Purpuric eruption of the breasts, abdomen, thighs, and buttocks

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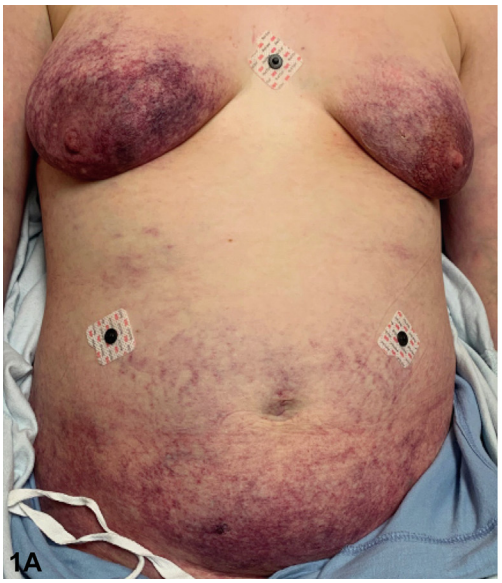
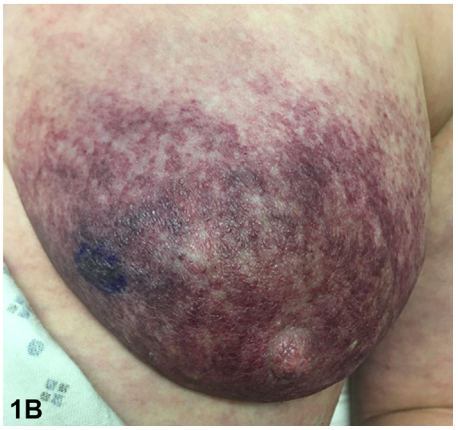
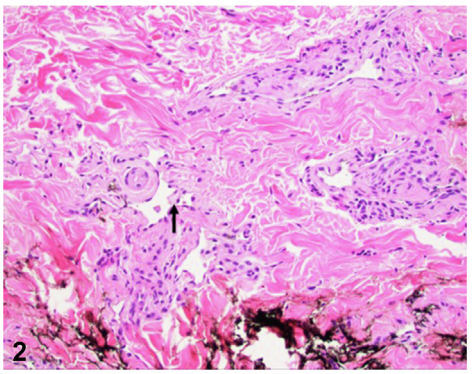
Key words: Reactive angioendotheliomatosis; systemic lupus erythematosus; lupus; SLE.







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Funding sources: None.
IRB approval status: Not applicable.
Prior presentation: None declared.

Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

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JAAD Case Reports 2022;30:140-2.
2352-5126

https://doi.org/10.1016/j.jdcr.2022.04.031

A 24-year-old female smoker presented to the emergency department with photosensitivity, rash, fatigue, and abdominal pain with diffuse lymphadenopathy. She noted 2 years of violaceous mottling of the skin, but in the months prior to admission had developed deeper, tender nodules in these areas. Laboratory workup was notable for anemia, leukopenia, and positive anti-Smith and antinuclear antibodies. Antiphospholipid antibodies and HIV screen were negative. Physical examination revealed purpuric patches and indurated nodules with superimposed petechiae and ecchymoses on the bilateral breasts, abdomen, thighs, and buttocks, with fixed, non-blanching livedoid patches of the trunk and legs (Fig 1). Superficial erosions on the hard palate, patchy erythema of the cheeks and neck, and a diffuse non-scarring alopecia of the scalp without erythema or scale were noted. A punch biopsy was performed (Fig 2).

Question 1: What cutaneous diagnosis is most likely?

A. Angiosarcoma

B. Kaposi's sarcoma

C. Reactive angioendotheliomatosis (RAE)

D. Polyarteritis nodosa (PAN)

E. Calciphylaxis

Answers:

A. Angiosarcoma — Incorrect. Angiosarcoma is more typically seen in older adults, and would most commonly appear as indurated purpuric or bruise-like lesions on the face and scalp.

