Case Report

Pyoderma Gangrenosum with Splenic Abscess- A Rare Association

Abstract

Pyoderma gangrenosum is a rare, chronic neutrophilic dermatosis of unknown etiology. The classical clinical feature of pyoderma gangrenosum is a pustule or plaque that rapidly progresses to a painful, necrotic ulcer with undermined violaceous margins. Pyoderma gangrenosum may be associated with underlying inflammatory bowel diseases, hematological malignancies, or rheumatologic disorders in 50–70% of the cases. The visceral involvement by pyoderma gangrenosum is rare. Sterile neutrophilic infiltrates in organs other than the skin are uncommon systemic manifestations of neutrophilic dermatoses, but have occasionally been reported. We report a case of a 38-year-old female with pyoderma gangrenosum and visceral involvement manifesting as splenic abscess.

Keywords: *Extracutaneous pyoderma gangrenosum, pyoderma gangrenosum, splenic abscess*

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Introduction

Pyoderma gangrenosum is a chronic inflammatory neutrophilic dermatosis originally described by Louis Brunsting *et al*.[1] in 1930 at the Mayo Clinic. It was incorrectly presumed to be an infectious disorder, and although the misnomer 'pyoderma' was never changed, pyoderma gangrenosum is now regarded as a result of immune dysfunction rather than infection. The classical clinical feature of pyoderma gangrenosum is a pustule or plaque that rapidly progresses to a painful, necrotic ulcer with undermined violaceous margins. The etiology is unknown, but more than 50% of the cases are frequently found in association with systemic diseases, mainly inflammatory bowel disease, arthritis, paraproteinemia, and hematological malignancy.[2] The visceral involvement by pyoderma gangrenosum in the form of sterile neutrophilic infiltrates in organs other than the skin has occasionally been reported.[2] These non-cutaneous findings are categorized into three subtypes: (1) systemic inflammation common to other diseases associated with neutrophilic dermatosis, such as gastrointestinal, hematologic, and rheumatologic conditions; (2) non-specific inflammatory findings such as myalgia, fever, and joint pain; (3) sterile neutrophilic infiltrates found in organs other than the skin.[3] The pulmonary involvement by these sterile neutrophilic infiltrates is most common followed by the eye and other organs. The spleen is a rare site of sterile abscesses found in association in patients with pyoderma gangrenosum. A total of 14 cases of pyoderma gangrenosum with splenic involvement have been reported to date with four cases affecting children.[4-17] We report a case of a 38-year-old female with pyoderma gangrenosum and extracutaneous manifestation presenting as asymptomatic splenic abscess.

Case Report

A 38-year-old female was admitted with an 8-month history of painful, non-healing ulcers over the right gluteal region and right lower leg, associated with malaise and low-grade fever off and on. Despite antibiotic therapy, no clinical improvement was achieved and the ulcers progressively increased in size. There was no significant medical history and the family history was non-contributory. A physical examination revealed a well-circumscribed, oval ulcer with violaceous, and undermined edges of size 8 cm^2 × 10 cm^2 over the right buttock. Its floor was covered with reddish granulation tissue and extensive purulent exudate. Adjacent to this toward the gluteal cleft, there was a smaller 2 cm^2 × 2 cm^2-sized oval ulcer with sharp undermined

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margins. Another similar ulcer was present near the ankle, measuring 4 cm^2 × 5 cm^2 [Figures 1a and b] These ulcers were markedly tender. There was no lymphadenopathy or organomegaly and the systemic examination was normal.

