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Case Report

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A Case Report of Stewart Treves Syndrome: A Rare Presentation

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ABSTRACT

We report the case of a 78-year-old female with a history of radical mastectomy for breast cancer in 2011, complicated by chronic left upper extremity (LUE) lymphedema. In September 2021, she was referred to plastic surgery for evaluation of a progressively enlarging, violaceous lesion on the LUE. Initially presumed to be a slow-healing hematoma, the lesion continued to enlarge, prompting a skin biopsy three months later that confirmed a diagnosis of cutaneous angiosarcoma.

The patient declined amputation but underwent multiple local resections, chemotherapy, and radiation therapy. In June 2024, nearly two years after her initial diagnosis, she presented to the emergency department with signs of a soft tissue infection of the LUE. Imaging and laboratory workup revealed no evidence of metastatic spread. During her hospitalization, she developed worsening infection, suffered multiple strokes, and became septic. After a transition to comfort care, she passed away the same day.

Angiosarcoma, a rare and aggressive subtype of soft tissue sarcoma (STS), is associated with poor prognosis and a high rate of early metastasis, with 20–40% of patients presenting with metastatic disease at diagnosis. Median survival is approximately 2.5 years with intervention, compared to 5–8 months with palliative care alone. Surgical resection remains the gold standard for localized disease. This case underscores the importance of early recognition and biopsy of suspicious lesions, particularly in patients with chronic lymphedema, to facilitate timely diagnosis and potentially improve outcomes in STS.

Keywords: Stewart Treves Syndrome, Angiosarcoma, Metastases, Lymphedema

Introduction

STS is a rare form of cutaneous angiosarcoma that arises in areas of chronic lymphedema. The disease was first described in 1948 by Drs. Fred Stewart and Norman Treves, after observing six female patients that developed angiosarcoma following radical mastectomy [1]. Although this syndrome is known to arise as a consequence of mastectomy, it can also occur in the setting of chronic infection, chronic venous stasis, morbid obesity, and malignant obstruction [2]. The classic presentation consists of enlarging purple papules arising in lymphedematous areas that eventually infiltrate tissue and lead to edema, ulcerations, infections and hemorrhage [3].

We report a case of a 78-year-old female who was diagnosed with angiosarcoma ten years after radical mastectomy for breast cancer complicated by chronic lymphedema consistent with STS.

Case Report

The patient was a 78-year-old, Caucasian female with an extensive oncologic history including colonic carcinoid tumor resected in 2002, ER+, PR+, HER stage 2 carcinoma of the left breast for which she underwent breast lumpectomy, adjuvant chemotherapy and radiation complicated by lymphedema that was managed with a lymphedema sleeve. Her oncologic history is further notable for a duodenal neuroendocrine tumor with regional nodal metastases diagnosed in 2019, endometrial adenocarcinoma of the uterus diagnosed in 2020 followed by intrauterine device placement and cessation of hormone therapy for breast cancer, and intraductal papillary mucinous neoplasm of the pancreas. Despite her multiple cancer diagnoses, germline testing did not find a cancer predisposition syndrome. Other medical problems included obesity, glaucoma, obstructive sleep apnea, hypothyroidism, and hypertension. Notably, the patient also had a history of prior taxane exposure with persistent neuropathy.

In September 2022, the patient presented to her primary care physician for subcutaneous lumps with color change on her left

