JOURNAL OF COMMUNITY HOSPITAL INTERNAL MEDICINE PERSPECTIVES
2018, VOL. 8, NO. 5, 300–302
https://doi.org/10.1080/20009666.2018.1513293

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CASE REPORT

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# Cutaneous angiosarcoma: a case report of picking the battles in geriatrics

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**ABSTRACT**

Cutaneous angiosarcoma (CA) is a rare form of cancer with limited treatment options and has a very severe prognosis. In this case report, a differential diagnosis, ranging from infection and neoplasia to autoimmune disease, was attributed to recurrent cellulitis centered on a purple lesion. The continuous pursuit of a diagnosis and treatment plan had to be tailored in accordance with patient goals of care. Careful anticipation of disease progression and complications was required following the diagnosis of CA and involved careful transition from palliation to hospice care. Given the aggressive nature of CA, the radical treatment required, and poor prognosis, clinical providers face a difficult task of balancing diagnostic and therapeutic steps with patient goals of care.

**ARTICLE HISTORY**
Received 12 May 2018
Accepted 9 August 2018

**KEYWORDS**
Cutaneous angiosarcoma; recurrent cellulitis; severe prognosis; limited treatment options

## 1. Introduction

The association between chronic lymphedema and cutaneous angiosarcoma (CA) was first described in 1948 by Stewart and Treves in a case series of breast cancer patients undergoing mastectomy [1]. Although CA is a rare pathology, the tumor has been reported to affect different sites, including the lower extremities [2–6], usually involving older patients. It is a very aggressive disease with a poor prognosis, particularly if it has metastasized [7]. Thus, treatment also has to be aggressive and includes radical surgery, chemoradiotherapy, and occasionally novel agents [6,8,9].

## 2. Case report

Ms A, a 90-year-old woman with a known past medical history of hypertension, chronic obstructive pulmonary disease, lower extremity edema, cognitive deficit, and urinary incontinence, was admitted to the hospital for cellulitis in her right lower extremity, that initially responded to intravenous vancomycin. She was readmitted for recurrence and growth of the discolored skin area, consistent with cellulitis that did not recede after the administration of oral antibiotics (doxycycline and sulfamethoxazole). Following the last admission, she had developed an extensive area of red-purple discoloration surrounding a dark purple, raised round lesion that was 2 cm in diameter, with a smooth surface and puffy consistency. The lesion was located in the anterior aspect of her right shin.

Despite extensive discoloration of the skin area, the patient was afebrile and did not have leukocytosis. Intravenous vancomycin was resumed, and the differential diagnosis was expanded to include vasculitis and neoplasia. A small fluid collection with internal echoes was identified on ultrasound of the dark purple lesion, leading to suspicion of organizing hematoma or bacterial abscess. However, needle aspirations of the lesions yielded only a few polymorphonuclear leukocytes and the absence of bacteria (an erythrocyte sedimentation rate [ESR] of 69).

The differential diagnoses were revised to include cutaneous malignancy. Given the purple discoloration and associated chronic lymphedema, consideration was also given to CA, and cutaneous B- or T-cell lymphoma. Wedge skin biopsy was recommended by the dermatologist. However, The patient would not consent to the surgical procedure owing to cognitive deficit (associated with Montreal Cognitive Assessment score of 11/30, with the identification of cerebral atrophy on head computed tomography).

Previously, the patient authorized one of her two daughters to be a surrogate medical decision-maker. The other daughter could not commit to a definitive decision because of her own medical history of traumatic brain injury. During discussions about the risks and the benefits of the procedure, the surgeon expressed significant concern about lack of healing at the surgical site, particularly in the context of chronic edema and the patient's very frail skin.

Oncology was consulted for alternative diagnostic approaches. Biopsy remained the optimal diagnostic test but was not pursued given the likelihood of a non-healing ulcer at the surgical site. Radiotherapy was

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recommended as the most suitable treatment for palliative purposes, based on suspicion of neoplasia.

The ESR increased from 69 to 86 and intravenous antibiotics were stopped after a 10-day course, by which time the C-reactive protein levels had normalized. The patient was discharged from the hospital, subject to clinical observation of the reported skin lesion, despite significant anxiety over the need to obtain a diagnosis and the inability to perform a biopsy. The patient's daughter did not consent to bringing Ms A for radiation therapy as the daughter believed that it would be too difficult for the patient and that it would impact negatively on her quality of life.

