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Recognizing Pyoderma Gangrenosum in a Patient with History of Essential Thrombocytosis

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Abstract

We describe a 62-year old man diagnosed with pyoderma gangrenosum following a traumatic injury to the hand. The patient was originally misdiagnosed with cellulitis and his condition did not improve with oral or intravenous antibiotics. The wound dramatically improved following institution of intravenous steroids. Pyoderma gangrenosum should be considered in the differential of a non-healing cellulitis following traumatic injury.

Keywords

Pyoderma gangrenosum; essential thrombocythemia; essential thrombocytosis; cellulitis

Abbreviations

PG: Pyoderma Gangrenosum

Introduction

Pyoderma gangrenosum (PG) is an ulcerating neutrophilic dermatosis commonly affecting the lower extremities and presenting in the second to sixth decades of life [1-2]. It is often a diagnosis of exclusion, and more than half of presenting cases are associated with an underlying systemic illness [2]. A variety of other conditions can present similarly including infection, vascular insufficiency ulcers, cancer, and autoimmune disorders [2]. A high degree of suspicion for PG should be maintained for chronic wounds that do not respond to standard treatments.

Case Presentation

A 62-year-old white male presented to the hospital with fever and a worsening wound on the left hand. The initial wound started after a trauma to the hand from a dump truck engine two weeks prior. He described the initial lesion as a blister which had progressively enlarged and reddened. His past medical history included seizure disorder, complicated diverticulitis requiring surgical intervention, and essential thrombocytosis treated with hydroxyurea in the past. His home medications consisted of levetiracetam, omeprazole, and sildenafil. He rarely smoked and consumed alcohol socially. He had been married to his wife for over 30 years and denied recent travel or a history of intravenous drug use. He was initially admitted for sepsis syndrome. On admission, he was a febrile with a blood pressure of 135/70 mmHg. He had an elevated white blood cell count of 18 x 10^9cells/L. He was prescribed intravenous antibiotics for suspected cellulitis, and was subsequently discharged home to continue intravenous

therapy. Blood and wound cultures from that admission remained negative.

He presented again four days after discharge due to persistent pain in his hand. It had become increasingly painful to touch, warm, red, with mild purulent discharge. On exam, he was febrile at 101.4°F (38.6°C) with a blood pressure of 163/88 mmHg. Covering the majority of the dorsum of his left hand was an erythematous plaque with ulcerations and crusting (Figure 1). There was edema of the hand with severe tenderness. Range of motion, sensation, and capillary refill was normal distally. The remainder of the physical exam was unremarkable. Laboratory values were significant for an elevated white blood cell count of $31x10^9$cells/L, platelet count of $271x\ 10^9$/L, and ESR 77 mm/hr. Magnetic resonance imaging did not reveal any signs of tendon involvement or osteomyelitis. Broad spectrum antibiotics were started. Vancomycin, meropenem, and oral trimethoprim/sulfamethoxazole were chosen to cover for non-tuberculosis mycobacteria, *Nocardia*, and conventional bacterial pathogens (including *Staphylococcus aureus* and *Pseudomonas*).

Given the intense pain and failure to improve with antibiotic therapy, alternative etiologies were entertained. Dermatology was consulted for biopsy which revealed pyoderma. His antibiotics were discontinued and he was started on pulse dose intravenous steroids with improvement in symptoms. He was diagnosed with PG. Over the course of three inpatient days his leukocytosis and fever resolved, and his wound improved (Figure 2A). He was discharged on a steroid taper and follow-up revealed a well-healing wound and normalized inflammatory markers. His ulcer continued to improve at one week (Figure 2B) and three weeks (Figure 2C) following discharge.

Discussion

Pyoderma gangrenosum is a cutaneous disorder marked by neutrophilic infiltrate on biopsy. The typical presentation is that of a papular or pustular lesion involving the lower extremity [1]. The appearance of PG can be subdivided into five subtypes—ulcerative, vegetative, bullous, pustular, and peristomal [2]. The ulcerative form is the classic presentation of PG, and presents as a single or a few small pustules that quickly ulcerate [2]. Hematological disturbances are most commonly associated with the bullous form, which present as rapidly spreading, superficial bullae that break down to form ulcers [2]. Ulcers typically start in locations of trauma, a phenomenon known as pathergy. The diagnosis is a clinical one; taking into account clinical and histologic findings, exclusion of other etiologies such as infection and malignancy, and is verified by a prompt response to steroids.

PG has been associated with a variety of systemic disease such as inflammatory bowel disease, hematological malignancies, and rheumatologic conditions (Table 1) [1,2]. Essential thrombocytosis (ET) has been associated with PG in few case reports, and does not appear to be a marker of disease activity [1,3,4]. Often times, PG does not fully resolve without specific treatment, even if the underlying associated disease has been controlled. Hydroxyurea, which is commonly used in the management of essential thrombocytosis, has been associated with apyoderma-like ulceration that commonly presents on the lateral malleoli of the feet [5]. This particular complication responds to discontinuation of the drug, versus immunosuppressive therapy, which is the mainstay of treatment for PG [5]. Our case highlights a patient with no history of preexisting PG, but a long-standing diagnosis of ET. His sudden development of a non-healing ulcer following trauma was not associated with pathergy given the poorly known association between ET and PG.

Localized or mild pyoderma gangrenosum is treated with topical corticosteroids or topical tacrolimus [6]. Systemic treatment is indicated for those with more aggressive disease or mild disease with failure of local therapies. This includes systemic glucocorticoids, dapsone, cyclosporine, tacrolimus, and azathioprine [6]. Infliximab is especially effective when PG is associated with underling inflammatory bowel disease [6]. Surgical debridement should be avoided secondary to pathergy.