The laboratory studies revealed a total leucocyte count (TLC) of 15.5 × 10^9/L with neutrophilia (93.8%), hemoglobin of 10.1 gm/dL, C-reactive protein 75.14 mg/L, and erythrocyte sedimentation rate of 74 mm in the first hour. Serum ferritin was 120.5 ng/mL and anti-tissue transglutaminase A was 0.1 U/mL. The liver and renal function tests, urine routine analysis, and serum electrophoresis were normal. The stool for occult blood was negative and the chest X-ray did not reveal any abnormality. The anti-nuclear antibodies and anti-neutrophil cytoplasmic antibody profile were negative, while the serology for hepatitis C virus, hepatitis B virus, and human immunodeficiency virus (HIV) were non-reactive. The Mantoux test was negative. The ultrasound of the abdomen showed multiple cystic ill-defined hypoechoic areas in the mid and lower pole of the spleen, the largest measuring 5.4 cm × 3 cm with no internal vascularity, suggestive of splenic abscess [Figure 2]. The spleen measured 13.5 cm in span, with a normal outline and parenchymal echotexture. The splenic vein was normal. No abnormality in any other intraabdominal organ was detected on ultrasonography (USG). The arterial and venous Doppler study of the bilateral lower limbs did not reveal any evidence of deep vein thrombosis, arterial stenosis, or incompetent valves. The MRI of the pelvis revealed an ill-defined irregularity in the right superior gluteal region, with changes restricted to subcutaneous and gluteal fat, minimal extension to gluteal fat, and no evidence of the involvement of the underlying bones or musculature. The skin biopsy taken from the edge of the ulcer showed dense neutrophilic infiltration extending through the dermis with extensive areas of necrosis. Histiocytic collections and a few giant cells were also seen surrounding the dense neutrophilic infiltrate. The extension of the infiltrate into the subcutaneous tissue was also seen. There were no granulomas or atypical cells. The periodic acid schiff (PAS) stain for fungus and 20% Ziehl-Neelsen (ZN) stain for acid-fast bacilli were negative [Figures 3–5]. The cultures for aerobic and anaerobic bacteria, fungi, and mycobacteria obtained from the skin samples were negative. On clinco-pathological correlation, a diagnosis of pyoderma gangrenosum with splenic abscess was made. The patient was managed with oral cyclosporine 100 mg twice daily for 3 months and pulse intravenous methylprednisolone (750 mg), administered for 3 days, repeated at monthly intervals for 3 pulses. The ulcers were cleaned with normal saline and the dressing was done using topical antibiotics. A cross consultation with a gastroenterologist and a surgical specialist for the splenic abscess was taken and no active intervention was advised. The patient is on regular follow-up and the ulcers are healing [Figure 6a and b]. The abdominal ultrasound on the follow-up showed a complete resolution of the splenic abscess [Figure 7].

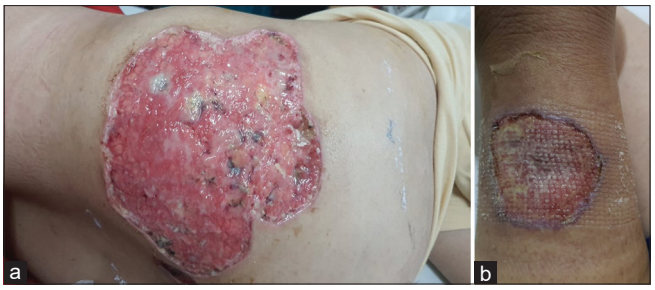

Figure 1: Well-circumscribed, oval ulcers with violaceous, undermined edges with the floor covered with reddish granulation tissue and extensive purulent exudate over the right buttock (a) and ankle (b)

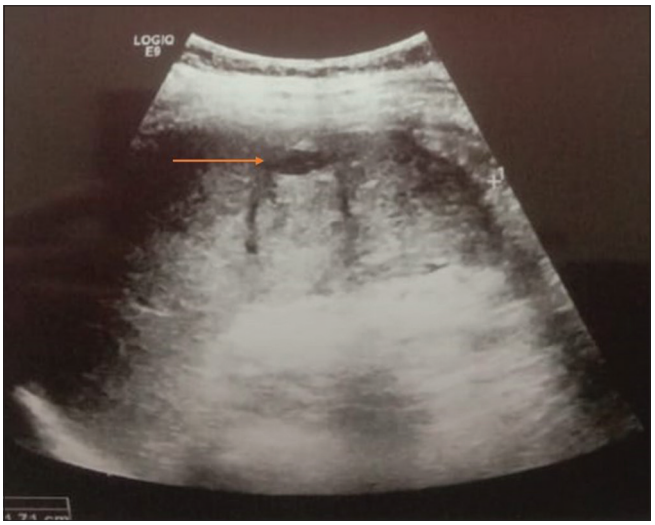

Figure 2: Ultrasound abdomen showing the cystic hypoechoic area (arrow) in the mid and lower pole of the spleen, suggestive of splenic abscess

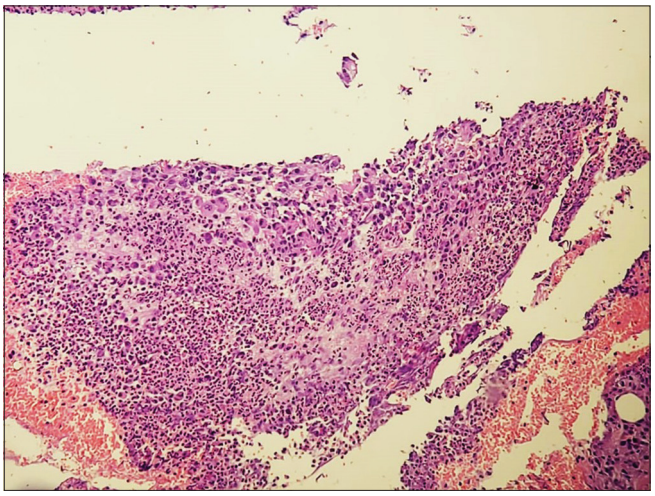

Figure 3: Biopsy from the ulcer showing dense neutrophilic infiltrates surrounded by mononuclear cell infiltrate comprising mainly of histiocytes along with the areas of hemorrhage (H&E, 100×)

