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Hidradenitis suppurativa and pyoderma gangrenosum: A common autoinflammatory broad spectrum

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Abstract

Hidradenitis suppurativa (HS) and pyoderma gangrenosum (PG) are chronic inflammatory skin conditions with distinct clinical presentations, but with an increasing recognition of their overlapping pathogenic mechanisms. HS primarily affects areas rich in apocrine glands and is characterized by painful nodules, abscesses, and draining sinus tracts, which often progress to scarring. PG, in contrast, presents as rapidly progressive, painful ulcers with neutrophilic infiltration, commonly affecting the lower limbs and trunk. Both conditions are associated with autoinflammatory syndromes, such as PASH and PAPASH, and are characterized by immune dysregulation involving cytokines such as TNF-α, IL-1β, IL-17 and IL-23.

We present two rare cases of concomitant HS and PG. The first case is a 51-year-old male with a 20-year history of severe HS who developed PG lesions on his lower limbs, later managed with dapsone and adalimumab. The second case describes a 58-year-old male with a 12-year history of HS who recently developed PG lesions associated with underlying inflammatory bowel disease (IBD). Both patients exhibited elevated inflammatory markers and histopathological confirmation of PG.

These cases highlight the importance of recognizing the co-occurrence of HS and PG, as this may represent a late or systemic manifestation of a common autoinflammatory disorder. Early identification and targeted treatment with biologics, particularly TNF-α inhibitors, may offer improved disease control and patient outcomes. Due to the rarity of these conditions and the associated diagnostic challenges, further case reports and studies are crucial for improving our understanding of their shared pathophysiological mechanisms and refining therapeutic strategies.

Keywords: Hidradenitis Suppurativa; Pyoderma Gangrenosum; Autoinflammatory Syndromes; Adalimumab; Dapsone; Inflammatory Skin Disease

1. Introduction

Hidradenitis suppurativa (HS), previously known as Verneuil's disease, is a chronic inflammatory skin disease characterized by recurrent inflamed subcutaneous nodules and abscesses, suppurating sinus tracts, subsequent dermal fibrosis and scarring [1,2]. HS most commonly affects areas where the apocrine glands are abundant, such as the axillary, inguinal, perineal and anogenital regions [1]. The condition is multifactorial and involves genetics, immunology, metabolic comorbidities and environmental factors such as smoking, obesity, hyperandrogenism, infectious agents and traumatic friction [1,2]. Moreover, HS tends to follow a relapsing course with suspensive treatment requiring long-term

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combined strategies and may be associated with acne, dissecting cellulitis of the scalp, pilonidal cysts and Crohn's disease [1].

On the other hand, pyoderma gangrenosum (PG) is a relatively rare autoinflammatory neutrophilic dermatosis. Its clinical presentation is characterized by an inflamed papule, papulo-pustule or nodule that progresses into a painful and rapidly expanding suppurative, cribriform ulcer with raised, irregular borders and peripheral erythema [3]. The distribution of PG lesions is predominantly localized in the lower extremities and trunk. However, they may manifest anywhere on the tegument and are frequently associated with other immune-mediated systemic diseases including inflammatory bowel disease (IBD), myelodysplasia or myeloproliferative disorders and rheumatoid arthritis.

A connection between HS and PG was recently described, as both diseases represent features of rare autoinflammatory syndromes such as PASH (Pyoderma gangrenosum, Acne, Suppurative Hidradenitis) and PAPASH (Pyogenic Arthritis, Pyoderma gangrenosum, Acne, Suppurative Hidradenitis) [2,4]. However, this correlation remains underrecognized and the coexistence of HS and PG in a single patient has rarely been documented in the literature. We describe two cases of concomitant HS and PG, highlighting the diagnostic and therapeutic challenges associated with this overlap.

2. Case 1

A 51-year-old male patient with a medical history of hypertension and chronic smoking was diagnosed with a Hurley stage III HS, which had been evolving for 20 years. The patient had initially been treated with conventional treatment protocols including antibiotics (ciprofloxacin, moxifloxacin, ceftriaxone), metronidazole and methylprednisolone, resulting in moderate clinical improvement and a relatively reduced Dermatology Life Quality Index (DLQI) from 29 to 19.

On clinical examination at follow-up (Figures 1 and 2), he presented with a new onset of painful, erythematous subcutaneous nodules and suppurative cribriform ulcers on the lower limbs, associated with multiple inflammatory nodules, abscesses, draining sinus tracts, polyfistulized plaques and hypertrophic rope-like scars in the axillary, inguinal and perineal regions.