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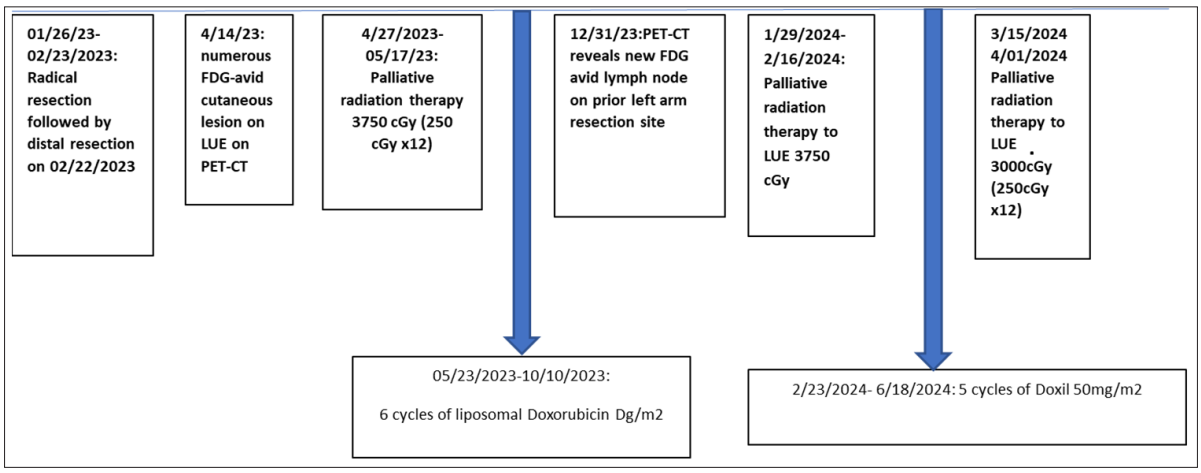
upper extremity for the past three months, prompting a referral to plastic surgery for potential biopsy. She saw plastic surgery in October 2022, who observed "bruising/hardening of the superficial subcutaneous tissue [on the posterior arm] without underlying palpable mass or fluid" and diagnosed a slow-healing bruise. In November 2022, the patient visited her hematologist with concerns that the lesion was now painful and larger than 10 centimeters. Given no history of easy bruising and an adequate platelet count (110,000) with normal INR, there was low suspicion for coagulopathy. A punch biopsy of the left upper arm, below the area of the lesion, was performed and revealed normal skin (see Figure 1). An MRI was then ordered by her oncologist, with notable findings including nodular and mass-like skin thickening at the dorsal ulnar aspect of the upper arm. By December, the lesion had continued to progress, so an excisional biopsy of the lesion was obtained, with pathology confirming high-grade angiosarcoma involving deep and peripheral margins with vascular invasion. The patient underwent two local resections in 2022 and 2023, completed eleven cycles of liposomal doxorubicin and three courses of radiation therapy, but consistently declined amputation as a definitive surgical treatment. The patient was longitudinally treated by a wound care clinic due to drainage from the superficial lesions.

Timeline of Treatment of STS



In June 2024, the patient presented to the emergency room with an infection of the left upper extremity. The option of amputation was again presented and refused by the patient. She received two weeks of empiric antibiotic treatment with overall improvement. However, the patient re-presented two weeks later with worsening drainage and was found to have multiple metabolic derangements in the setting of gram-negative bacteremia. Despite treatment with broad spectrum intravenous antibiotics, the patient continued to decline and developed multiple acute strokes. Ultimately, the decision was made to transition her to comfort care and she died a few hours later.

