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Ulceratice Pyoderma Gangrenosum With Aspectic Splenic Abscess - A Rare Association

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Abstract
Pyoderma gangrenosum (PG) is a rare, non-infectious, inflammatory, ulcerative skin disease that mainly involves the lower extremities and the exact etiology remains unknown. .The classical clinical feature of pyoderma gangrenosum is a pustule or plaque that rapidly progresses to a painful, necrotic ulcer with undermined violaceous margins. It is often associated with systemic diseases, including inflammatory bowel disease, rheumatoid arthritis, and hematological disorders in 50–70% of the cases . Sterile neutrophilic infiltrates in organs other than the skin are uncommon systemic manifestations of neutrophilic dermatoses, but have occasionally been reported .However, its association with aseptic splenic abscesses is extremely uncommon and underreported in the literature. We report a case of a 81-year-old patient with pyoderma gangrenosum and visceral involvement manifesting as splenic abscess. The diagnosis was confirmed based on clinical data, histological analyses , and medical imaging results Increased systemic corticosteroid therapy produced a successful outcome.

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

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I. Introduction

Pyoderma gangrenosum (PG) is a neutrophilic dermatosis characterized by chronic, non-infectious skin ulcers. Although the exact etiology remains unknown, more than 50% of cases are often associated with systemic diseases such as chronic inflammatory bowel disease, arthritis, paraproteinemia, and certain malignant hematological conditions.In rare instances, PG can affect internal organs in the form of sterile neutrophilic infiltrates, though these cases have been reported infrequently. These extracutaneous manifestations are classified into three subtypes: (1) systemic inflammation related to other diseases associated with neutrophilic dermatosis, such as gastrointestinal, hematological, and rheumatological conditions; (2) nonspecific inflammatory symptoms, such as myalgia, fever, and joint pain; (3) sterile neutrophilic infiltrates found in organs other than the skin.The lungs are the most commonly affected organ by these infiltrates, followed by the eyes and other organs. The spleen, although rarely, can also be the site of sterile abscesses in patients with pyoderma gangrenosum.

We report the case of an 81-year-old man with pyoderma gangrenosum and extracutaneous manifestations, presenting as an asymptomatic splenic abscess

II. Case Report

This is an 81-year-old hypertensive patient admitted for multiple ulcers on the lower limbs. The condition began three weeks prior with pustules on the right foot and both thighs, which gradually developed into painful ulcers, accompanied by a decline in general condition with unspecified fever.

The physical examination revealed multiple ulcers located on the lower third of the right leg and above both the right and left popliteal fossae. These ulcers varied in size, with fibrinous bases and granulating tissue in some areas. Their edges were elevated, violaceous, tender, and bled upon contact (Figure1).The laboratory tests revealed a total leukocyte count (TLC) of 15.5 × 10^9/L with neutrophilia at 85%, hemoglobin at 10 g/dL, a CRP level of 168 mg/L, and a polyclonal increase in gamma-globulins. The skin biopsy taken from the edge of the ulcer showed subepidermal bulla and neutrophil and mononuclear cell infiltration in the mid- to lower dermis . He was referred to our department for an abdominal CT scan as part of the paraneoplastic workup, revealed a spleen of normal size, with intrasplenic and subcapsular collections of varying sizes. These collections appeared hypodense with liquid density and showed peripheral enhancement after contrast injection, suggesting splenic abscesses with vesicular macrolithiasis without signs of complication (Figure2). An

additional abdominal-pelvic MRI also confirmed the splenic abscesses. Based on a clinico-pathological and radiological correlation, a diagnosis of pyoderma gangrenosum with aseptic splenic abscesses was made and confirmed (Figure3). The patient was started on high-dose oral corticosteroid therapy at 1 mg/kg/day, a rapid radio-clinical and biological improvement was observed after the introduction of corticosteroid therapy.

