When a Rash Goes Deeper Than the Surface: A Case of Angiosarcoma

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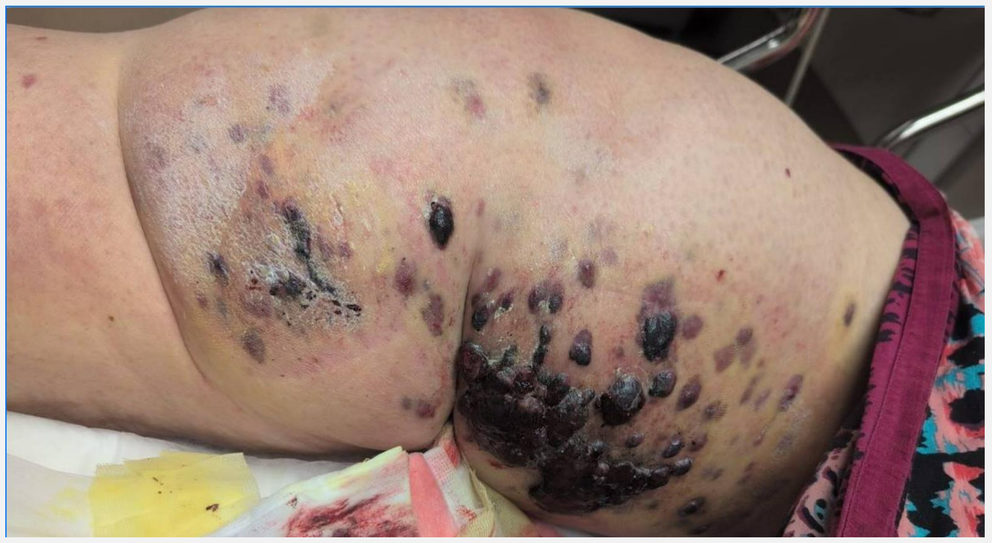
Background

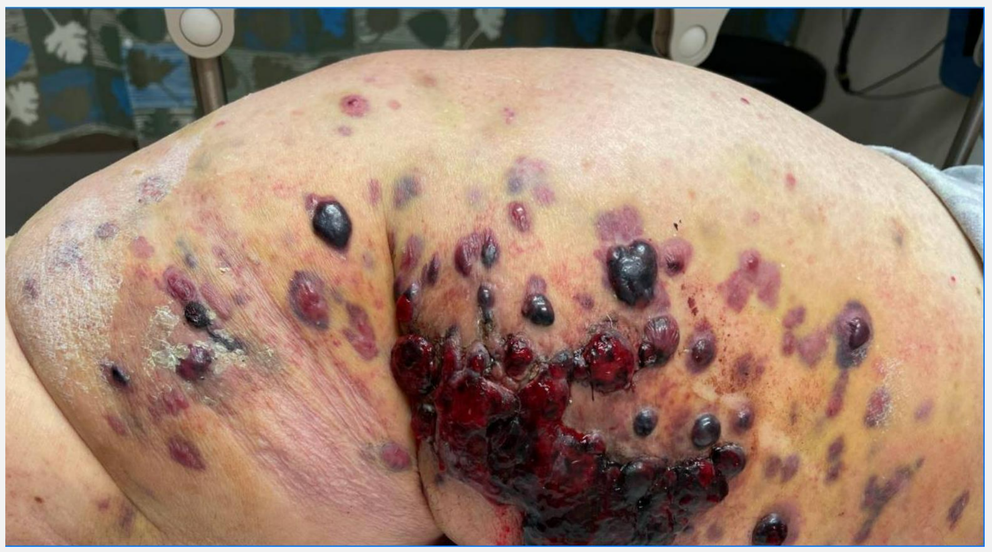
- Angiosarcomas are rare malignant tumors of endothelial origin, most often presenting as cutaneous lesions of the head and neck in elderly patients.
- Prognosis is poor due to aggressive behavior and frequent metastasis.
- Diagnosis is often delayed - initial presentation as purplish papules or erythematous plaques may mimic benign conditions such as dermatitis or shingles.

Case Report

- 78-year-old female presented to ED with shortness of breath and a flat, purpuric rash on the right medial thigh that developed over 4–6 weeks, associated with burning, later becoming pruritic, painful, and bullous.
- Past medical history significant for multiple malignancies: right breast DCIS (2002) treated with lumpectomy, radiation, and anastrozole; recurrence with invasive ductal carcinoma (2012) treated with mastectomy, radiation, and letrozole; and stage I endometrial cancer (2011) treated with TAH+BSO.
- Initial workup revealed right lobar consolidation and spiculated left lung nodules; she was treated empirically for pneumonia and presumed shingles with valacyclovir, doxycycline, and cefpodoxime.
- Rash continued to enlarge and bleed, prompting multiple ED visits in the following month and subsequent hospital admission for acute blood loss anemia.
- Punch biopsy initially showed a poorly differentiated malignant neoplasm concerning for cutaneous metastasis from prior cancer.
- However, second opinion revealed positive staining for ERG, D2-40, CD34 (multifocal), and CD31 (focal) consistent with high-grade epithelioid angiosarcoma.

Images





Case Report (cont'd)

- Lung nodules seen on prior imaging were interpreted as metastatic disease.
- Her hospital course was complicated by hypotension, acute kidney injury and significant deconditioning.
- Though initially unclear due to body habitus, she was later noted to have chronic lymphedema of the lower limbs, likely congenital.
- Due to poor functional status and advanced disease, she was not a candidate for surgery or systemic chemotherapy and goals of care discussions were pursued.
- She then underwent palliative radiation therapy for local bleeding control, with role of dose-adjusted chemotherapy to be reconsidered after radiation.

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Discussion

Risk factors:

- Prior radiation exposure, chronic lymphedema (Stewart–Treves syndrome), and chemical exposure such as vinyl chloride or arsenic.
- Chronic inflammation and lymphatic obstruction are thought to play a key role in tumor development.

Pathologic characteristics:

- High-grade endothelial tumors with rapid growth, local recurrence, and early hematogenous metastasis.
- Common metastatic sites include the lungs, liver, and bone.

Treatment approaches:

- Localized disease is managed primarily with wide surgical excision with negative margins, often followed by adjuvant radiation therapy to reduce recurrence risk.
- Unresectable or metastatic disease is treated with systemic chemotherapy, most commonly taxane-based (paclitaxel, docetaxel) or anthracycline-based (doxorubicin) regimens.
- Anti-angiogenic agents such as TKIs that inhibit VEGF (bevacizumab) and VEGFR (pazopanib) may offer benefit in select patients, though responses are variable.
- Clinical trials are ongoing to explore the role of immunotherapy and targeted therapies.

Clinical significance:

- Requires a high index of suspicion for persistent, atypical, or treatment-refractory rashes, especially in elderly patients or those with known risk factors.
- Delayed diagnosis is common and contributes to poor prognosis and limited treatment options

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