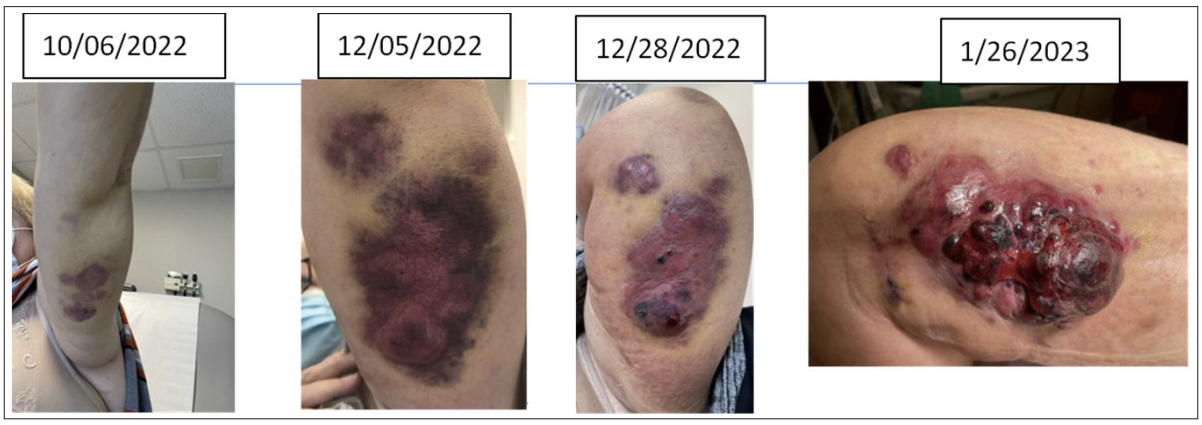


Figure 1: Picture of Lesion Progression from 10/2022-1/26/2023

Pathology

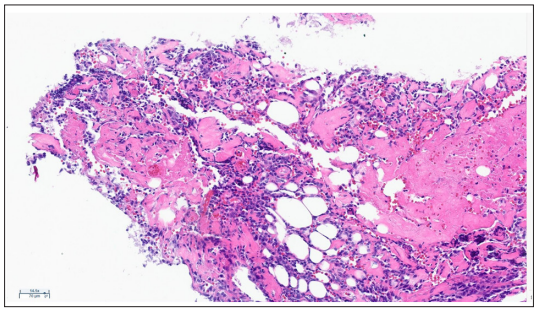


Figure 2a: H&E Stain

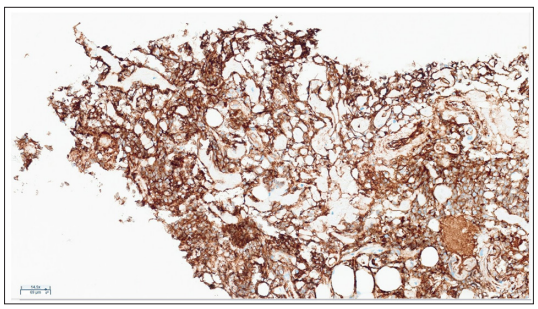


Figure 2b: CD31 stain

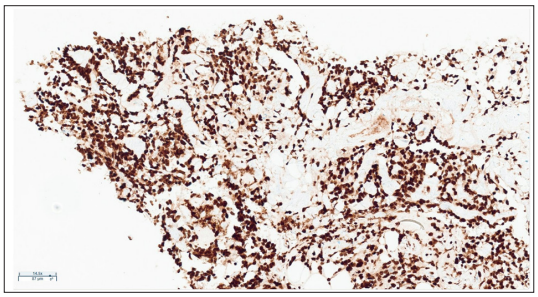


Figure 2c: ERG Stain

Initial Punch Biopsy: "normal skin with minimal superficial lymphohistiocytic inflammation with mild mucin. Prussian blue was negative for iron."

Excisional Biopsy Report: "Excisional Biopsy revealed CD31+, CD34+, ERG+, Pan CK+, and CK7+, negative for cytokeratin, OSCAR, chromogranin, synaptophysin, CDX2, PAX8, CD34, and GATA3. She has extensive lobular/tumor vascular proliferation with high grade cytological atypia, epithelioid as well as spindle cell morphology, involving the entire dermis and the subcutaneous fat. Tumor invasion into a large blood vessel is also identified. Tissue margins, including the deep margin, are invaded by the tumor extensively. This confirms the diagnosis of angiosarcoma."

