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Pyoderma gangrenosum presenting as bilateral leg and foot ulcers

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Abstract

Pyoderma Gangrenosum (PG) is a rare disease on the neutrophilic dermatosis disease spectrum. It most commonly occurs as a secondary complication of diseases such as gastrointestinal, rheumatologic, autoimmune and malignancies. We present the case of a 67-year-old woman who presented to the Emergency Department with complaints of extremely painful bilateral leg ulcers which rapidly progressed in size, fever, abdominal pain, and bloody diarrhea for 4 days.She had a past medical history significant for a longstanding history of colitis which was eventually diagnosed as Crohn's disease (CD), hypertension, and uncontrolled diabetes mellitus. Given her history of CD and her presenting symptoms,she was likely experiencing exacerbation of CD and the suspicion of the ulcers being PG, although rare, was very high. Diabetic foot ulcers were also on the differential. Biopsy and histological examination confirmed the ulcers to be PG. The diagnosis of PG is one of exclusion. Good history, physical exam, biopsy and histological examination, along with high clinician suspicion are the hallmarks of the diagnosis. Initiation of steroids should be done based on the suspicion even before obtaining a definitive diagnosis to decrease its morbidity.

Keywords

pyoderma gangrenosum; ulcers; crohn's disease; ulcerative colitis; inflammatory bowel disease

Introduction

Pyoderma Gangrenosum is a rare disease that can present as a secondary manifestation of a primary gastrointestinal disease such as inflammatory bowel disease (IBD), although it can also occur as a complication of rheumatologic diseases, autoimmune diseases, such as connective tissue disorders, and hematological malignancies [1,2]. This disease entity was first described in 1916 by Louis-Anne-Jean Brocq, a French dermatologist [3]. It can cause large, painful ulcers to develop on the skin but can also involve underlying soft tissues structures. It is an uncommon neutrophilic dermatosis but in the current literature it is still considered a part of a neutrophilic dermatosis disease spectrum [3,4,5]. The exact incidence for its occurrence is unknown. It is estimated to occur in 3 to 10 cases per million people worldwide yearly [3].

Case Report

A 67-year-old woman was admitted to the hospital in January 2017, with complaints of extremely painful bilateral leg ulcers which rapidly progressed in size over a period of 2 months, fever, abdominal pain, and bloody diarrhea for 4 days. She had a past medical history significant for a long standing history of colitis which was eventually diagnosed as Crohn's disease, hypertension and uncontrolled diabetes mellitus. The ulcers were worst on the left leg and initially thought to be diabetic ulcers given her uncontrolled diabetes mellitus.

On physical examination, the skin was warm, dry and tender to palpation. On the left leg, she had a partial thickness ulcer which was asymmetric with inflamed borders measuring 10cm x 5cm on the anterior left distal tibia (Figure 1, 2) and 5cm x 4cm on the anteromedial right distal tibia. There was no purulent drainage, no fluctuance, no crepitus, and no probe to bone. Her initial laboratory examination was significant for HbA1c of 8.0%, elevated erythrocyte sedimentation rate (ESR) at 110 mm/hr (normal value 0 to 30 mm/hr), elevated C-reactive protein (CRP) at 127.3 mg/L (normal value <10 mg/L) and acute kidney injury (AKI) with creatinine of 1.37 mg/dL (baseline <1.1 mg/dL).Her hemoglobin remained stable at her baseline despite her having bloody diarrhea. Stool polymerase chain reaction (PCR) was not obtained as the patient had formed stools in the form of diarrhea.

Imaging studies during her hospitalization included a plain radiograph of her lower extremities as well as a MRI of her lower extremity joints. Radiographs showed focal skin irregularity at the lateral aspect of the distal left shin and anterior mid aspect of the right shin without underlying evidence of osteomyelitis. She also underwent a MRI which was negative for osteomyelitis. She was initially started on broad spectrum antibiotics with vancomycin & piperacillin-tazobactamas she was febrile and the ulcers were thought to be of a secondary infectious etiology. Over the preceding day, her condition did not improve and in fact, her abdominal pain and the pain around the ulcer site increased in intensity despite receiving morphine for pain. We suspected that the patient was in fact in a Crohn's flare up and the lower extremity ulcers may in fact be PG so, based on this suspicion, we initiated intravenous methylprednisolone. Antibiotics were discontinued as she was no longer febrile and all cultures, including blood cultures and wound cultures, were negative in the final read.

