Background research does not reveal any articles within the Emergency Medicine literature regarding STS. The purpose of this case report is to introduce for emergency medicine physicians the highly lethal form of angiosarcoma that can arise in patients after radical mastectomy with lymph node dissection or those with chronic lymphedema.

Case Report

A 68 year old female with history of hypertension, hyperlipidemia, left MCA aneurysm diagnosed one month before, obesity, left breast cancer status post modified axillary lymph node dissection, radiation and chemotherapy six years prior who presented to the Emergency Department for evaluation of bleeding from her left upper extremity lesion. She reported burning pain and bleeding from the left arm lesion, fatigue, lightheadedness, and shortness of breath. Two months prior to presentation, the patient injured her arm while cleaning a litter box and noticed a small bruise that had enlarged since then. Nine days prior, she was evaluated by plastic surgery and had biopsy of the lesion, which showed atypical vascular neoplasm consistent with angiosarcoma.

In ED triage, patient was hypotensive to 86/57 and sent to the Critical Care area for evaluation. The remaining vital signs were Tmax 36.8, HR 102, RR 16, and O2 saturation 96% on room air. On physical exam, the patient had a 12cm x 8cm x 4cm red violaceous lesion on the left upper arm. She was resting comfortably in bed in no acute distress.

Initial ED labs were significant for lactic acid 2.5, BNP 11201, ESR 43, CRP 5.9, potassium 3.4, glucose 262, Mg 1.5, Ca 8.1, troponin 0.38, and anion gap 20. Hemoglobin decreased from 12.9 two months prior to 8.9. Chest x-ray showed pulmonary vascular congestion and left pleural effusion. CT pulmonary angiogram was negative for pulmonary embolism with extensively calcified, but normal caliber, aorta. CT chest showed a 2.3cm x 1.7cm x 2.7cm enhancing fluid collection within the left anterior chest wall and diffuse skin thickening of the left breast.

Despite 2 liters of IV fluids, the patient was persistently hypotensive with SBP in the 70s. The ED team placed a midline catheter and arterial line, and the patient was subsequently started on norepinephrine and vasopressin with im-

Corresponding Author: Cosmin Florescu **e-mail:** sbu.florescu@gmail.com
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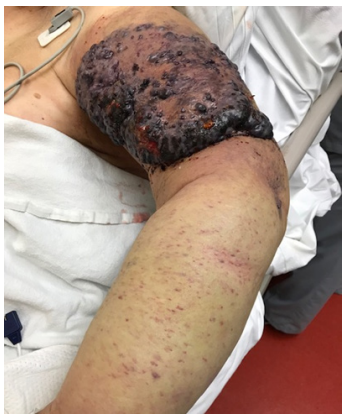

Figure 1a: Patient's lesion: note new petechiae on forearm

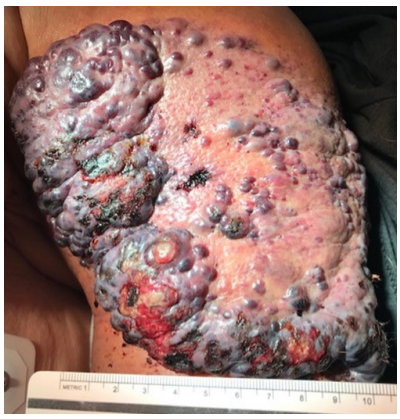

Figure 1b: Patient's lesion: width of 12cm: note draining and bleeding areas

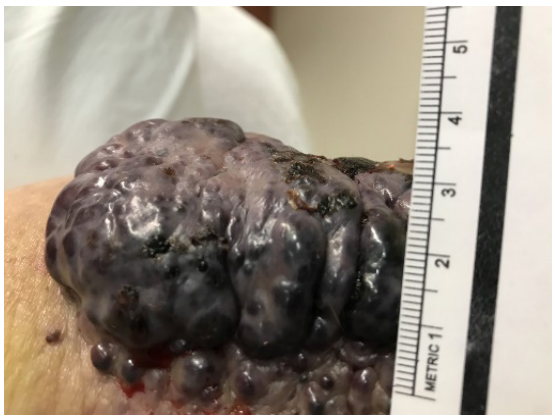

Figure 1c: Patient's lesion: height of 4cm: note dark purple blue characteristics

provement in SBP and MAP. Additionally, over the course of three hours, the patient's left forearm started developing petechiae and she became febrile. Antibiotic coverage with cefepime and vancomycin was started due to concern for septic shock. Plastic surgery and ICU were consulted and the patient was admitted to the ICU for further management.

After admission to the ICU, our patient's troponins started to increase, so cardiology was consulted. Transthoracic echocardiogram showed EF 22% (previous TTE six years prior to presentation showed EF 64%). The following day, the patient suffered an acute stroke, confirmed on CT and MRI, and the patient required intubation. Neurosurgery evaluated the patient and reviewed imaging, which showed the left MCA aneurysm had enlarged over the past month. However, due to patient's clinical condition, worsening multi-organ system failure, and poor prognosis in the setting of STS, the patient's family elected to pursue palliative comfort measures. The patient died three days after presenting to the ED and admission to the ICU.

Discussion

Stewart-Treves Syndrome is lymphangiosarcoma seen in patients after breast cancer treatment with radiation and lymph-node dissection. The eponymous syndrome was first described by Drs. Stewart and Treves in 1948 in a case series of six patients[1,2]. There are approximately 400 cases reported worldwide in the literature[3]. Lymphangiosarcoma is one of the rarest and most aggressive forms of soft tissue neoplasms[4]. It is therefore important for physicians to consider this syndrome in patients with a history of breast cancer with radiation and lymph-node dissection who present with limb edema.

