

Pyoderma Gangrenosum: Case Presentation and Review of Literature

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Abstract

Case Presentation: 80 y-o male who presented to the burn unit for management of a non-healing wound to right hand/ and forearm X 1 month. PMH was significant for leukemia, and the patient gave a history of having had chemotherapeutic infusion through a peripheral intravenous site on the affected extremity prior to onset of symptoms. He underwent surgical debridement twice at an outside hospital for presumed chemical burn from infiltration of chemotherapeutic agent.

Presentation to Burn Unit : He subsequently presented to the burn center stating that his wound was worsening over the past month, and he could not tolerate the wound care due to severe pain. Pt was septic and hypotensive with necrotic open wound to R arm/ forearm and leukocytosis (Figure 1). He was taken emergently to the operating room for debridement and source control, and was admitted to the ICU for further management including local wound care and antibiotic treatment (Figure 2). Punch biopsy was obtained which was consistent with the diagnosis of pyoderma gangrenosum.

Treatment and Hospital Course: System steroid treatment as well as local wound care and topical steroids were initiated. Pts pain was managed with a multimodal approach including local blocks as well as ketamine and pt was able to tolerate local wound care. Given the large open wound, and location over his dominant hand and forearm, pt was subsequently taken to the OR for split thickness autograft application.

Pathophysiology / Diagnosis/ Treatment: Pyoderma gangrenosum is a rare idiopathic neutrophilic dermatosis first described in 1930 by Brunsting et al (1). The diagnosis is often difficult to ascertain as there are no specific laboratory or histopathologic findings, and it remains a diagnosis of exclusion (2). The disease typically affects patients in the 3rd-6th decade of life and is often associated with systemic diseases such as ulcerative colitis, irritable bowel syndrome, and leukemia (greater than 50% of cases are associated with underlying malignancy (3,4)).

Pathophysiology: The pathophysiology is not clear, although it is believed to involve loss of the innate immune regulation and altered neutrophil chemotaxis. Increasing reports suggest cytokines such as IL-8 and TNF alpha (involved in neutrophil chemokine signaling) are dysregulated.

Diagnosis: Diagnosis remains difficult without specific serologic or histologic markers, however important aspects include: histologic findings of neutrophil predominance, severe pain associated with chronic non-healing ulcers, hx of prior trauma in affected area, presence of systemic disease of auto-inflammation , and underlying malignancy.

Treatment: Treatment is based on suppression the acute inflammatory response, and is aimed at systemic use of corticosteroids as well as local wound care and use of topical steroids (1). Other immune-modulating agents such as IVIG. Additionally, in the presence of infection, antibiotics such as minocycline and other tetracycline's can be used as they additionally inhibit neutrophil chemotaxis (1). Surgery Surgical treatment of pyoderma gangrenosum has historically been controversial as 30-50% of lesions demonstrate pathergy or worsening of the lesions with trauma. Additionally, concern that donor sites from split thickness autograft may be susceptible to the disease has made surgical therapy local wound care a mainstay of treatment. Local wound care, however, is often limited by the severe pain associated with the disease and lack of tolerance to local wound care, leading to infection and chronic non-healing wounds. Additionally, if the overall TBSA is large, and located in anatomic areas of functional importance, split thickness autografting should be given consideration. In the patient presented, overall TBSA was 4%, and the location was over the patients R hand and R forearm—thus decision was made to proceed with split thickness autograft application after initial source control and treatment with systemic and topical steroids.

Summary: Pyoderma gangrenosum is a rare neutrophilic dermatitis, without clear diagnostic / serologic/ or microscopic criteria. Diagnosis should be considered in the setting of chronic non-healing wounds marked by significant pain and in the setting of systemic inflammatory disease and malignant processes. Finally, surgical treatment is not contra-indicated, and patients may greatly benefit in closure of the wound and skin grafting procedures

Learning Objectives

1. Describe pyoderma gangrenosum as a disease entity
2. Understand the clinical presentation of the disease as well as the treatment modalities currently available

References:

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Figure 1. Non-healing wound of R hand/ R forearm not responsive to local wound care.



Figure 2. R forearm/ R hand wound after initial debridement/ source control.