Subsequently, the dark purple lesion continued to grow and eventually ruptured. A biopsy was performed by the surgeon during a house call visit. Given her cognitive deficit, the patient was not fully able to understand her condition. In addition, despite learning about the poor prognosis attributed to the disease, the two daughters initially refused to admit their mother in hospice. At that stage, potential future clinical complications were envisioned, including infection, bleeding, and deep vein thrombosis.

Wound care was pursued using silver alginate to prevent infection at the lesion site. By then, the clinical course was complicated by bleeding and pain. Opiates were used for pain control and ambulation became progressively more difficult, despite the patient having assistance at home.

A hemoglobin level of 10 g/dL was recorded on patient discharge. Once home, the bleeding increased in intensity. Ambulation was still possible, but with considerably more effort and increased shortness of breath. The surrogate decision-maker continued to refuse a hospice admission. During a subsequent house call visit, when asked if the patient had any chest pain, the daughter agreed that she did and that it was a new symptom. The physician then re-discussed the need for hospice care with the daughters and explained that it was likely that myocardial infarction (MI) would complicate the bleeding. It was explained that MI is associated with significant distress and an increased need for pain- and anxiety-reducing medication, which eventually helped to overcome the family's resistance to the idea of admitting her to a hospice.

The patient continued to experience pain that increased in intensity once in hospice and the bleeding worsened to the point where the leg wound dressings were becoming saturated daily. The subsequent clinical course was complicated by delirium, at which point the daughters called 911 and had the patient was sent to the emergency room. The patient received a blood transfusion for a hemoglobin level of 4.6 g/dl Complications relating to the potential need for repeated blood transfusions were pivotal to discussions about patient treatment. Although blood transfusions would reduce the patient's shortness of breath, chest pain, and delirium, they would also artificially prolong her life, thereby exposing the patient to additional complications, and, in particular, worsening pain. After careful and involved counsel with the patient and her family, a decision was taken not to subject her to further blood transfusions. Further treatment centered exclusively on symptom control and she died peacefully at home, surrounded by her family.

## 3. Discussion

CA, alternatively known as Stewart-Treves syndrome, is a rare disease, has a poor prognosis, mainly affects elderly populations, and requires aggressive surgical therapy. The current case study did not focus on the pathological mechanisms nor treatment options available for this devastating disease. Instead, it was concerned with how patients and their family, as well as clinicians, face significant dilemmas in terms of its diagnosis and treatment, including the need to adjust the latter to meet reasonable, patient/family goals of care.

In the current case, cognitive deficit prevented the patient from making medical decisions. As a further logistical complication, one of her daughters could not participate meaningfully in the medical decision-making process as a consequence of her own neurological condition. This placed the burden of decision-making solely on the other daughter, further placing her at risk of emotional burnout. As expected, both daughters went through the Kübler-Ross stages of grief (i.e., denial, anger, bargaining, depression, and acceptance).

In addition, the patient and her family experienced a difficult transition from aggressive care to palliation. Initially, the decision-making process centered on whether to adhere to diagnostic procedures, given the expected lack of healing after a biopsy. As surgery was not an option, and as palliative radiation therapy was also considered too burdensome for the patient, a decision was taken to proceed with home-based palliative care, with the physician providing house call visits.

Once the lesion broke spontaneously, the surgeon graciously performed a biopsy during a house call visit. Once the diagnosis had been established, the care shifted from palliation to admission into hospice. Judicious steps were taken to help the patient and her family navigate the decisions taken regarding the care goals. The clinician helped by attempting to foresee potential complications, actively searching for anticipated symptoms and explaining that these were part of the natural course of the disease for which hospice care was required. This approach helped to prevent patient suffering and actively involved the family in the management of her disease, while helping the family members in their transition toward the loss of their loved one.

Thus, clinicians should not only focus on the epidemiology, diagnosis, prognosis, and treatment of

distinct pathologies. Instead, they need to remain actively aware of how diagnostic and clinical dilemmas impact patients and their families, especially in the case of terminal illness. Continuous consideration of the care goals is required in such cases.

The current case report emphasizes the need to constantly assess care goals during the CA diagnostic process and to carefully balance the anticipated treatment with patient frailty and preferences. This is especially warranted in the mostly frail populations affected by this disease, particularly because CA has such a poor prognosis.

## Disclosure statement

No potential conflict of interest was reported by the authors.

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