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Skin Angiosarcoma Masquerading as Hematoma in an Elderly Woman

Case Presentation

A 78 year old Caucasian, bed-bound, nursing home female, presented with non-tender, violaceous, indurated plaques of the skin of her anterior left thigh. Patient's past medical history was significant for coronary artery disease, atrial fibrillation, diabetes mellitus type II, and stroke with left hemiplegia. On physical examination of her thigh, a 5X8 cm diffuse dusky red, woody hard, hematoma-like plaque was noted without inguinal lymphadenopathy or leg edema (Figure 1). Laboratory studies (comprehensive metabolic panel, complete blood count, and, coagulation factors) were unremarkable. Initial assumption was the lesions were hematomas, possibly secondary to trauma and heat packs were applied for patient comfort.

Over the next two weeks, despite conservative management, the lesion on the anterior thigh extended to the lateral and posterior thigh. In addition, dark serous drainage appeared from 3-4 open areas (Figure 2). A vascular pathology rather than hematoma was suspected. Dermatology consultant performed a skin biopsy and initial results suggested a Kaposi sarcoma, but were not conclusive. HIV/AIDS studies were negative. A deeper second biopsy was conducted and favored the diagnosis of angiosarcoma. The diagnosis was further supported by a third biopsy showing the morphology of the spindle cells and negative HHV-8 staining, consistent with angiosarcoma (Figure 3 and Figure 4). Skin lesions continued to spread locally with emergence of more satellite lesions (Figure 5). Available treatment options including surgical, chemotherapeutic, and radiation interventions were discussed with the resident and daughter (medical power of attorney). Given the presence of multiple underlying co-morbidities, poor baseline functional status, and widespread local disease, resident elected not to pursue any further aggressive diagnostic or therapeutic options. At a meeting requested by the resident to review goals of care, she elected comfort care with hospice services. Due to the increased frequency of dressing changes



Figure 1: Initial appearance of thigh lesion.

Case Report



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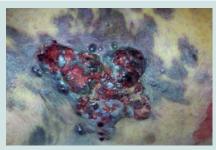


Figure 2: Appearance of thigh lesion one month later.

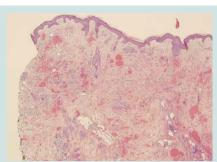


Figure 3: Overview image- epidermis and dermis stained with hematoxylin and eosin (Courtesy of Dominion Pathology Laboratories).

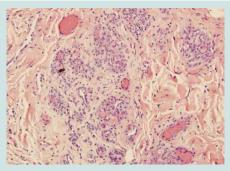


Figure 4: Solid sheets of atypical spindle cells organized as islands throughout dermis (Courtesy of Dominion Pathology Laboratories).

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Figure 5: Appearance of thigh lesion after 3 months.

from wound drainage, she expressed mild pain to hip which was treated effectively with Tramadol, prior to dressing changes. One month later, with daughter at bedside, the resident died comfortably.

Discussion

Angiosarcomas are rare malignant tumors of endothelial origin that carry a poor prognosis and high tendency for local and metastatic spread [1]. The most common sites, in decreasing order, are the skin, breast, deep soft tissues, visceral organs and bone [2]. Cutaneous angiosarcoma is a great mimicker with many clinical presentations that include hematoma-like lesions, violaceous nodules and plaques. It is confined to the face and scalp region in more than 50% of cases [3]. It is an uncommon, malignant neoplasm that contains rapidly proliferating, extensively infiltrating anaplastic cells from blood vessels. Angiosarcomas in general have a similar distribution between sexes, can develop at any age, but are more common in older patients; with the cutaneous form most common in elderly white men [4-6]. Most angiosarcomas arise spontaneously, but there are a few reports of malignant transformation within pre-existing benign vascular lesions [7]. Radiotherapy is an independent risk factor, specifically in breast lesions [8]. Various chemicals such as vinyl chloride and thorium dioxide are associated with hepatic angiosarcomas [9,10]. Lymphedema is one of the well-known risk factors for the development of cutaneous angiosarcomas [11]. Our patient had no known risk factors for angiosarcoma.

Angiosarcomas have high rates of local recurrence and metastases in part because they are unrecognized or misdiagnosed [10], leading to a poor prognosis and a high mortality rate. Differential diagnoses of angiosarcoma include hemangioblastoma, Kaposi's sarcoma, and metastatic cancer of unknown origin [12]. Histological features of angiosarcoma can vary among cases. The hallmark of angiosarcoma is malignant endothelial cells that are round, polygonal or fusiform and can have an epithelioid appearance. Angiosarcomas usually express endothelial markers such as von Willebrand factor, CD34, CD31, and vascular endothelial growth factor [10]. Immunohistochemistry is important in confirming the diagnosis of skin lesions.

The prognosis of cutaneous angiosarcoma was considered extremely poor in the past, with an overall 5-year survival rate less than 20% [13]. In recent years, several prognostic factors have been analyzed. Poor prognostic factors include old age, metastatic disease at presentation, poor patient performance status at baseline, presence of other soft-tissue sarcomas, large tumors (>5cm), and high tumor

grade [1,14]. The site of the sarcoma is also important. Angiosarcoma of the viscera (liver, heart) and retroperitoneal disease have poorer outcomes [1]. Diagnosis relies on biopsy to confirm histology and MRI to delineate extent of primary lesion and endothelial markers. Treatment modalities are limited. For local disease, radical surgery with complete resection is the primary modality, complemented by adjuvant radiotherapy and, in some cases, adjunct chemotherapy. For metastatic disease, cytotoxic chemotherapy is the primary treatment modality [10].

In summary, cutaneous angiosarcoma of the elderly is a great mimicker with widely variable clinical presentations. This condition needs to be differentiated from several other common pathologies. The diagnosis may be challenging as illustrated in our case where several skin biopsies were required to ascertain the diagnosis of angiosarcoma of the skin. Given the poor prognosis of angiosarcoma in frail older patients, treatment options for extensive disease should include palliative care rather than aggressive interventions.

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