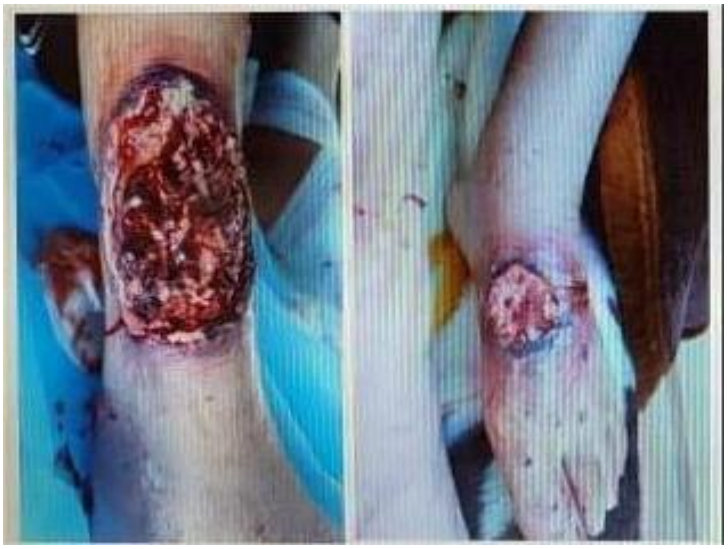
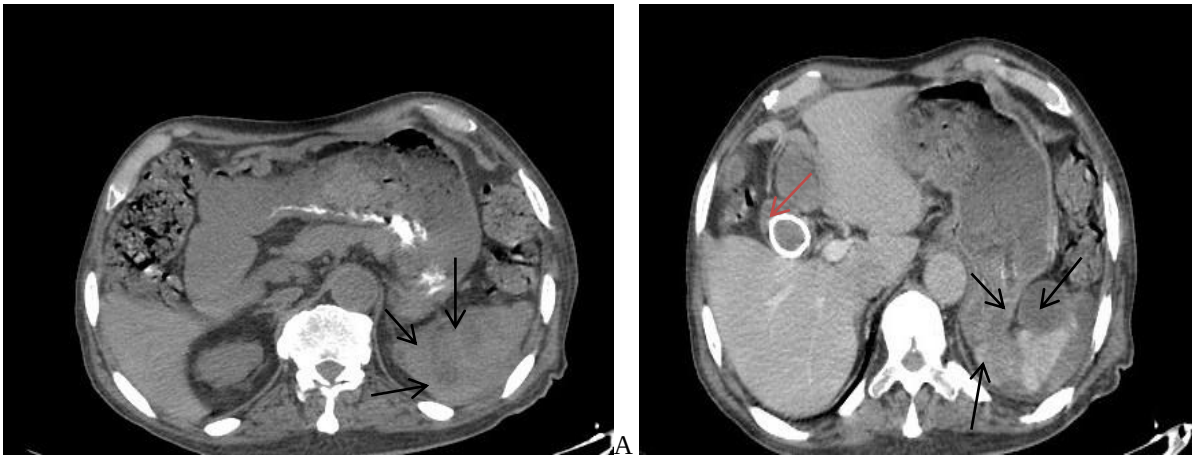
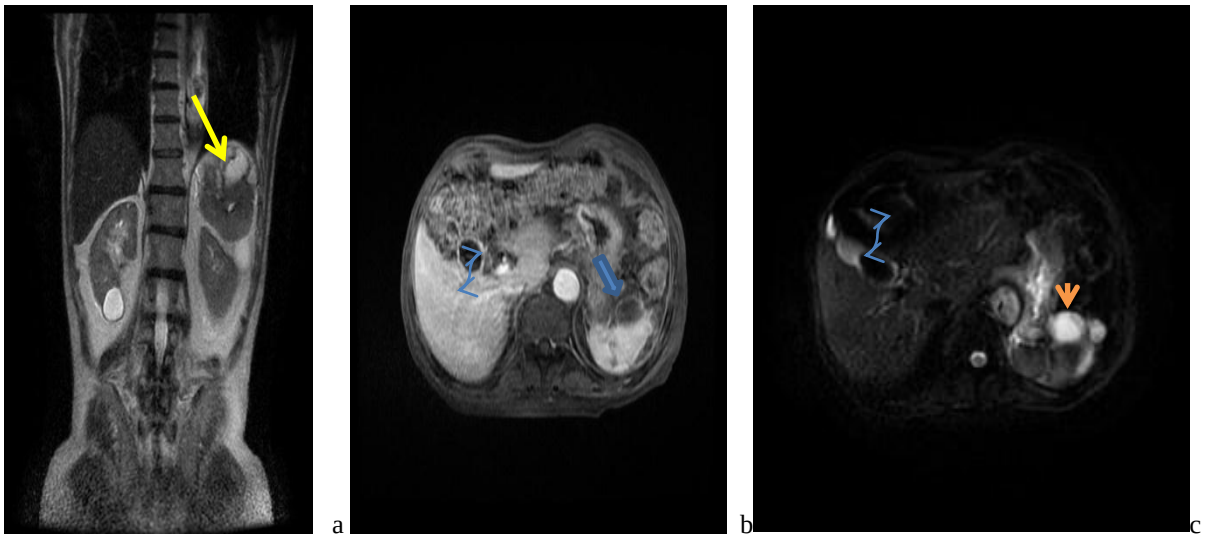


Figure 1 : Ulcers on the lower third of the right leg and left foot, well-defined, oval-shaped, with granulating tissue, violaceous borders, and surrounded by an erythematous halo.