Discussion

Pyoderma gangrenosum (PG) is a rare, chronic neutrophilic dermatosis of unknown etiology. The worldwide incidence is estimated to be around 3–10 cases per million population per year.[3] Although PG affects both sexes, a slight female

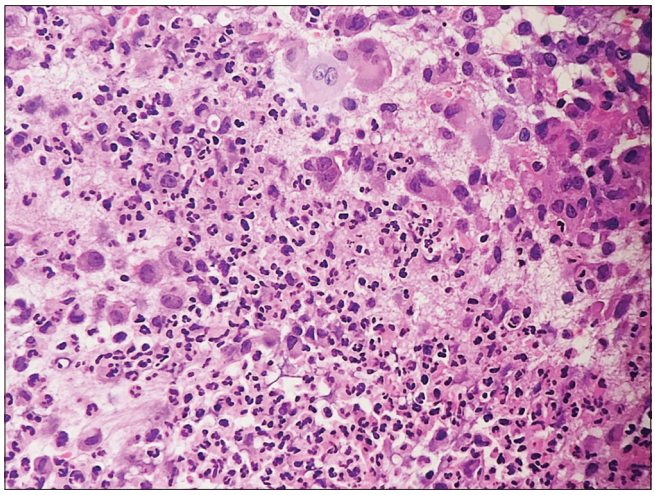

Figure 4: Higher magnification showing the presence of neutrophilic collections surrounded by many histiocytes and giant cells in the deeper dermis (H&E, 400×)

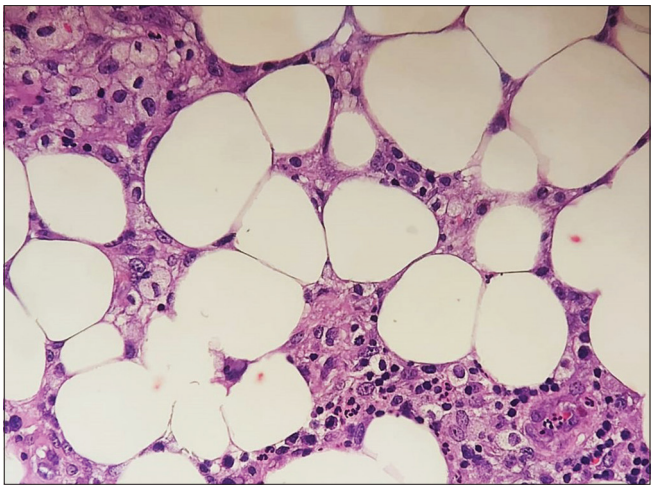

Figure 5: Photomicrograph showing neutrophilic infiltrate with histiocytes extending into the subcutaneous tissue (H&E, 400×)

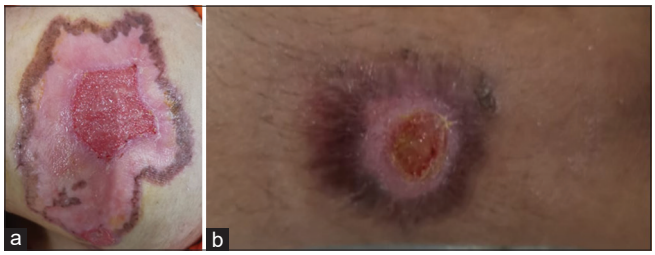

Figure 6: Post-treatment images—ulcers over the gluteal region (a) and ankle (b) showing healing at 3 months of follow-up

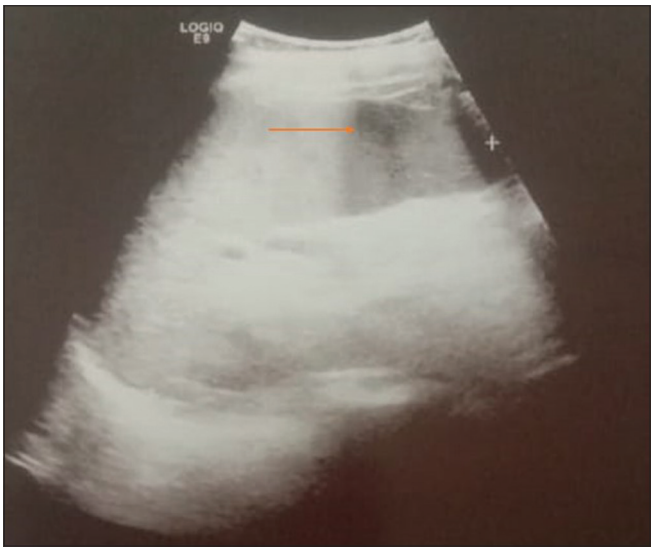

Figure 7: Abdominal ultrasound on the follow-up showing complete resolution of the splenic abscess (arrow)