B. Kaposi's sarcoma — Incorrect. Kaposi sarcoma would typically present with slowly enlarging vascular patches or plaques with more defined demarcation than this case.

C. RAE — Correct. RAE is a rare, benign entity characterized by proliferation of endothelial cells within vascular lumina.[1,2] It is often seen in patients with coexistent systemic disease, such as autoimmune disease (as in our patient, who met criteria for systemic lupus erythematosus), chronic infection, lymphoproliferative disorder, cryoproteinemia, severe atherosclerotic disease, or valvular heart disease.[1-4] RAE may present with erythematous to violaceous patches, plaques, or nodules, with or without a livedoid pattern, necrosis, blisters, or ulceration.[1-4] The presence of superimposed focal purpura, petechiae, or ecchymoses may serve as a clinical clue.

D. PAN — Incorrect. The clinical features of cutaneous PAN can be similar to RAE, commonly presenting with lower extremity subcutaneous nodules, livedoid changes, and occasionally ulceration.

E. Calciphylaxis — Incorrect. Retiform purpura and marked pain would be more classic for a diagnosis of calciphylaxis. The lesions of calciphylaxis are most commonly noted on the lower legs, though a large portion of patients can also exhibit involvement on central, adipose-rich areas of the body; acral and genital lesions are also possible.

Question 2: Which of the following histopathologic findings would be most compatible with this diagnosis?

A. Proliferation of endothelial cells within vascular lumina

B. Fibrin deposition within vessel walls with perivascular neutrophils and leukocytoclasia

C. HHV8 expression by immunohistochemistry

D. Deposition of calcium in eccrine gland basement membranes

E. A granulomatous infiltrate involving medium-sized vessels

Answers:

A. Proliferation of endothelial cells within vascular lumina — Correct. Histopathologic features may vary among lesions, even within the same patient. Findings may include dilated blood vessels, a benign intraluminal proliferation of endothelial cells that often occludes lumina, and/or associated fibrin thrombi.[1-4] A broad range of additional histologic phenotypes have been reported, including glomeruloid hemangioma-like, tufted angioma-like, diffuse dermal angiomatosis, and others.[4]

B. Fibrin deposition within vessel walls with perivascular neutrophils and leukocytoclasia — Incorrect. These would be the histologic features of a small vessel vasculitis, which most typically presents as palpable petechiae on the lower extremities.

C. HHV8 expression by immunohistochemistry — Incorrect. This would be a key feature in the histologic diagnosis of Kaposi sarcoma.

D. Deposition of calcium in eccrine gland basement membranes — Incorrect. This would be a diagnostic feature of cutaneous calciphylaxis.

E. A granulomatous infiltrate involving medium-sized vessels — Incorrect. This would typify a diagnosis of cutaneous polyarteritis nodosa.

Question 3: Which of the following treatment options would be most appropriate?

A. Systemic steroids

B. Sodium thiosulfate

C. Warfarin

D. Resection

E. Radiation therapy

Answers:

A. Systemic steroids — Correct. RAE may respond to therapies targeting the underlying disorder,[2] including the treatment of systemic lupus erythematosus such as in our patient. There are also reports of spontaneous resolution in cases of RAE.[4] In addition, a workup for causes of vascular thrombosis is recommended.

B. Sodium thiosulfate — Incorrect. This would be the treatment of choice for a diagnosis of calciphylaxis.

C. Warfarin — Incorrect. This is not a recommended treatment option for RAE.

D. Resection — Incorrect. Resection is a treatment option in the management of both angiosarcoma and Kaposi sarcoma.

E. Radiation therapy — Incorrect. Radiation therapy is a potential treatment in the management of both angiosarcoma and Kaposi sarcoma.

Abbreviations used:

ANA: antinuclear antibody
HHV8: human herpesvirus-8
HIV: human immunodeficiency virus
RAE: reactive angioendotheliomatosis
SLE: systemic lupus erythematosus

Conflicts of interest

None disclosed.

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