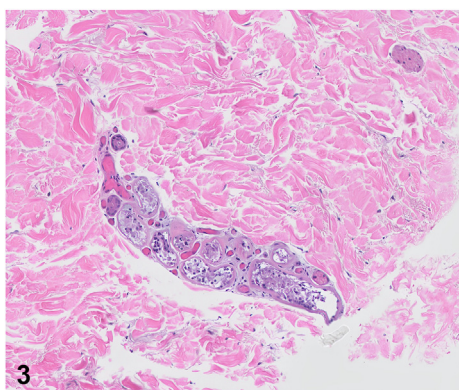


Acral palpable purpura with rapidly progressing multiorgan involvement



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A 36-year-old man from Southeast Ohio presented in August with fever, diarrhea, rapidly progressing multiorgan failure, and palpable purpura on the malar cheeks with periorbital and nasolabial sparing (Fig 1), ears, penis, and fingertips (Fig 2). He reported bug bites a week prior but denied tick exposure. He denied illicit drug use. Labs showed kidney and liver injury, microangiopathic hemolytic anemia, thrombocytopenia, and elevated fibrinogen. D-dimer was normal. Serum cryoglobulins and toxicology screen were negative. Skin biopsy demonstrated intravascular thrombi (Fig 3). Special stains for organisms were negative. Additional infectious workup, including blood and skin tissue cultures, was negative.

Question 1: What is the most likely diagnosis?

- A. Angioinvasive fungal infection
- B. Disseminated intravascular coagulation (DIC)
- C. Levamisole-induced vasculopathy
- D. Mixed cryoglobulinemia (type II/III)
- E. Rocky Mountain spotted fever

Answers:

A. Angioinvasive fungal infection – Incorrect. Extensive infectious workup was negative, organisms were not evident on histology, and tissue cultures were sterile.

B. Disseminated intravascular coagulation (DIC) – Incorrect. While microangiopathic hemolytic anemia, rapidly progressing organ failure, and palpable purpura are characteristic of DIC, localization to acral regions, a normal D-dimer, and elevated (rather than decreased) fibrinogen levels are less consistent with this diagnosis.

C. Levamisole-induced vasculopathy – Incorrect. Ingestion of cocaine contaminated with levamisole may result in acral palpable purpura, most commonly on the face and ears. On skin biopsy, intravascular thrombi are common and features of leukocytoclastic vasculitis may also be observed. In this case, the patient denied use of illicit drugs, and toxicology screen was negative.

D. Mixed cryoglobulinemia (type II/III) – Incorrect. Mixed cryoglobulinemia may present with acral purpura due to small and medium vasculitis instead of intravascular occlusion, which was observed on skin biopsy in this case. Of note, type I cryoglobulinemia occurs in the setting of protein-secreting monoclonal gammopathies and is characterized by intraluminal obstruction rather than vasculitic changes.

E. Rocky Mountain spotted fever (RMSF) – Correct. Palpable purpura on acral sites with rapidly progressing multiorgan involvement is characteristic of RMSF. RMSF is transmitted by ticks, which require at least 4–6 hours of attachment before they infect the victim. Nevertheless, up to 40% of patients do not

report tick exposure at the time of presentation, in part because the bites typically cause no symptoms.¹

Question 2: Which of the following is the most appropriate next step?

- A. Initiate treatment with doxycycline
- B. Confirm diagnosis with serologic rickettsial antibody titers
- C. Obtain additional skin sample for direct immunofluorescence
- D. Contact the local or state health department
- E. Order contact precautions to avoid person-to-person transmission

Answers:

A. Initiate treatment with doxycycline – Correct. After an incubation period of 2–14 days, symptoms of headache, fever, myalgias, and arthralgias occur. Cutaneous involvement typically develops 3–5 days later. Because antibodies are not detectable until 7 to 10 days after disease onset, patients with suspected RMSF should receive empiric doxycycline, since treatment delayed after day 5 is associated with an increased mortality rate.²

B. Confirm diagnosis with serologic rickettsial antibody titers – Incorrect. Diagnosis can be confirmed by a four-fold rise in serum IgG antibodies drawn 2–4 weeks apart, but empiric therapy should be started immediately in patients with suspected RMSF. *Rickettsia rickettsii* IgG returned elevated at 1:128 5 days after hospitalization and 1:256 2 weeks later. The patient had no previous baseline serologic rickettsial antibody. Therefore, the elevated titers were not diagnostic, but strongly supportive of RMSF in conjunction with the patient's history and clinical findings.³

C. Obtain additional skin sample for direct immunofluorescence – Incorrect. Direct immunofluorescence staining via skin biopsy may be supportive diagnostically, but appropriate treatment should be initiated first. Of note, sensitivity of direct immunofluorescence declines after doxycycline therapy is initiated.⁴

D. Contact the local or state health department — Incorrect. All cases of RMSF should be reported to state or local health department according to local laws, but reporting should take place after appropriate therapy is initiated.

E. Order contact precautions to avoid person-to-person transmission — Incorrect. RMSF is transmitted via tick bite. Person-to-person transmission does not occur.

Question 3: Of the following, which 3 ticks are vectors for transmission of RMSF in the United States?

- A.** *Amblyomma aureollatum* (yellow dog tick)
- B.** *Amblyomma cajennense* (Cayenne tick)
- C.** *Dermacentor andersoni* (Rocky mountain wood tick)
- D.** *Dermacentor variabilis* (American dog tick)
- E.** *Rhipicephallus sanuineus* (brown dog tick)

Answers:

A. *Amblyomma aureollatum* (yellow dog tick) — Incorrect. The yellow dog tick is a vector for RMSF transmission in Brazil.⁵

B. *Amblyomma cajennense* (Cayenne tick) — Incorrect. The Cayenne tick is a vector for RMSF transmission in Central and South America.⁵

C. *Dermacentor andersoni* (Rocky mountain wood tick) — Correct. The Rocky mountain wood

tick is the primary vector for RMSF transmission in the Rocky Mountain region of the United States.⁵

D. *Dermacentor variabilis* (American dog tick) — Correct. The American dog tick is the primary vector for RMSF transmission in the eastern and southern central United States.⁵

E. *Rhipicephallus sanuineus* (brown dog tick) — Correct. The brown dog tick is a vector for RMSF transmission in the Southwest region.⁵

Abbreviations used:

DIC: disseminated intravascular coagulation
RMSF: Rocky Mountain spotted fever

Conflicts of interest

None disclosed.

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