The patient's lesion was further subjected to biopsy and histological examination. It showed skin ulceration with perivascular and interstitial neutrophilic infiltrate in the superficial, deep dermis, and extending into the subcutaneous fat. It also showed perivascular lymphocytic infiltrate without fibrin deposition or mural necrosis. Given the history of Crohn's disease and the findings of the histological examination, a diagnosis of pyoderma gangrenosum (PG) was made. The steroids provided her significant pain relief and her diarrhea resolved. Over these preceding days of her hospitalization, both of her inflammatory marks, ESR & CRP trended down to the normal limits respectively stated above and her AKI, which was most likely secondary to dehydration due to diarrhea, resolved. She was started on subcutaneous insulin injections for her DM. The patient was discharged from the hospital in a much improved condition on a steroid taper with a referral to follow up with her Rheumatologist and Gastroenterologist, in anticipation to be started on steroid sparing drug modalities for her condition.

Discussion

Inflammatory bowel disease is characterized by chronic inflammatory changes and encompasses two main disease entities namely, Crohn's disease and Ulcerative Colitis (UC). Pyoderma gangrenosum is a relatively uncommon skin condition which is thought to have immune-mediated lesions which affects only about 0.4%-2% of the inflammatory bowel disease patient population [6]. Conversely, PG can also follow a course that is independent to that of the bowel disease [7]. It can even occur in patients with a history of long standing colitis of unknown etiology [7].

The PG ulcer lesions are comprised of different characteristics and there are 4 different subtypes in general – pustular, ulcerated, bullous, and vegetative [5, 6]. They usually start as a swelling or blister, which maybe single or in a group and can even occur at different anatomical locations [8]. The lesions are usually very painful and the healing process is a slow one. In particular, the edges of this specific type of ulcer are not well defined and the surrounding surface is usually erythematous. As in the patient described above, these PG ulcers are strongly related to flare up of IBD where in a flare up of bowel symptoms may exacerbate the growth and pain potential of these lesions [8]. Furthermore, the pathogenesis of PG is fascinating and starts at the molecular level. One of the purposed pathogenesis is that there is an abnormal immune response which triggers cross reaction of antibodies. These antibodies cross react with the common antigens which are found in bowel and skin and intensify the inflammatory response [9]. Additionally, in neutrophilic dermatoses there is over-expression of pro-inflammatory cytokines such as TNF-alpha, IL-8, IL-16, IL-17, and IL-23 along with neutrophil dysfunction and abnormal T-cell response [10].

In the above patient, the diagnosis of PG was very high on our differential diagnosis list, although it is nevertheless a rare complication of IBD and even more rare with just a history of long standing colitis. She had a history of CD and presented with bloody diarrhea and abdominal pain, which was likely an exacerbation of CD coupled with extremely painful lower extremity ulcers and elevated inflammatory markers such as ESR and CRP. Although she did have a history of uncontrolled DM and also had polyneuropathy, her lower extremity ulcers were most likely PG lesions, given that they were extremely painful and responded very well to intravenous steroids rather than antibiotics. Even though she was also started on broad spectrum antibiotics, they are of minute value in these ulcers unless there is an associated superimposed bacterial infection. Lastly, early biopsy of these ulcers helped confirm our clinical suspicion.

