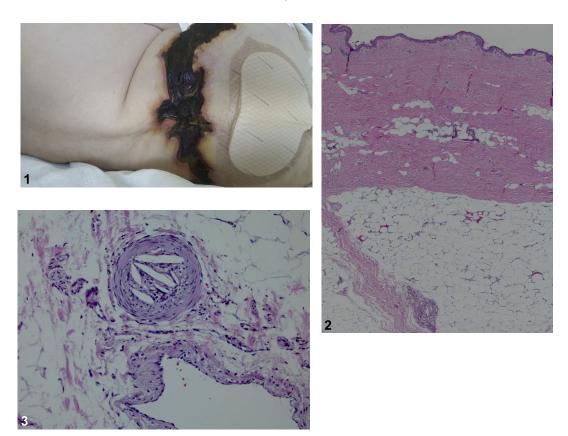
Extensive skin necrosis



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A 62 year-old woman was hospitalized for progressive, painful, skin necrosis of the lower back, bilateral hips, and abdomen and acute kidney injury and altered mental status.

Her medical history included coronary artery disease, hypertension, hyperlipidemia, type II diabetes mellitus, stage III chronic kidney disease, seizure disorder, and emergency percutaneous coronary intervention via right femoral artery, 2 months prior. Physical examination found extensive stellate eschars on the lower back and hips, focally scattered on abdomen, and right proximal lower extremity. She also had transient livedo reticularis (Fig 1). Laboratory workup was remarkable for mild peripheral eosinophilia of 3.9%, blood urea nitrogen (BUN), 52; creatinine, 2.2; erythrocyte sedimentation rate (ESR), >30; and C-reactive protein (CRP), 7.9.

Figures 2 and 3 show the findings from an incisional skin biopsy from a purpuric nodule on the right lateral thigh.

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Question 1: What is the most likely diagnosis?

- A. Antiphospholipid antibody syndrome
- B. Calciphylaxis
- **C.** Cholesterol embolization syndrome (CES)
- D. Cryoglobulinemia
- E. Warfarin necrosis

Answers:

- **A.** Antiphospholipid antibody syndrome—Incorrect. In antiphospholipid antibody syndrome, stroke and transient ischemic attack are the most common arterial events, whereas lower extremity deep vein thrombosis is the most common venous event. ¹
- **B.** Calciphylaxis—Incorrect. Calciphylaxis classically occurs in patients with end-stage renal disease; however, nonuremic calciphylaxis has been reported in literature.
- C. CES-Correct. Cholesterol embolization syndrome is a life-threatening condition that results from atherosclerotic plaque embolization from the aorta or its major branches. It can occur spontaneously, but is generally secondary to invasive vascular procedures and anticoagulant therapies.²⁻⁴ There can be a significant delay between the causative factor and symptom onset, making diagnosis difficult. Furthermore, cutaneous findings can mimic those seen in more commonly encountered dermatologic diseases, such as calciphylaxis and vasculopathy. Biopsy specimens should be obtained by elliptical incisions over blanched portions of livedo reticularis or firm purpuric nodules and include fat. Histologically, cholesterol emboli produce elongated clefts within the lumen of small subcutaneous vessels (Figs 2 and 3).
- **D.** Cryoglobulinemia—Incorrect. Cardinal signs of cryoglobulinemia include retiform, purpuric, or necrotic lesions affecting acral sites.
- **E.** Warfarin necrosis—Incorrect. Warfarin necrosis occurs 2 to 5 days after starting warfarin in the absence of heparin.

Question 2: What is the *most common* laboratory finding in CES?

- A. Neutropenia
- **B.** Peripheral blood eosinophilia

- C. Leukocytosis
- **D.** Elevated ESR
- E. Thrombocytopenia

Answers:

- **A.** Neutropenia—Incorrect. Neutropenia is not a finding in CES.
- **B.** Peripheral blood eosinophilia—Correct. Peripheral blood eosinophilia is common, occurring in up to 80% of cases.³ Fluctuations in eosinophilia count can be a clue to recurrent showering of emboli.
- **C.** Leukocytosis—Incorrect. Leukocytosis can occur in CES but is not the most common laboratory abnormality.
- **D.** Elevated ESR—Incorrect. ESR elevation can occur in CES, but is not the most common finding.
- **E.** Thrombocytopenia—Incorrect. Thrombocytopenia is not associated with CES.

Question 3: Potential therapies for CES include which of the following?

- **A.** Systemic corticosteroids
- **B.** Pentoxifylline
- **C.** HMG-CoA reductase inhibitors
- **D.** Antiplatelet agents
- **E.** All of the above

Answers:

- A. Systemic corticosteroids
- **B.** Pentoxifylline
- **C.** HMG-CoA reductase inhibitors
- **D.** Antiplatelet agents
- **E.** All of the above—Correct. Treatment is supportive with a multidisciplinary approach to optimize care. All of the above therapies have been advocated to be effective in select cases, but there are no standard therapeutic guidelines.^{3,5}

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