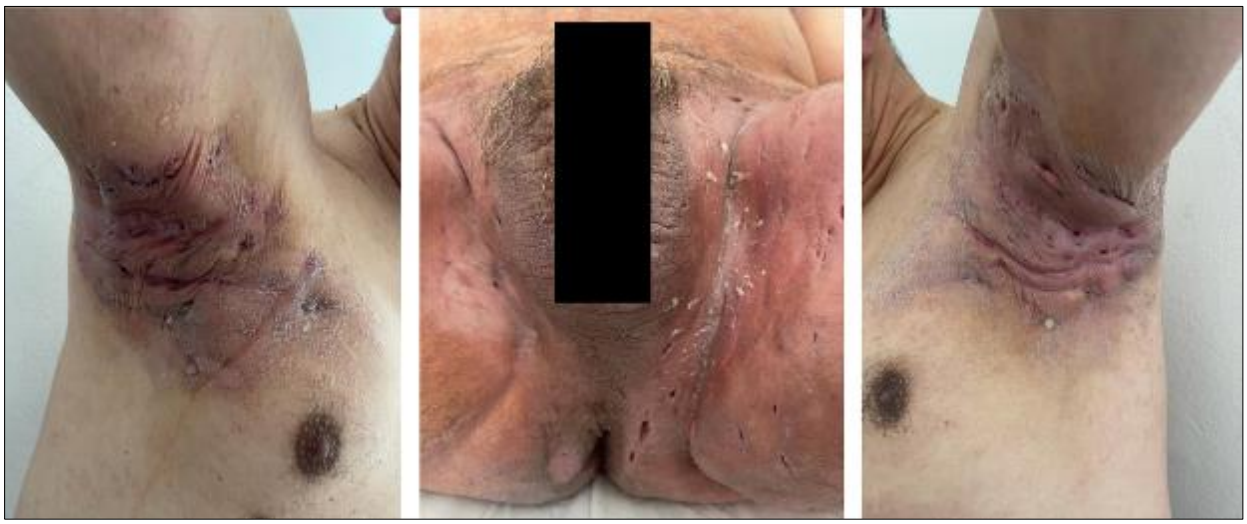

Figure 1 Multiple inflammatory nodules, abscesses, draining sinus tracts, polyfistulized plaques and hypertrophic rope-like scars in the axillary, inguinal and perineal regions