The incidence of STS in patients who survive more than 5 years after radical mastectomy with axillary node dissection is between 0.07% to 0.45%[2]. The mean time of onset is approximately 10-11 years after radical mastectomy. The mean age of presentation is 60 years and the highest incidence occurs in patients between 50 and 70 years old[5]. After diagnosis, survival is low, typically 8-15 months[5].

The pathophysiology of STS is complex, and research has elucidated some of the process by which a patient can develop angiosarcoma after lymphedema. Sarcomas of the soft tissue account for less than 1% of all cancers, and angiosarcoma is a subset that is particularly aggressive[2] with a high rate of local recurrence and potential for metastasis[6]. Angiosarcoma includes malignant sarcomas originating from either lymphatic (lymphangiosarcoma) or capillary endothelium (hemangiosarcoma), although the clinical differences have not been formalized due to the rarity of both tumors. The cutaneous sub-type accounts for 50-60% of cases[2]. The etiologies of cutaneous angiosarcoma are idiopathic, post radiation treatment, and chronic lymphedema after mastectomy (also known as STS)[7]. The exact mechanism by which Stewart-Treves syndrome arises is currently unclear, although research has suggested systemic carcinogenic factors and neoplastic transportation that arises in edematous tissue as collateral circulation develops after radiation therapy[2].

There are multiple risk factors for the development of STS. The most important cause of STS is congenital or acquired chronic lymphedema[8]. While the majority of angiosarcomas are idiopathic, notable risk factors include exposure to ionizing radiation and chronic lymphedema[2]. In approximately 90% of cases, the angiosarcoma is associated with lymphedema after mastectomy[2]. Additional risk factors include chronic infections, chronic venous stasis, morbid obesity, malignant obstruction, surgical procedures that disrupt lymphatic flow, and hereditary lymphatic malformations such as Noonan syndrome and Milroy disease[2].

There are no pathognomonic physical exam findings for STS. There are some common characteristics that emergency medicine physicians should consider. Cutaneous angio-

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sarcoma appears as a "spreading bruise" with subsequent edema, ulceration, and hemorrhage[2]. The lesions of angiosarcoma can be purplish cutaneous nodules or reddish blue macules that enlarge and coalesce[8,6]. Lesions are typically 3-6cm, although if left untreated, they can grow to 20cm or larger and begin to drain[2]. The most common site of metastasis is the lung, and patients may present with pleural effusion, pneumothorax, or pleural disease, although the cancer can also spread to the liver, bone, soft tissue, and lymph nodes[2].

The differential diagnosis includes hemangioma, hemangioblastoma, squamous cell carcinoma, Kaposi sarcoma, anaplastic melanoma, cutaneous telangiectatic metastatic breast disease[2], and venous ulcer of the extremity[7]. Kaposi sarcoma is difficult to distinguish from STS[8]. Kaposi sarcoma is positive for human herpesvirus 8 (HHV-8) and does not have lymphedema as much as Stewart Treves syndrome does[8].

Since STS is so rare, there is no standard treatment[8]. Emergency medicine management of STS has not been described in the literature. As is the case with an undifferentiated patient with a skin lesion, the emergency physician should obtain vital signs and begin resuscitation efforts as needed. A set of labs should include a CBC, BMP, LFTs, inflammatory markers such as lactic acid, ESR, and CRP. An HIV test is warranted if status is unknown. Imaging should be obtained in consultation with specialists, typically hematology-oncology and/or plastic surgery. Evaluation with CT or MRI helps to evaluate the tissue, including lymph node involvement[2]. The mortality rate for STS is high, and as such, most patients will require close observation in a telemetry unit or an intensive care unit. Our patient required hemodynamic support with vasopressors. Placing a central line or midline, if available, and arterial line are prudent measures in a hypotensive patient suspected of STS. The health care team should engage the patient and family early to discuss goals of care if the diagnosis of STS is already known. It is important to consult specialists early, as evaluation requires immediate biopsy. Inpatient management includes surgery, radiation therapy, and chemotherapy[2-4].

Prognosis of STS is grim, with poor long-term survival. The median survival is 7-19 months[2] and mean survival is 19-31 months[8]. The 3 year survival rate is 55% and by 5-years, survival decreases to 8.5-13.6%[2,3]. Between 20-45% of patients have metastatic disease when they present to a health care provider[2]. Survival outcomes are significantly worse with high grade tumors and tumor size greater than 5cm[9]. Adverse predictors of survival include metastasis at presentation, visceral/deep soft tissue location, tumor size greater than 5cm, tumor necrosis, and absence of surgical excision[10]. Good prognostic factors include age less than 50, localized tumor stage, and tumor located on the trunk rather than the extremities[2].

Conclusion

Emergency physicians should be aware of Stewart Treves Syndrome, a rare, but highly lethal, syndrome. A patient with a history of breast cancer and radiation therapy who presents with an enlarging red-purple plaque should be admitted or referred for rapid follow up with surgical oncology or plastic surgery for immediate biopsy and early discussion with the patient regarding goals of care.

Conflict of Interest

The authors declared no conflict of interest.

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Author Contributions

Concept – CF, CA; Design – CF, CA; Supervision – CF, CA; Resource – CF, CA; Materials – CF, CA; Data Collection and/or Processing – CF, CA; Analysis and/or Interpretation – CF, CA; Literature Search – CF, CA; Writing – CF, CA; Critical Reviews – CF, CA

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