Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Dermatology

Pyoderma Gangrenosum Revealing Splenic Abscesses: A Rare Clinical Case

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i10.105 | **Received:** 18.06.2025 | **Accepted:** 12.08.2025 | **Published:** 31.10.2025

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Abstract **Case Report**

Pyoderma gangrenosum is a neutrophilic dermatosis characterized by aseptic skin ulcerations and may reveal an underlying systemic disease. Extracutaneous manifestations of pyoderma gangrenosum and other neutrophilic dermatoses are rare, most often involving the lungs. Through this work, we report the case of a patient presenting with a pyoderma gangrenosum associated with splenic abscesses. We report the case of an 81-year-old patient who was hospitalized for multiple lower limb ulcerations in the context of general malaise and unquantified fever. Biological tests revealed an inflammatory syndrome. Abdominal CT scan and MRI showed intrasplenic and subcapsular collections suggestive of splenic abscesses. The diagnosis of pyoderma gangrenosum associated with splenic abscesses was made. The patient was started on oral corticosteroids at 1mg/kg/day with good clinical improvement.

Keywords: systemic disease association, neutrophilic dermatoses, splenic abcess\, extracutaneous manifestations, pyoderma gangrenosum (pg).

INTRODUCTION

Neutrophilic dermatoses (ND) refer to a group of conditions characterized histologically by a cutaneous infiltration of mature polymorphonuclear neutrophils, without an identifiable cause. In 1990, several conditions previously described independently, including pyoderma gangrenosum (PG), subcorneal pustular dermatosis of Sneddon-Wilkinson (SCPD), erythema elevatum diutinum (EED), and neutrophilic eccrine hidradenitis (NEH), were integrated into the spectrum of ND.

Pyoderma gangrenosum is a rare condition (3 to 10 cases per million inhabitants per year). It predominantly affects adults between 40 and 60 years of age (average age of 44 years) [1].

Associations with systemic conditions are observed in 57% of PG cases. These are mainly Inflammatory Bowel Diseases (IBD), monoclonal gammopathies, and arthritis. Extracutaneous manifestations are mainly oropharyngeal involvement, joint involvement (sterile pyoarthrosis), and pulmonary inflammatory infiltrates.

We report a case of pyoderma gangrenosum with splenic abscesses.

CASE PRESENTATION

We report the case of an 81-year-old hypertensive patient under treatment who was hospitalized for multiple lower limb ulcerations (Figure *1*). His history dated back 3 weeks with pustules on the right foot and right and left thighs, progressing to painful ulcerations in a context of general malaise and unquantified fever.

FIGURE 1: Ulcers on the lower third of the leg, budding, surrounded by an erythematous, violaceous halo

Physical examination revealed multiple ulcerations (numbering 5), located on the lower third of the right leg and above the right and left popliteal fossae, of variable size, the largest measuring 10×11cm, with a fibrinous and budding base in places, with an elevated and violaceous border, sensitive and bleeding on contact (Figure 2).

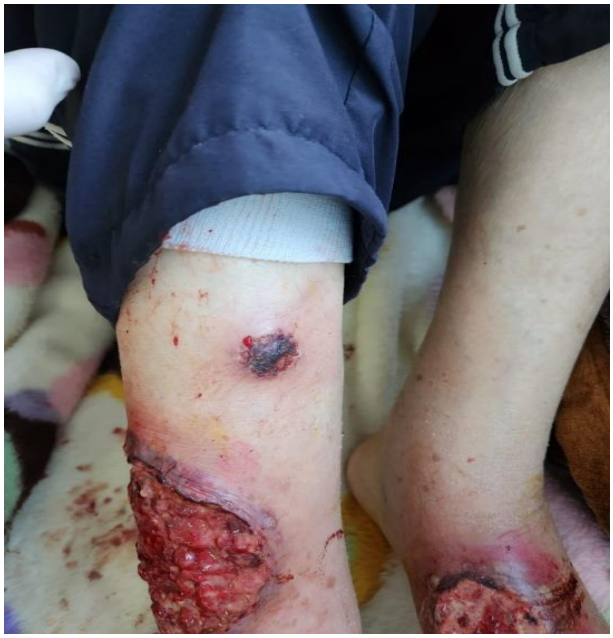

FIGURE 2: Multiple Ulcers on the lower third of the leg, budding, surrounded by an erythematous, violaceous halo

Biological tests revealed an inflammatory syndrome with hyperleukocytosis at 15,300 with 85% neutrophils, a CRP of 168 mg/l, and a polyclonal increase in gamma globulins. Superficial bacteriological sampling showed numerous neutrophils with a negative culture. Histology showed a subcorneal pustule and discrete spongiosis in the epidermis. The dermis showed a very dense inflammatory infiltrate, composed exclusively of neutrophils forming a true abscess (Figure 3).