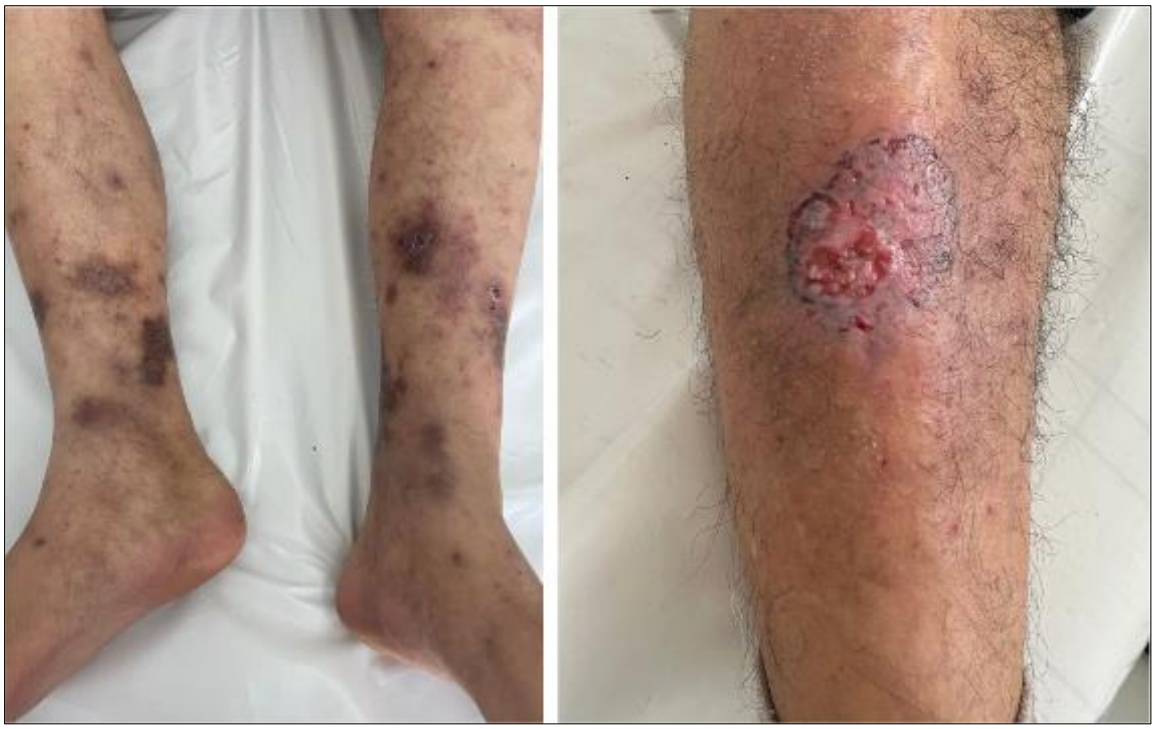

Figure 2 New onset of painful, erythematous subcutaneous nodules and suppurative cribriform ulcers on the lower limbs healing into hyperpigmented atrophic scars

Laboratory findings showed hyperleukocytosis (16690/mm^3) with neutrophilic predominance (11182/mm^3), elevated erythrocyte sedimentation rate (ESR, 120 mm/h), a fibrinogen level of 7.35 g/L and increased C-reactive protein (CRP, 190 mg/L), while procalcitonin (PCT) was negative.

Histopathological examination of the inflammatory nodules and ulcers of the lower limbs revealed a neutrophilic dermo-hypodermitis consistent with PG. Inflammatory bowel disease (IBD) work-up was negative. The patient was treated with dapsone (100 mg/day) resulting in the healing of ulcers into hyperpigmented atrophic scars and no new PG lesions were observed after two months follow-up.

Despite combination therapies for the concomitant HS and PG, the patient had an insufficient response marked by cycles of remission and flare-ups. After 14 months and due to the persistent disease activity, he was started on adalimumab (160 mg initially, followed by 80 mg every two weeks) in combination with dapsone for PG.

3. Case 2

A 58-year-old male patient with a history of chronic smoking and anal fistulas was followed in our dermatology department for a Hurley stage III HS, that had been evolving for 12 years. Initially, he was started on a combination protocol of prednisone (20 mg/day) with adjuvant treatment, ciprofloxacin (500 mg x 2/day) and sulfamethoxazole/trimethoprim 800 mg/160 mg for 10 days per month but with poor adherence and response.

Subsequently, the patient received a new treatment regimen consisting of ceftriaxone (2 g/day) and metronidazole (500 mg x 3/day) for 21 days, followed by doxycycline (100 mg/day). A favorable clinical evolution was observed with a desinfiltration and size reduction of the polyfistulized nodular lesions in the axillary, inguinal and perineo-scrotal regions, as well as drying of painful abscesses and an improvement of the DLQI score (9 vs 15).

On dermatological examination (Figures 3 and 4), the patient presented with new inflammatory nodular lesions centered with pustules on the lower limbs. The evolution was marked by the ulceration of lesions with erythematoviolaceous raised borders and a centrifugal extension on the anterior surfaces of both legs.