Imaging

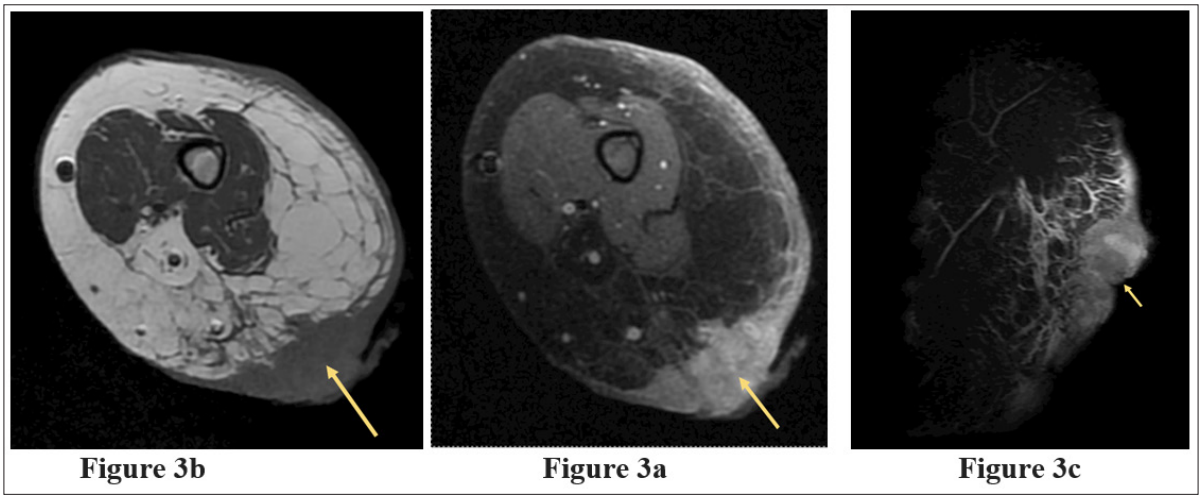


Figure 3 a,b,c: MRI of the left arm with and without contrast from 1/7/2023

Magnetic resonance imaging (MRI) performed after the initial incision and drainage is shown above. Axial T1 (no fat suppression) (image 1), axial T1 post-contrast with fat suppression (image 2), and sagittal STIR (image 3) images demonstrate an ill-defined, mildly exophytic, nodular mass centered within the skin and extending into the subcutaneous soft tissues of the dorsal, ulnar aspect of the distal upper arm. There is adjacent skin thickening and subcutaneous edema. The depth/thickness of the mass has increased from 1.3 cm on the prior motion-degraded, prematurely-terminated MRI from 1 month ago, to 3.0 cm. The mass is heterogeneously T2 hyperintense and predominately T1 isointense to slightly hyperintense to muscle. The mass likely demonstrates heterogeneous nodular enhancement.

Fusion FDG PET/CT image (figure 3d) and the corresponding noncontrast CT in soft tissue window (figure 3e) demonstrates an irregular, nodular, intensely FDG-avid cutaneous/subcutaneous mass within the distal left upper arm [corresponding to the primary mass seen on MRI]. Fusion FDG PET/CT (figure 3f) and noncontrast CT in soft tissue window (figure 3g) depict an adjacent separate intensely FDG-avid satellite nodule inferolateral to the dominant lesion. Maximum intensity projection (MIP) PET image (figure 3h) shows both the dominant and satellite lesion within the left upper arm. [Both lesions demonstrate intense FDG uptake, causing them to blend into one another on the PET MIP.]

Discussion

STS is a rare form of angiosarcoma with poor prognosis that arises in patients with chronic lymphedema. Although most documented cases develop at sites of chronic lymphedema after radical surgery for breast cancer, it may arise in other forms of congenital and acquired lymphedema such as morbid obesity, venous stasis, and surgical trauma [4].

Presentation

STS presents with progressing red and purple ecchymoses which can lead to pain and bleeding in the later stages. Sometimes, STS can present as a violaceous papule that can be mistaken as a hematoma, as in our patient's case. However, these lesions rapidly progress to ulcerating purple-red papules that can necrotize and bleed. There may also be atrophic skin, hyperkeratosis, and telangiectasias associated with the classical dermatological findings of STS [5].

A key differential diagnosis for this syndrome is radiation induced angiosarcoma. The key finding differentiation this from STS is that in STS, there is a key clinical finding of chronic lymphedema without radiation whereas in the radiation induced angiosarcoma,

the patient only has radiation exposure without any edema [6]. Clinically, it is very difficult to diagnose the difference as they present the same way. However, using the clinical history and timeline can help differentiate between the conditions.