Although pyoderma gangrenosum is classically associated with inflammatory bowel disease, being mindful of less well known associations is useful in patient with non-healing ulcerated wounds. This is especially helpful when these lesions present in atypical locations, such as the hand. Prompt recognition and treatment of pyoderma gangrenosum is the most vital step to prevent further manipulation of the wound.

Table

Frequency of Association	Associated Conditions
Common	Inflammatory bowel disease Ulcerative Colitis Crohns disease
Less common	Hematologic malignancy Leukemia Lymphoma Myeloproliferative disorders Polycythemia vera Essential Thrombocytosis Chronic myeloid leukemia Myelofibrosis Myelodysplastic syndrome Paraproteinemia Monoclonal gammopathy Multiple myeloma Arthritis Rheumatoid arthritis Ankylosing spondylitis Seronegative non-IBD arthritis
Rare	Sarcoidosis Systemic Lupus Erythematosis Behcet's Hidradenitis Suppurativa Autoimmune hepatitis

Figures

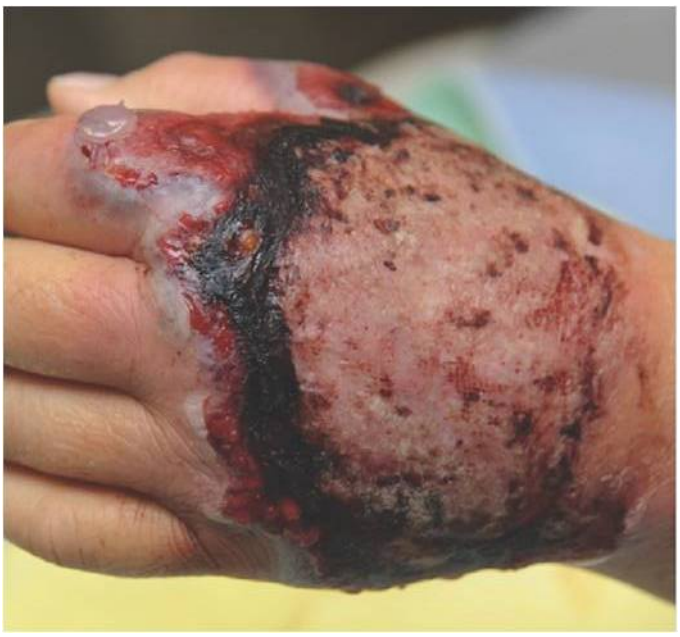

Figure 1: Non-healing wound on dorsum of left hand

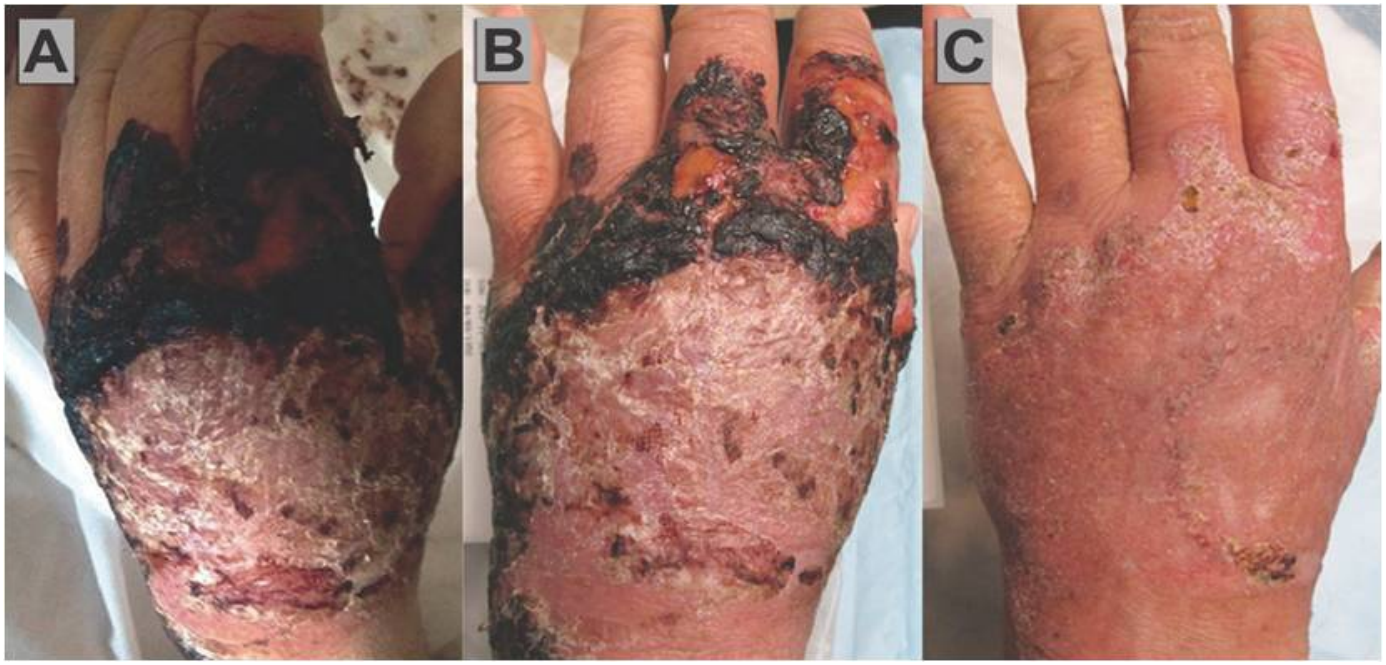

Figure 2: Improvement in pyoderma gangrenosum following three days of intravenous steroids (A), one week of oral steroids (B), and three weeks of oral steroids (C).

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