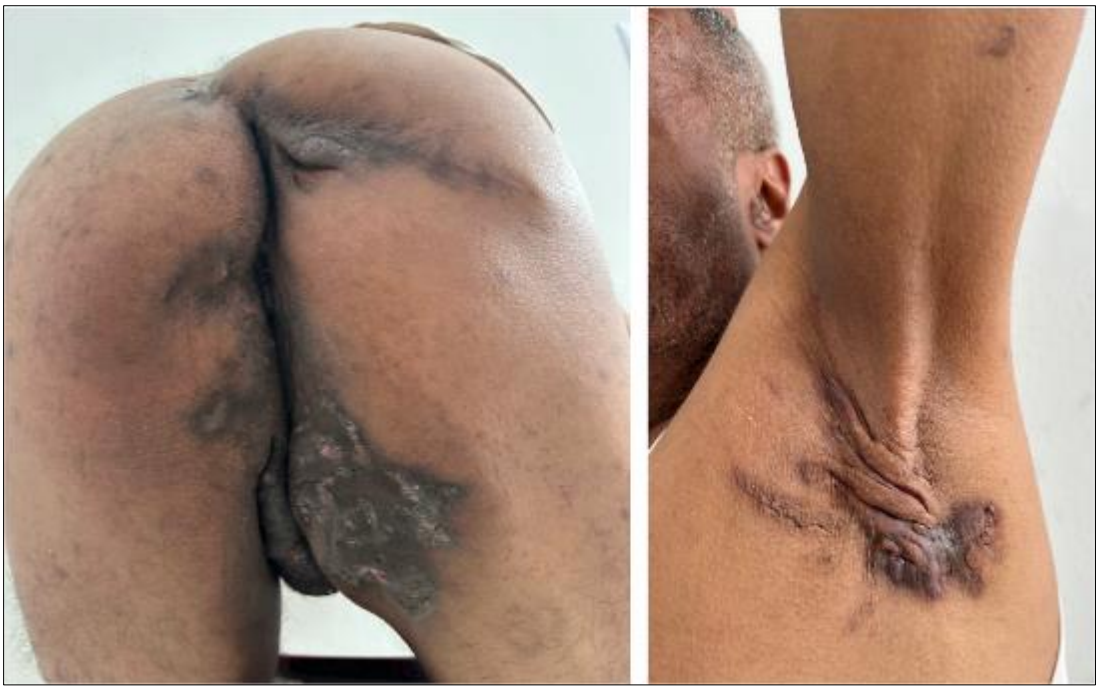

Figure 3 Polyfistulized nodular lesions and painful abscesses in the axillary, inguinal and perineo-scrotal regions

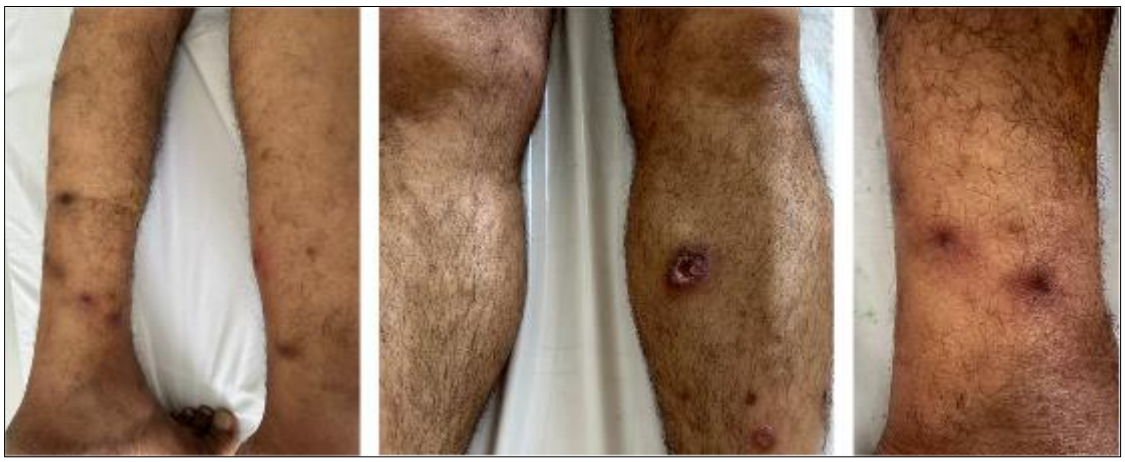

Figure 4 New inflammatory nodular lesions centered with pustules on the lower limbs, and ulcerations with erythematoviolaceous raised borders and a centrifugal extension

Laboratory tests revealed hyperleukocytosis with neutrophilic predominance, elevated inflammatory markers (ESR 140 mm/h, CRP 110 mg/L) and histopathological findings showed a mixed inflammatory infiltrate including neutrophils in favor of PG. Considering the patient reported a chronic constipation and diarrhea alternation, further investigations were conducted and revealed a positive fecal calprotectin and biopsy specimens from colonoscopy were consistent with a chronic colitis in favor of an IBD.

The patient was started on dapsone for PG, resulting in healing of the lesions on the lower limbs. A pre-biologic therapy work-up was ongoing to initiate adalimumab and the patient was referred to gastroenterology for IBD management and interdisciplinary follow-up.

4. Discussion

While the exact pathogenesis of HS and PG remains incompletely elucidated, current understanding strongly suggests that HS development is triggered by an abnormal secretion of apocrine glands, leading to follicular occlusion and cyst

formation [1,2]. This results in an exaggerated inflammatory response and chronic suppuration, destroying pilosebaceous units and leading to confluent nodular lesions and draining sinus tracts. Over time, the healing process can cause dermal fibrosis, hypertrophic rope-like scars and contractures [1].