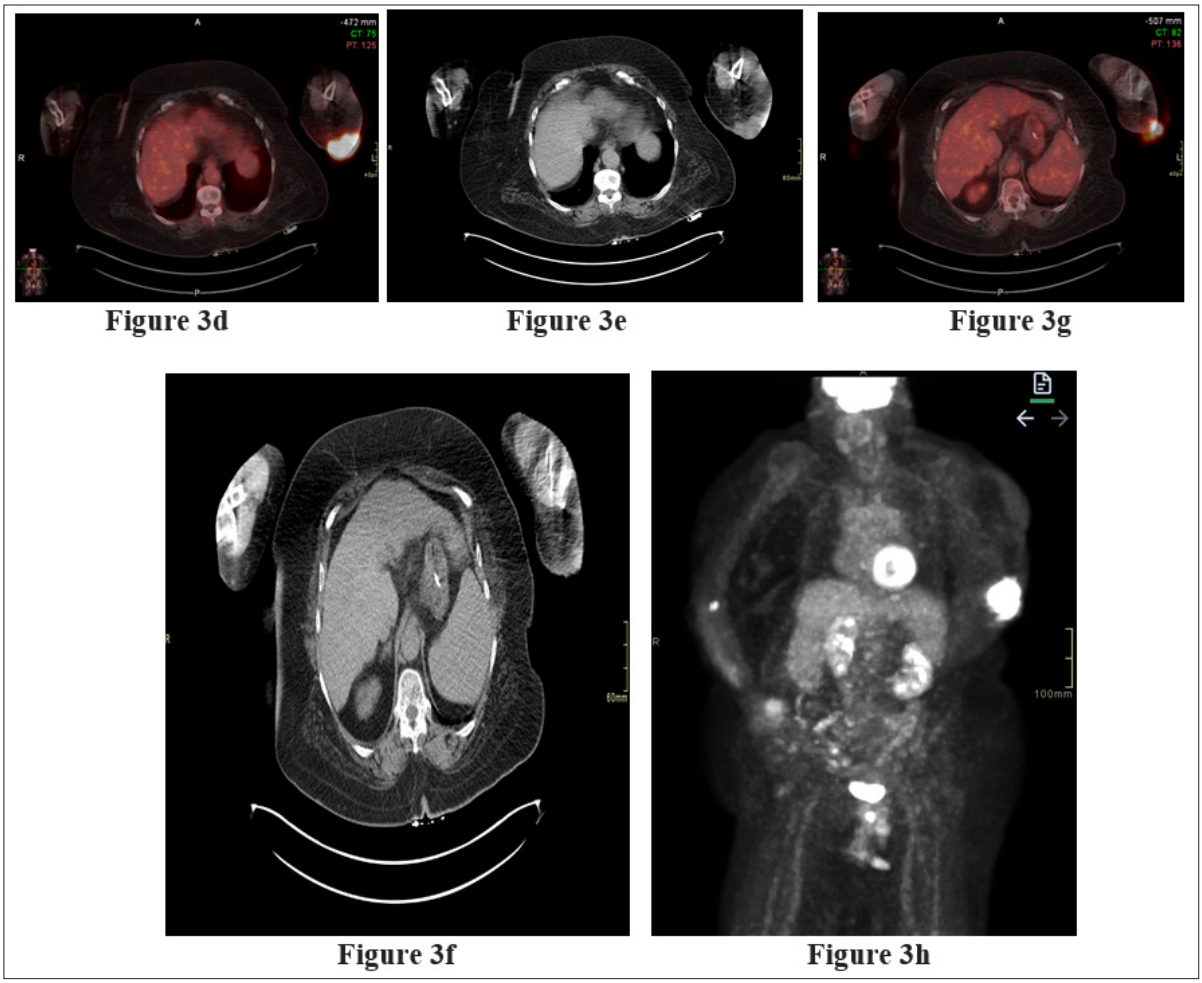


Figure 3 d,e,f,g,h: Initial fluorodeoxyglucose (FDG) PET/CT from 1/13/2023

Diagnostic Testing

The diagnosis of STS is a clinical one supported with imaging and immunohistochemical findings.