predominance exists. All ages may be affected by the disease, but it predominantly occurs in the fourth and fifth decades of life. The pathogenesis of PG is multifactorial and involves neutrophilic dysfunction, inflammatory mediators, and genetic predisposition.[3] In 50–70% of the cases, inflammatory bowel diseases, hematological malignancies, or rheumatologic disorders are associated with PG.[2]

Additionally, patients with PG may have extracutaneous manifestations in the form of sterile neutrophilic infiltrates in various organs.[3] The culture-negative pulmonary infiltrates are the most common and potentially fatal extracutaneous manifestations.[2] Other organs systems that may be involved include the heart, the central nervous system, the gastrointestinal tract, eyes, liver, bones, and lymph nodes.[2] The splenic and renal involvement of PG is extremely rare.[2] A systematic review of the literature by Borda *et al*.[3] identified 96 cases of PG with extracutaneous manifestations. The pulmonary involvement was reported most frequently in 42.7% of these patients, followed by ocular involvement in 35.4%, bone involvement in 7.2%, and spleen and renal involvement in 6.2% and 3.1%, respectively. The less commonly involved organs were the brain, gastrointestinal, muscle, and heart in 5.2% of the cases.

The splenic involvement in the course of PG has been reported at all ages, from 22 to 73 years, corresponding to 10 adult cases reported in the literature.[4-13] There are four reports of pediatric PG with associated splenic involvement ranging from a 16-month child to 14 years of age.[14-17] It is slightly more frequent in males.[5-8,13-16] PG with splenic involvement can be associated with additional involvement of other organs including the liver, kidney, lung, and bone.[3] The skin manifestations can present before, during, and after splenic involvement. Three cases of PG with splenic involvement had associated disorders in the form of multiple myeloma,[5] immunoglobulin A (IgA) gammopathy,[7] and chronic monomyelocytic leukemia.[8] Splenic abscess in PG is asymptomatic in a majority of cases and only two cases have been reported with abdominal pain.[11,15] Laboratory abnormalities observed in these cases include leukocytosis with neutrophilia, anemia, raised erythrocyte sedimentation rate (ESR) and c-reactive protein (CRP).[4-17] The differential diagnoses for splenic lesions in PG include

vasculitis, tumor, and infections. The imaging studies aid in the diagnosis and follow-up for these patients. However, invasive diagnostic procedures are generally avoided due to concerns of pathergy.[3] Fine-needle aspiration of the spleen abscesses has been done in three cases in the literature, which yielded purulent fluid; both Gram and Ziehl–Neelsen stains were negative.[5,8,13]

The therapy for PG in patients with underlying visceral involvement involves the use of anti-inflammatory agents, including antibiotics, corticosteroids, immunosuppressive agents, and biologic agents.[4-17] In most cases, the disease responded well to glucocorticosteroids, whereas a few patients were managed with immunosuppressants like cyclosporine[8-13] and dapsone.[4] In refractory cases, biologicals such as infliximab, adalimumab, and etanercept were used.[13-14]

The prognosis is generally good; however, the disease can recur and residual scarring is common. The recurrence of PG in patients with splenic involvement has been reported in two patients.[4,14] One case was fatal following the development of bowel infarction of unknown etiology after the recurrence of pyoderma gangrenosum.[4]

PG is a diagnosis of exclusion. Our patient was thoroughly investigated to exclude any other cause of ulceration and associated disorders. The splenic abscess was an incidental finding on abdominal USG. The patient was afebrile during admission, with no other signs of sepsis, without any abdominal symptoms or signs. According to the organ involved, the relevant investigations to exclude out other causes like vasculitis, infective pathology are advised. The ulcers showed an excellent response to the corticosteroids and cyclosporine. The remission of the splenic abscess was also achieved with corticosteroid therapy. This case highlights that PG can be associated with visceral involvement. A systemic immunosuppressant is indicated in the management of such patients irrespective of the size of cutaneous ulcers.[3]

Conclusion

The visceral involvement of PG is rare, comprising non-infective sterile abscesses. The presence of these infiltrates in organs other than skin supports the idea of PG being a systemic disease. The management of the underlying PG results in the remission of the associated visceral abscesses.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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