As a response to various stimuli in genetically predisposed individuals, a dysregulation of the immune system is increasingly recognized to play a pivotal role in the uncontrolled inflammatory process associated with HS, involving a range of cytokines such as tumor necrosis factor-alpha (TNF-α), Interleukin-1 beta (IL-1β), IL-17 and IL-23 [1,2,5]. Particularly through the implication of the IL-1β pathway and type 1 immune response, including neutrophilic inflammation, inflammasomes and the activation of caspase-1 [1,2].

Furthermore, there is growing evidence supporting the efficacy of early initiation of the FDA approved biologics: adalimumab (TNF-α inhibitor) and more recently secukinumab (IL-17A inhibitor) for the management of moderate-to-severe HS compared to antibiotics, immunosuppressants or other adjuvant treatments [1,2].

For patients with PG, genetic and immunological factors such as dysregulation of the innate and adaptive immune systems due to neutrophil chemotaxis dysfunction, have also been identified as potential causative factors. The physiopathology of PG lesions is characterized by a perifollicular inflammation involving type 1 and type 3 immune responses, and increased inflammasome activation [3,6]. This cascade leads to a mixed inflammatory cell infiltrate dominated by neutrophils with increased levels of various cytokines including TNF-α, IL-12, IL-15, IL-17, IL-23 and IL-36, followed by intradermal abscesses and dermoepidermal necrosis [3].

Additionally, some recent studies suggest that elevated concentrations of the IL-8 cytokine are often found in the cutaneous ulcers of PG lesions, with or without underlying IBD [6]. IL-1 may also play a crucial role in the pathogenesis of PG, regardless of its association with the Pyogenic Arthritis, Pyoderma gangrenosum and Acne (PAPA) syndrome [4].

Treatment for PG usually begins with rapid-acting systemic immunosuppressants to control inflammation, such as systemic corticosteroids and/or cyclosporine. TNF-α inhibitors offer a more effective and favorable safety profile. Steroid-sparing therapies include dapsone, wound care and management of associated comorbidities and/or underlying disorders [1,5].

Despite their clinical differences, increasing evidence supports that HS and PG share some similar physiopathological pathways. Both diseases are characterized by immune system dysregulation and an enhanced inflammatory response mediated by neutrophils and proinflammatory cytokines [1,4,6]. They are also frequently associated with other autoinflammatory and immune-mediated conditions, particularly IBD [3,6].

Furthermore, in our cases, the timing of PG onset relative to the development of HS varies between patients: 20 versus 12 years. This raises the possibility that PG occurred as a late manifestation of the same underlying systemic inflammatory process that triggered HS [2,4]. Although the inflammatory process is predominantly local, our patients with coexisting HS and PG lesions had evidence of systemic inflammation as indicated by the presence of concomitant comorbidities (hypertension, obesity, IBD) and elevated acute inflammatory markers [2]. In addition, Jennifer L. Hsiao et al. suggest that HS and PG may be considered part of a broad spectrum that includes the cutaneous disorders observed in PASH and PAPASH autoinflammatory syndromes [4].

A deeper comprehension of the underlying pathogenesis of both conditions is, therefore, essential for optimizing treatment strategies and improving the advancing roles of biologic and targeted therapies, particularly in the management of patients with concomitant HS and PG that has proven challenging [1,2,4].

5. Conclusion

In summary, overlap cases of HS and PG are not frequently reported in the literature; however, there is a discernible connection between both diseases. A better understanding of their common underlying pathogenesis may provide insightful directions for diagnosis, assess more effective treatment strategies that target the overlapping features of their cytokine-driven inflammatory pathways and immune dysregulation to improve patients' outcome.

Compliance with ethical standards

Disclosure of conflict of interest

Authors have no conflicts of interest to declare.

Statement of ethical approval

This study was conducted according to the principles specified in the Declaration of Helsinki and the local ethical guidelines of the Ethics Committee for Biomedical Research of the Faculty of Medicine and Pharmacy, Hassan II University of Casablanca, Morocco (International Review Board 00002504). The confidentiality of personal and clinical data was guaranteed.

Statement of informed consent

Consent was obtained from all individual participants included in the study.

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