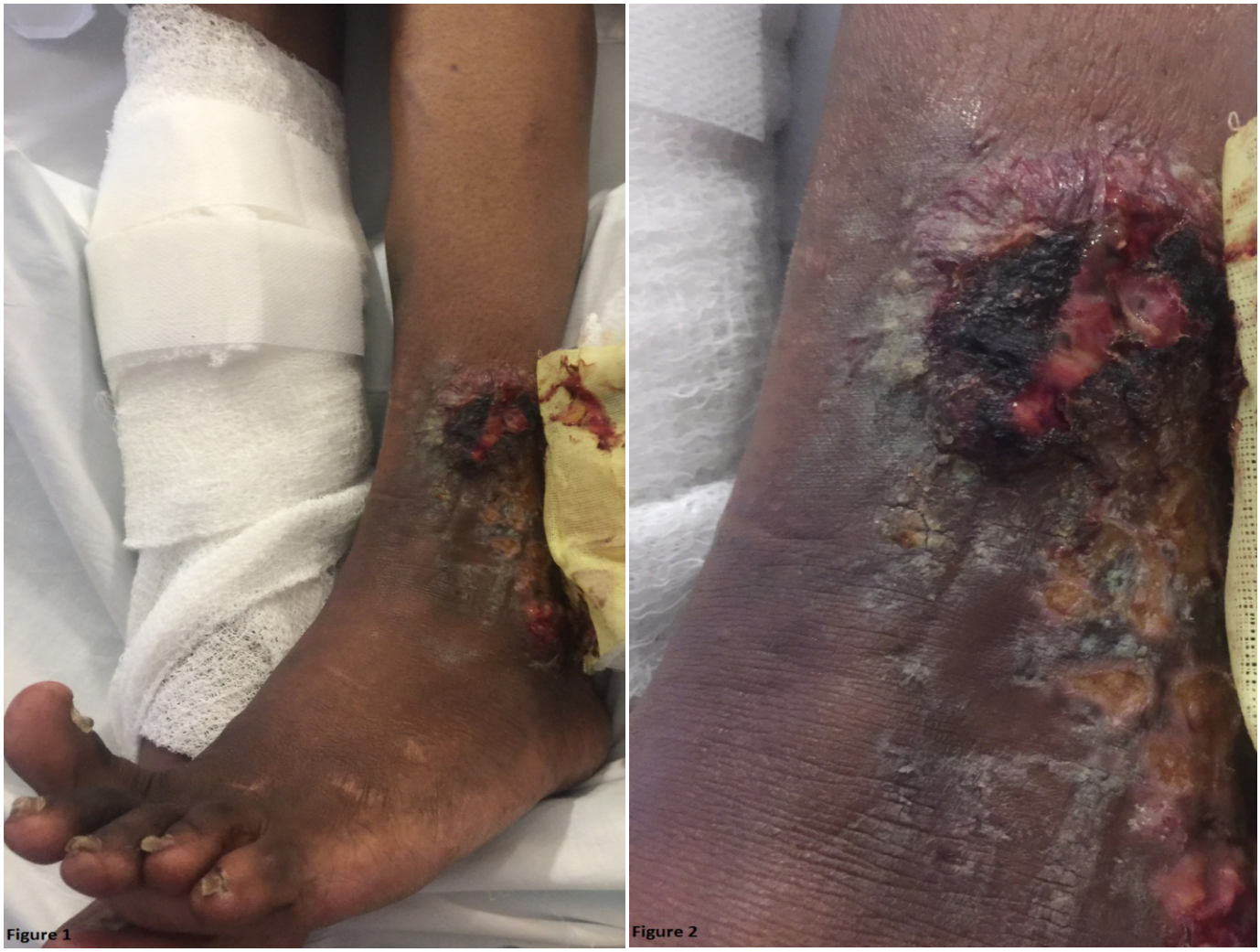
Treatment of these ulcerative lesions is non-specific. It is comprised of proper wound care and dressings, intravenous steroids in severe lesions with transition to oral steroids and steroid sparing drug modalities such as immunosuppressive or cytotoxic drugs. These lesions can be very severe and refractory to treatment despite the aforesaid treatment modalities resulting in considerable morbidity.

Conclusion

In conclusion, the diagnosis of PG is challenging as it is a diagnosis of exclusion. It can even occur in patients who merely have colitis of unknown etiology. Good clinical history, including the past medical and surgical history, physical examination and biopsy, along with a high suspicion, are the hallmarks for diagnosing PG. Clinicians should maintain a high level of suspicion and seek an early tissue diagnosis so as

to not miss this already under diagnosed disease entity, especially in patients with IBD or any form of long standing colitis.

Figures



Figures: Pyoderma Gangrenosum presenting as foot ulcers on the anterior left distal tibia; (1) superior and (2) close up views.

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