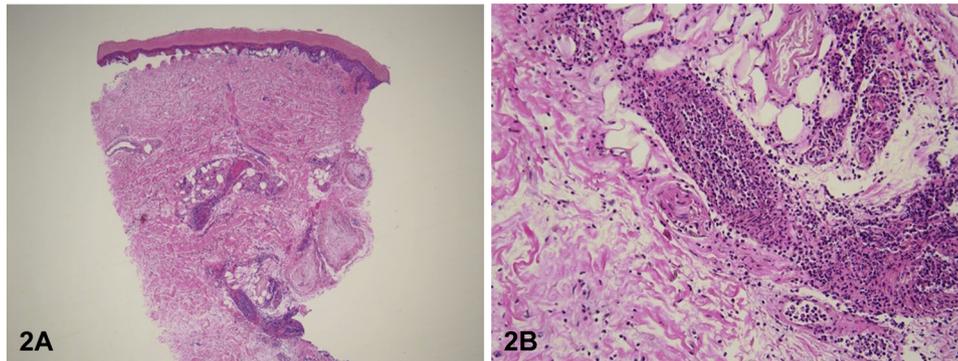


Painful violaceous bullae of the hands



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A 63 year-old-woman with a medical history of aortic valve replacement on anticoagulation, type II diabetes, and recently diagnosed ovarian cancer presented to the emergency department. She had fever, chest pain, a lactic acid level of 7.2 mmol (normal, 0.5-2.0 mmol), pulse rate of 154, and several painful violaceous bullae of the palms with surrounding stellate macules and rare splinter hemorrhages (Fig 1). Results of a punch biopsy are shown in Fig 2.

Question 1: What is the most likely diagnosis?

- A. Coumadin-induced necrosis
- B. Drug-induced vasculitis
- C. Calciphylaxis
- D. Septic emboli
- E. Porphyria cutanea tarda

Answers:

A. Coumadin-induced necrosis — Incorrect. The classic distribution often involves areas with excessive adipose tissue, like the abdomen, thighs, or breasts. The duration of anticoagulation was not specified in the history but would typically be within the first 2 weeks of anticoagulation.

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B. Drug-induced vasculitis — Incorrect. The patient history does not indicate any recent changes in medications, and the distribution would be more widespread on the trunk and extremities. Additionally, signs and symptoms of sepsis would remain unexplained.

C. Calciphylaxis — Incorrect. Calciphylaxis commonly occurs in the setting of end-stage renal disease and secondary hyperparathyroidism. The distribution is typically more localized to the abdomen, thighs, and legs and would not typically be on the hands or feet. This would also not account for the acute illness depicted in the question stem.

D. Septic emboli — Correct. The patient presented with characteristic findings of septic emboli including splinter hemorrhages and distal extremity stellate purpura. Our patient's symptoms support an infective process. Also, her history of aortic valve replacement and recent cancer diagnosis make her susceptible to bacteremia with endocarditis complicated by *Streptococcus pyogenes*.^{1,2}

E. Porphyria cutanea tarda — Incorrect. The diagnosis of porphyria cutanea tarda would appear more commonly on the dorsal aspect of the hands, forearms, chest, and face. The patient history would most likely include the development of skin findings after sun exposure and a history of liver disease. Lastly, this answer choice did not account for the acute illness of the patient.

Question 2: Which features would you expect to see on the histopathologic examination?

A. Subepidermal cleft formation, minimal inflammatory infiltrate with festooning of the dermal papillae, and few caterpillar bodies

B. Calcified small vessels in the subcutaneous fat with epidermal necrosis

C. Affected and unaffected areas with dermal neutrophilic vasculitis and subepidermal cleft formation

D. Subepidermal cleft formation with scattered neutrophils and eosinophils and mild perivascular lymphocytic inflammation

E. Full-thickness epidermal necrosis with subepidermal cleft formation with minimal inflammatory infiltrate

Answers:

A. Subepidermal cleft formation, minimal inflammatory infiltrate with festooning of the dermal papillae, and few caterpillar bodies (porphyria cutanea tarda) — Incorrect. Although a subepidermal cleft appears on histopathology (Fig 2, A), the lack of inflammatory infiltrate does not correspond with what is seen histologically, a robust neutrophilic vasculitis.

B. Calcified small vessels in the subcutaneous fat with epidermal necrosis (calciphylaxis) — Incorrect. Calciphylaxis is defined by characteristic calcified small vessels, typically located in the subcutaneous tissue. There was no calcium identified upon histologic examination.

C. Affected and unaffected areas with dermal neutrophilic vasculitis and subepidermal cleft formation (septic emboli) — Correct. Histopathology (Fig 2, A) represents a punch biopsy of acral skin with a subepidermal cleft. Histopathology (Fig 2, B) shows a neutrophilic vasculitis with infiltration of deeper dermal vessels with bacteria. The histologic pattern and clinical presentation are most consistent with septic vasculitis.

D. Subepidermal cleft formation with scattered neutrophils and eosinophils and mild perivascular lymphocytic inflammation (linear IgA dermatosis) — Incorrect. Although there are a subepidermal split and focal collections of neutrophils, they are located in the reticular dermis with destruction of small vessels. For linear IgA dermatosis, the infiltrate is typically at the dermoepidermal junction.

E. Full-thickness epidermal necrosis with subepidermal cleft formation with minimal inflammatory infiltrate (Steven-Johnson syndrome) — Incorrect. Although there is some epidermal necrosis noted, the inflammatory infiltrate is too robust with a neutrophilic vasculitis present. Those features are not typically seen in Steven Johnson syndrome.

Question 3: What is the best initial management for this condition?

A. Removal of the offending medication

B. Corticosteroids

C. Sodium thiosulfate

D. Appropriate antibiotic therapy

E. Discontinuation of Coumadin

Answers:

- A.** Removal of the offending medication — Incorrect. Withdrawal of the suspected offending agent is critical in the management of Steven-Johnson syndrome. Our patient's clinical picture was secondary to an infective process, not a medication.
- B.** Corticosteroids — Incorrect. Corticosteroids are considered a reasonable treatment for drug-induced vasculitis such as cutaneous small vessel vasculitis but have limited efficacy in treating septic emboli secondary to infection.
- C.** Sodium thiosulfate — Incorrect. Sodium thiosulfate is the preferred treatment for calciphylaxis but has a limited role in treating septic emboli.
- D.** Appropriate antibiotic therapy — Correct. Recommended management of septic embolism caused by infective endocarditis is with long-term intravenous antibiotics with the appropriate bacterial

coverage. In suspected endocarditis, patients are started on broad-spectrum antibiotics and switched to antibiotics with appropriate coverage after culture results return.³

E. Discontinuation of Coumadin — Incorrect. There is no role for discontinuation of Coumadin in the setting of septic emboli secondary to infective endocarditis. In fact, if there is evidence of a vegetation on a mechanical valve, the anticoagulation should be optimized for best results.

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