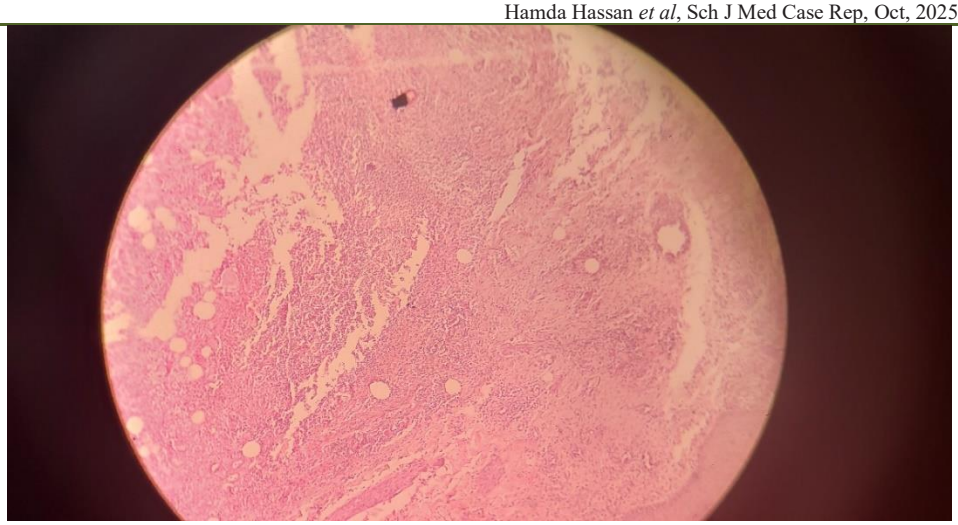

FIGURE 3: **The dermis shows a very dense inflammatory infiltrate, composed exclusively of neutrophils forming a true abscess**

The abdominal CT scan showed intrasplenic fluid-density formations, one of which was subcapsular and the largest measuring 23×19 mm (Figure *4*). MRI showed restricted diffusion in all intrasplenic and subcapsular collections, suggestive of splenic abscesses.