A B

Figure 2 : Axial section of an abdominal CT scan without (A) and following contrast injection in the portal phase with axial slices (B) demonstrating intrasplenic and subcapsular hypodense collections, some encapsulated and others not, with liquid density and thickened, enhanced wall, suggestive of splenic abscesses (arrow), Noted gallbladder stones (red arrow)



a b c

Figure 3 : The MRI images showed that the intrasplenic and subcapsular lesions are heterogeneously hyperintense on T2WI coronal images (yellow arrow), with restriction on the diffusion sequence (c) (orange arrow head). On the contrast-enhanced MRI images (b), peripheral wall enhancement is noted (double blue arrow). Additionally, the signal void of the gallstone is visible on all sequences (curved tail arrow)

III. Discussion

Pyoderma gangrenosum (PG) is a rare chronic neutrophilic dermatosis of unknown etiology. The global incidence is estimated to be around 3 to 10 cases per million people per year. Although PG affects both sexes, there is a slight female predominance. The disease can occur at any age but is most commonly seen during the fourth and fifth decades of life. In 50–70% of the cases, inflammatory bowel diseases, hematological malignancies, or rheumatologic disorders are associated with PG.

Extracutaneous locations of pyoderma gangrenosum are rare. Several organs can be affected, but isolated pulmonary involvement is the most common [2]. Other visceral locations have been reported, particularly multiple intra-abdominal sites, with hepatic ,splenic [3 ,4], pancreatic [6], and lymph node abscesses [10], as well as extra-abdominal locations, such as ocular or bone involvement [,4,5]. These manifestations can also be found in other neutrophilic dermatoses, which may sometimes occur concurrently during the disease course [4].

The eruption initially appears are a pustule which enlarges and ulcerates with ill-defined undermined borders that are bluish or violaceous in color. The surrounding skin also becomes edematous. Secondary bacterial infection also occurs with the ulcer becoming purulent. The ulcers are typically painful. Most commonly they occur over the pretibial region, however any skin surface can be affected. Rarely mucous membrane and airway involvement can occur. Splenic abscess in PG is asymptomatic in a majority of cases .

Currently, ultrasound has been replaced by CT scanning, which offers higher sensitivity and specificity for the diagnosis of splenic abscesses, estimated at 95% and 92%, respectively. The abscess appears as a hypodense image with slight peripheral contrast enhancement (7), (8). Occasionally, this contrast enhancement may be absent, and the clinical context is necessary to suggest the diagnosis of an abscess . CT scanning also allows for guided puncture, with a morbidity rate of 5% and a mortality rate below 1% for experienced teams (9). The risks of surgical puncture are higher, with morbidity ranging from 11% to 28% and mortality from 6% to 14% . MRI is not routinely performed due to the excellent sensitivity and specificity of CT scanning, which also offers faster results and lower costs. However, MRI has the advantage over CT in detecting hyper-vascularized lesions that carry a risk of hemorrhagic complications.

The diagnosis of aseptic splenic abscess and pyoderma gangrenosum is one of exclusion, based on several criteria: clinical appearance of a deep abscess on radiological examination, with neutrophilic characteristics confirmed by surgical pathology, aspiration, or skin biopsy of the ulcer; (10), negative infectious bacterial and parasitological serologies (notably for Yersinia and mycobacteria), ineffectiveness of antibiotic therapy, and rapid clinical and radiological improvement following the introduction of corticosteroid therapy.

Differential diagnoses for splenic lesions in pyoderma gangrenosum include vasculitis, tumors, and infections. Imaging studies are crucial in the diagnosis and follow-up for these patients.

The first-line treatment for extracutaneous manifestations of pyoderma gangrenosum is high-dose corticosteroid therapy (1 mg/kg per day of prednisone), which proved effective in our patient. However, in some previously reported cases of splenic involvement, cyclosporine was combined with systemic corticosteroids

from the outset. Additionally, a splenectomy was performed for both diagnostic and therapeutic purposes before starting treatment with dapsone alone, followed by systemic corticosteroids.

IV. Conclusion

The visceral involvement of PG is rare, comprising non-infective sterile abscesses and requiring thorough diagnostic evaluation and targeted treatment. Successful management often involves high-dose corticosteroids and, in some cases, additional therapeutic agents or interventions like splenectomy to achieve clinical and biological improvements. Corticosteroid dependence and relapses are common (approximately 60% of cases), justifying the use of other immunosuppressants, notably anti-TNFα agents.

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