Histological findings usually include proliferating vascular channels lined by tumor cells undergoing mitotic changes and displaying hyperchromatism and pleomorphism. The immunohistochemical stains for angiosarcoma include MYC, CD31, CD34, Factor VII, FL-1, ERG (II).

In addition to these tests, the medical history and nature of the lesions should be taken into account. Our patient's progressive skin lesion was initially mistaken for a hematoma. In retrospect, the lesion, in addition to its location (upper extremity), lack of preceding trauma, underlying lymphedema, and prior history of chemotherapy, radiation, and radical mastectomy for breast cancer, raised suspicion for STS. It is important for physicians to be cautious of lesions and skin discolorations in patients who have had a history of chronic lymphedema and mastectomy or radiation

Treatment

Standard treatment for STS is a radical surgical approach due to the aggressive nature of the cancer. The mainstay treatment with the greatest chance of survival is radical amputation of the area of angiosarcoma to minimize the extent of metastatic disease and infection [7]. The five-year survival rate for those who undergo radical amputation is 80%. Patients can also choose to take a conservative approach with chemotherapy and radiation, although this approach is associated with a shorter survival of 15 to 18 months [8]. Many chemotherapy agents such as pembrolizumab, paclitaxel, doxorubicin, gemcitabine, and ifosfamide/anthracycline can be used therapeutically to manage STS, however, overall survival is limited to 8-31 months [9-11].

Table 1: Summary of Outcomes from Case Reports with Varying Treatments for STS with Upper Limb Edema

Case Study (reference)	Treatment Approach	Follow Up	Metastasis
1[X]	TNF- alpha Inhibitor	Died 10 months	yes
2[V]	ifosfamide	Died in 4 months	yes
3[XXII]	doxorubicin	Died in 18 months	yes

4(X)	Extended resection + amputation	Survived for 72 months	no
5(X)	Extended resection chemotherapy	Died in 31 months	no
6(X)	4 patients with Forequarter amputation	Disease free for 3,16, 135 months	no

Prognosis

Despite the various treatment options, the overall prognosis of STS is poor due to the high rate of recurrence and metastatic disease.\ Some common areas of metastasis include the lungs, thoracic wall, liver, bones, soft tissue, brain and lymph nodes. With the frequently delayed diagnosis of STS, approximately 20–40% of patients have metastasis at the time of diagnosis. Metastatic disease at the time of diagnosis plays a significant role in its prognosis. The five-year survival rate for localized angiosarcoma ranges from 30-40% compared to 10-15% for metastatic angiosarcoma [12], thus, early diagnosis is critical and can positively influence survival rate [13]. Earlier diagnosis may facilitate complete surgical resection. With later diagnosis or metastatic spread of tumor, complete surgical resection is often not feasible, leading to recurrence as seen in our patient, and metastasis [14]. Our patient never developed detectable distant metastases, but despite undergoing two local resections, she did develop local recurrence throughout the LUE. Infection is another complication seen in STS. Our patient developed a life-threatening infection of the LUE requiring hospitalization twice within the course of one month that ultimately triggered other sepsis-related complications which she died from.

This case demonstrates the outcome of STS in a patient with a delay in diagnosis, and who chose non-surgical management throughout the disease course [15]. The case highlights the importance of clinicians to understand the clinical presentation and course of STS in order to diagnose it promptly. Early diagnosis can mitigate tumor progression and metastasis. STS must be part of the differential diagnosis in any patient with a history of chronic lymphedema who presents with progressively worsening red-purple raised papules [16]. Given the rapid progression and poor prognosis, it is also important to discuss the risks and benefits of the different treatment options at initial diagnosis [17]. We hope that this case continues to draw attention to this rare disease and offer guidance to physicians who come across these patients in practice [18].

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none

Conflict of Interest

None

Consent

The authors obtained written consent from patients for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available. Patient consent forms were not provided to the journal but are retained by the authors

Additional References

This paper is both a literature review and case report on Stewart Treves Syndrome, a rare condition, hence more than 15 references were used to cite material on the unknown pathophysiology behind this disease.

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