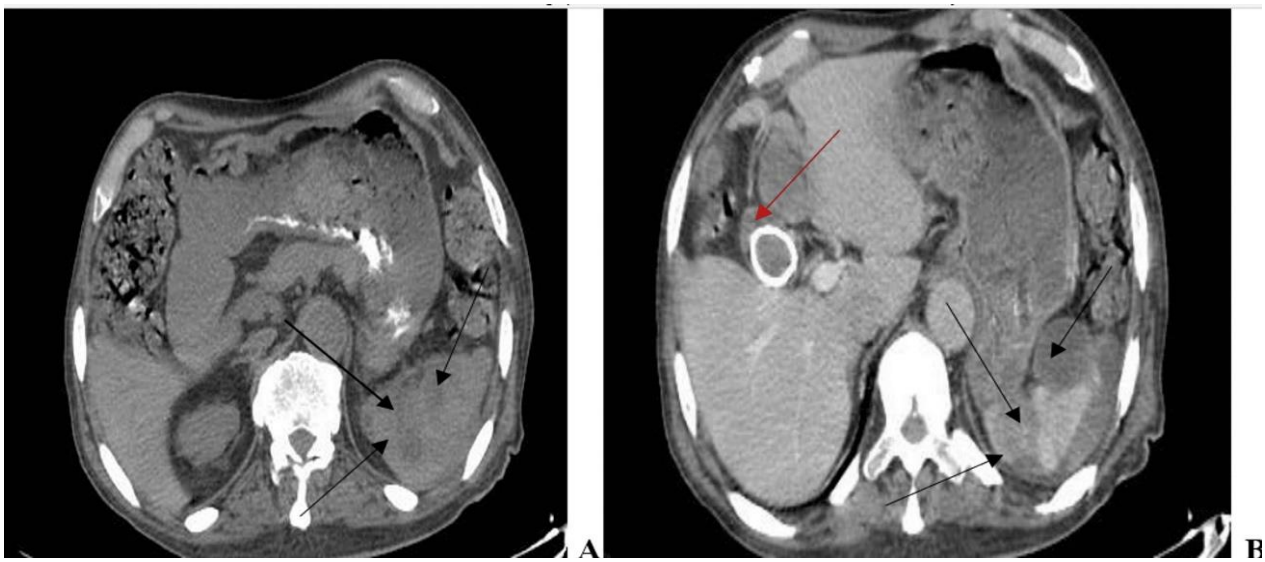

FIGURE 4: Axial section of an abdominal CT scan without (A) and following contrast injection in the portal phase with axial slices (B) demonstrating intrasplenic and subcapsular hypodense collections, some encapsulated and others not, with liquid density and thickened, enhanced wall, suggestive of splenic abscesses (black arrow). Noted gallbladder stones (red arrow)

The diagnosis of pyoderma gangrenosum associated with splenic abscesses was made. The patient was started on oral corticosteroids at 1mg/kg/day with good clinical improvement, re-epithelialization of the ulcers, and disappearance of the inflammatory syndrome with CRP at 1.2 mg/l. A follow-up CT scan showed residual imaging.

DISCUSSION

Neutrophilic dermatoses (ND) refer to a group of conditions characterized histologically by a cutaneous infiltration of mature polymorphonuclear neutrophils, without an identifiable cause [1]. These conditions are characterized by the frequent association of ND with systemic conditions (monoclonal gammopathies, inflammatory bowel diseases (IBD), autoimmune rheumatic diseases, and hematological malignancies), the possibility of specific visceral extracutaneous involvement characterized by a mature neutrophilic infiltrate that can affect various organs (lungs, bones, liver...), and the usual therapeutic sensitivity to systemic corticosteroids and dapsone.

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis (3 to 10 cases per million inhabitants per year). It predominantly affects adults between 40 and 60 years of age (average age of 44 years),

with a female predominance (female/male sex ratio of 3:1). Several clinical variants have been described, including the classic ulcerative, bullous, pustular, vegetative, peristomal, and postoperative forms [2]. The diagnosis of PG is often clinical. The diagnosis is suspected based on characteristic skin ulcers. Biopsies can be helpful to support a diagnosis of pyoderma gangrenosum; however, particularly in long-standing ulcerations, biopsies are unlikely to reveal characteristic acute signs such as a dense neutrophilic infiltrate [3].

Ulcerative PG is the most common clinical variant. It begins with small sterile pustules surrounded by an erythematous halo; the pustular lesions extend and progressively ulcerate within a few days to evolve into a necrotic and mucopurulent ulcer. The pyoderma gangrenosum (PG) ulceration is delimited by a violaceous, raised border, sometimes with undermined edges. The borders are circular, inflammatory, and undermined in their inner part with purulent tracts. The ulceration is surrounded by an erythematous halo. PG lesions can be single or multiple and preferentially affect the lower limbs and trunk, less commonly the neck, upper limbs, or external genitalia [4,5].

Morbid associations are observed in 50-70% of PG cases [6]. In the ulcerative form, these are mainly IBD, arthritis, and monoclonal gammopathies. Reported extracutaneous locations include aseptic abscesses, arthritis, aseptic meningitis, neutrophilic lung disease, and multifocal osteomyelitis. The location of splenic, hepatic, and lymph node abscesses associated with pyoderma gangrenosum has been reported in the literature [7-9].

The choice of pyoderma gangrenosum (PG) treatment depends on the size, number, location, and progression of the ulcers. Mild PG can be treated with topical corticosteroids or topical tacrolimus. Severe PG is primarily managed with systemic corticosteroid therapy. This is initiated with prednisone at a daily dose of 0.5 to 1 mg/kg and typically leads to healing of skin lesions within a few weeks. Corticosteroid doses must be gradually reduced to prevent relapses or corticosteroid dependency. Cyclosporine, at an initial dose of 2 to 6 mg/kg/day, can be used alone or in combination with corticosteroids as a steroid-sparing agent [10].

Conclusions

Extracutaneous locations of pyoderma gangrenosum are rare. Several organs can be affected, but isolated pulmonary involvement is the most frequent. Other visceral locations have been reported, particularly multiple intra-abdominal locations, with hepatic, splenic, pancreatic, and lymph node abscesses, as well as extra-abdominal locations. When visceral involvement precedes skin involvement, the diagnosis is difficult, with the clinical picture raising suspicion of infection. In our case, splenic involvement was concomitant with skin involvement.

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