

Abstract:

A 17-year-old male adolescent presents to the emergency department with recurrent ulcerative skin lesions of the extremities, as well as weight loss, hematochezia, and postprandial abdominal pain. As diagnosis and treatment of dermatologic lesions are mainstays of pediatric emergency medicine, his initial presentation and broad differential provide valuable teaching lessons regarding the etiology and workup of ulcerative skin lesions. Following multiple long hospitalizations and extensive testing, the ultimate diagnosis was eventually revealed through attention to subtle details in the patient's history and presentation. A keen eye for similar presentations has the potential to provide future patients with less extensive workups, minimizing invasive patient testing and saving significant health care dollars.

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Keywords:

ulcerative skin lesions; factitious disorder; Munchausen syndrome

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Funky Rash in Nashville

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A 17-year-old male adolescent with a history of Asperger syndrome and seizure disorder presented to the emergency department (ED) with new lesions to his left knee. About 1 year ago, he first developed recurrent abscesses, the most recent of which was 1 week ago. At that time, he was seen at an outside hospital, where an abscess of the left knee was incised and drained and he was admitted for intravenous antibiotics. Records from the other hospital were not available, but his father recalled that the patient was discharged on clindamycin (300 mg every 8 hours) and doxycycline (100 mg twice daily). Despite reported compliance to this antibiotic regimen, he developed a second lesion, also to his left knee, on the day of presentation to the ED.

Both the patient and his father described the lesions as always evolving the same way. They began as “red bumps” that quickly became swollen and turned dusky. They then developed a raised dark purple area that “burst” with blood and pus. The patient was not sure how many lesions had developed over the past year, but he did state that his previous lesions were located on the left arm and left knee.

On review of systems, he endorsed a 12 to 15-lb unintentional weight loss for the past 2 months, with accompanying loss of appetite and night sweats. He reported intermittent low-grade subjective fever but had not noticed a particular pattern to the fever. He had also experienced alternating diarrhea and constipation during this period, including episodes of hematochezia. He described diffuse abdominal pain, which seemed to be worse after meals.

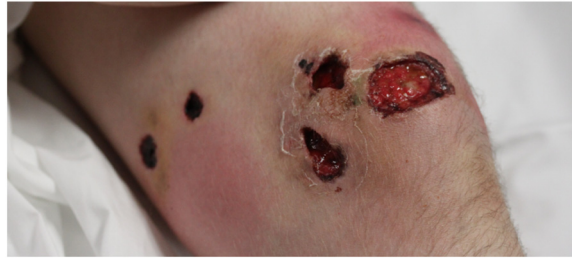


Figure 1 Appearance of left knee on initial presentation.

On examination, he was anxious appearing and mildly tachycardic with a heart rate of 110 beats per minute, respiratory rate of 22 breaths per minute, blood pressure of 125/77 mm Hg, temperature 36.8°C, oxygen saturation of 98% on room air, and a weight of 86 kg. Despite his tachycardia, he had a regular rhythm, with no murmur and 2+ pulses. His abdominal examination was benign, without tenderness or distension. He had full range of motion of his extremities but had difficulty ambulating without support due to left leg pain. His left knee was notable for erythema and fluctuance to both the medial and lateral aspects of his patella. He had 3 ulcerated open wounds with granulation tissue overlying his patella and 2 smaller open wounds with granulation tissue on the medial aspect of the patella. No foul odor or purulent drainage was present on examination (Figure 1).

In the ED, a complete blood count, erythrocyte sedimentation rate, C-reactive protein, and blood culture were obtained, as well as bacterial, acid-fast bacilli, and fungal cultures of the wound site. The complete blood count was significant for a white blood cell count of $21.7 \times 10^3/\mu\text{L}$, hemoglobin of 15.1 g/dL, mean corpuscular volume of 43 fL/red cell, and platelet count of 387 000/ μL , with a differential of 83.0% neutrophils, 7.8% lymphocytes, 6.7% monocytes, 0.9% eosinophils, and 0.2% basophils. The erythrocyte sedimentation rate was within normal limits at 15 mm/h (reference, 0-15 mm/h), and the C-reactive protein was elevated at 46.7 mg/L (reference, <10 mg/L). A radiograph of the knee was obtained, which demonstrated soft tissue defects consistent with his skin lesions but no underlying bony abnormality and no fractures or dislocations. He was admitted to the hospital on an increased dose of intravenous clindamycin.

While admitted, he underwent an extensive workup, with the ultimate diagnosis obtained over 1 month after his initial presentation to the ED.

DIFFERENTIAL DIAGNOSIS

Particularly in the ED, skin lesions in the febrile patient often suggest an infectious etiology. Typical bacterial causes, such as methicillin-sensitive *Staphylococcus aureus*, methicillin-resistant *S aureus*, and group A *Streptococcus* are routine pathogens to consider in the setting of presumed cellulitis and usually respond to empiric antibiotic coverage.¹ In cases of chronic lesions not improving on typical antimicrobial coverage, physicians should consider fungal etiologies, such as blastomycosis, histoplasmosis, as well as atypical mycobacteria.² Broadening coverage to include fungal pathogens, as well as obtaining fungal and bacterial cultures, can aid in the diagnosis of chronic lesions.

In the case of ulcerative lesions, more atypical pathogens, such as *Pseudomonas*, should be considered. *Pseudomonas* in the form of ecthyma gangrenosum can present as ulcerative lesions.³ These lesions typically progress over the course of 24 hours from an erythematous nodule or vesicle to an indurated ulcer with central necrosis followed by a black eschar.³ Although it can occur anywhere on the body, ecthyma gangrenosum most commonly occurs on the buttocks and extremities and rarely on the face.³ Prompt coverage of these lesions should be considered, as *Pseudomonas aeruginosa* is classically associated with septicemia.³ However, new evidence suggests that ecthyma gangrenosum may actually be caused by a variety of bacteria and fungi, sometimes without bacteremia.⁴

In patients with ulcerative lesions isolated to extremities, pyoderma gangrenosum should be considered. Pyoderma gangrenosum is a sterile neutrophil disorder that is relatively rare in children, accounting for <4% of all cases.⁵ These lesions are typically painful, starting as erythematous pustules or nodules that enlarge concentrically to form deep ulcerations with necrotic plaques.⁵ In adults, these lesions are classically isolated to the lower extremities.⁶ Recent evidence suggests that this is also the most common location for isolated pyoderma gangrenosum in children but that childhood disease overall is more commonly associated with disseminated ulcerations.⁶ Pyoderma gangrenosum can be associated with a variety of conditions, such as IgA monoclonal gammaglobulinopathy, specific forms of arthritis, inflammatory bowel disease, and PAPA (pyogenic arthritis, pyoderma gangrenosum, and acne) syndrome.⁶ Those with IgA monoclonal gammaglobulinopathy would be expected to present with hematologic disorders, whereas those with PAPA syndrome manifest their autosomal dominant disorder with sterile arthritis and acne.⁷ Most classically and particularly in patients with abdominal pain, inflammatory bowel disease

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