

## A rapidly progressive painful skin lesion

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A 48-year-old woman presented to our emergency department with fever and a severe painful erythematous and ecchymotic skin lesion on the third finger of the left hand, that rapidly evolved into haemorrhagic bullae with diffuse swelling and edema of all her left hand, associated with the onset of lymphangitis in her left arm and crescendo-like pain. She denied trauma or insect bite. Her past medical history was unremarkable, except for chilblains with no sequelae. She did not take any medication. She was not a smoker. Laboratory tests showed neutrophilic leucocytosis (WBC 15,000/mm³; N 14,500/mm³), a slight thrombocytopenia (139,000/mm³), elevated C-reactive protein (12 mg/dL, normal value <0.5), increased procalcitonin (1.3 ng/mL, normal value <0.5) and transaminases (AST 64 U/L; ALT 70 U/L, normal value 0-31). HIV was excluded.

## Question

Given the patient's history and her skin lesions, what is the most likely diagnosis?

- 1. Raynaud's ulcer
- 2. Pyoderma gangrenosum
- 3. Necrotizing fasciitis
- 4. Scleroderma skin ulcer

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## **Answer**

Necrotizing Fasciitis (NF) is the right answer. The patient was immediately treated with parenteral empiric broad-spectrum antibiotics (piperacillin/tazobactam and clindamycin) and surgical debridement. Blood and tissue culture grew methicillin-sensitive Streptococcus pyogenes.

NF is a rare life-threatening bacterial soft tissue infection that rapidly spreads along soft tissue planes, causing progressive destruction of the muscle fascia and subcutaneous fat.1 Prompt diagnosis and timely treatment are crucial to avoid devastating consequences, including limb loss, organ damage and death. Patients with comorbidities such as immunosuppression, malignancy, vascular disease, diabetes, alcoholism, and obesity are at an increased risk of NF with poor prognosis.<sup>2</sup> Diagnosis is made clinically with the presence of skin discoloration, bullae, palpable crepitus, and pain disproportionate to the clinical findings. Fever is sometimes present. Elevated concentrations of CK or AST suggest deep tissue infection. Confirmatory diagnosis is based upon a culture and Gram stain of specimens collected from deep tissue, or by positive blood cultures. NF is classified into two types: polymicrobial (type 1) and monomicrobial (type 2) NF.<sup>3</sup> Type 1 NF is more common in older patients with risk factors, and typically located at the trunk, abdominal wall, perianal and groin areas, and in postoperative wounds. Type 2 NF is commonly caused by invasive Group A Streptococci (GAS)-pyogenesis in healthy immunocompetent patients, and usually located in the head, neck, or extremities. Initial superficial injury may be undetectable and severe signs and symptoms may not manifest until the underlying tissue damage has progressed extensively, as in our case. Plain X-ray is not recommended as an initial or definitive imaging study for NF. CT and MRI may show edema extending along the fascial plane, although these findings may be absent in early stages of NF.4 Surgical intervention should not be delayed in order to perform diagnostic imaging. Surgery is the gold standard treatment: exploration and debridement of the affected tissue should be performed promptly, and subsequent debridement should be continued daily until all necrotic tissue has been removed.<sup>5,6</sup> Antibiotic treatment is mandatory to support the surgical therapy. The most recent guidelines recommend vancomycin or linezolid in combination with piperacillin-tazobactam, a carbapenem, or ceftriaxone-metronidazole. Clindamycin should also be included in empiric therapy due to its effect on toxins released by S. aureus and GAS.5 The role of hyperbaric oxygen therapy and I.V. immunoglobulin G remains controversial.7

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