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PASH Syndrome (Pyoderma Gangrenosum, Acne, and Hidradenitis Suppurativa): Presentation of Two Cases and Literature Review

Liliana Herrera-Martínez[1], Diana Elizabeth Medina Castillo[2]*, Victor Jaimes Hernandez[3], Diana Alejandra Aguilar Medina[4] and Carlos Humberto Sandoval Jimenez[5]

[1]*Internal Medicine Resident, "Dr. Nicolás San Juan" General Hospital, Toluca, State of Mexico.*

[2]*Dermatologist, Medical-Surgical Clinic at D'Corium Dermatológica Metepec, Mexico.*

ORCID: 0009-0009-3616-2750

[3]*Internist, Dermatologist, and Dermatopathologist at Centro Médico de Toluca Metepec, Mexico.*

[4]*Dermatologist, Cutaneous Surgery and Oncology, Mohs Surgeon at D'Corium Dermatológica Metepec, Mexico.*

[5]*General Surgeon and Colorectal Surgeon at Centro Médico Toluca Metepec, Mexico.*

***Correspondence:**

Diana Elizabeth Medina Castillo, D'Corium Dermatológica, Manuel Doblado 2560, Fracc. Pilares, Metepec, State of Mexico, C.P. 52179, Mexico, Tel: +52 722 3969747 / +52 722 2130071.

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ABSTRACT

Background: *PASH syndrome (pyoderma gangrenosum, acne, and Hidradenitis suppurativa) is a rare autoinflammatory disorder within the spectrum of neutrophilic dermatoses. Early recognition is essential due to its chronic course and variable therapeutic response.*

Clinical Cases: *We describe two adult male patients who presented the characteristic triad. The first case showed a refractory course despite conventional therapies and tumor necrosis factor (TNF) inhibitor therapy, with histopathological confirmation. The second case presented with lower-limb pyoderma gangrenosum associated with cicatricial acne and Hurley stage III hidradenitis suppurativa, showing favorable response to systemic corticosteroids.*

Conclusions: *These cases highlight the relevance of PASH syndrome in the differential diagnosis of neutrophilic dermatoses. Early diagnosis allows individualized therapeutic strategies and may improve clinical outcomes.*

Keywords

PASH syndrome, Pyoderma gangrenosum, Hidradenitis suppurativa, Acne, Neutrophilic dermatoses.

Background

PASH syndrome (pyoderma gangrenosum, acne, and hidradenitis suppurativa) was first described in the 2010s as a rare association of three chronic inflammatory conditions: pyoderma gangrenosum (PG), severe acne, and hidradenitis suppurativa (HS), without joint involvement. It falls within the spectrum of neutrophilic dermatoses mediated by innate immunity and shares clinical and pathophysiological characteristics with other autoinflammatory syndromes, such as PAPA (pyoderma gangrenosum, pyogenic arthritis, and acne) and PAPASH (pyoderma gangrenosum, acne, hidradenitis suppurativa, and sterile arthritis).

Initially described in young adults, with a male predominance, its recognition is essential for the comprehensive management

of patients and the appropriate selection of immunomodulatory therapies. Abnormal neutrophil activation and excessive production of proinflammatory cytokines play a central role in its pathogenesis. Although its prevalence is low and probably underestimated, timely recognition of PASH syndrome aids in early diagnosis and rapid treatment with targeted immunomodulatory therapies.

Clinical Cases

Case 1

A 30-year-old male patient, originally from and residing in Toluca, State of Mexico, with a history of type II cystic fibrosis (due to a cystic fibrosis transmembrane conductance regulator mutation) treated with ivacaftor, tezacaftor, elexacaftor, and pancreatic enzymes, type 1 diabetes mellitus managed with insulin glargine, as well as a history of acne and hidradenitis suppurativa without prior treatment. He presented with painful lesions in the intergluteal region of three week´s duration.

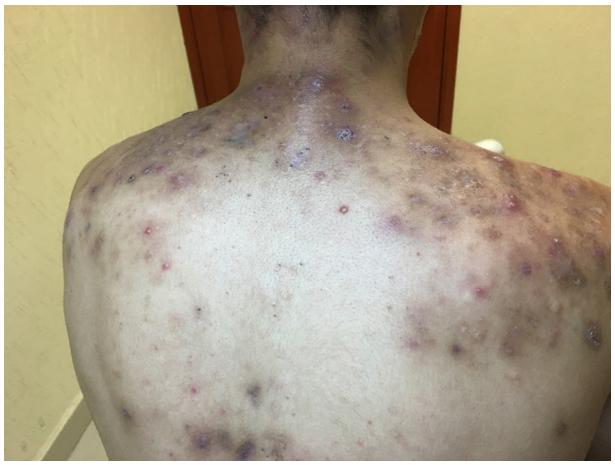

Figure 1: Severe truncal acne.

Physical examination revealed a disseminated polymorphic dermatosis affecting the face and trunk, as well as anterior and posterior upper thorax (Figures 1 and 2). This dermatosis consisted of a few open comedones, papules, pustules, and cysts. In the axillae and groin, fistulous tracts were found draining scant malodorous seropurulent discharge. In the intergluteal region, three ulcers approximately 2 cm in diameter, with well-defined borders and purulent discharge, were also present (Figure 3). An initial diagnosis of nodulocystic acne and hidradenitis suppurativa was established, and systemic antibiotic and isotretinoin treatment was initiated with a partial response. Subsequently, due to the progression of the hidradenitis (Hurley II), treatment with subcutaneous adalimumab 80 mg every two weeks was started for 6 months without clinical improvement. Surgical management of the intergluteal lesions was proposed by coloproctology due to intense pain (unroofing of the lesions) and repair with an island flap or V-Y flap (Figure 4). The histopathology study with routine staining showed discrete hyperkeratosis and acanthosis (Figure 5), superficial, mid and deep dermis with predominant polymorphonuclear infiltrate (Figures 6 and 7), neutrophil microabscesses, dilated and congested vessels compatible with pyoderma gangrenosum, ruling out hidradenitis. Based on these findings, the diagnosis of PASH syndrome was established, and currently continuing treatment with clindamycin 300 mg, isotretinoin 60 mg weekly, copper sulfate compresses with improvement of acne and persistent hidradenitis suppurativa of the left axilla. Four years after surgery, he has not presented pathergy.

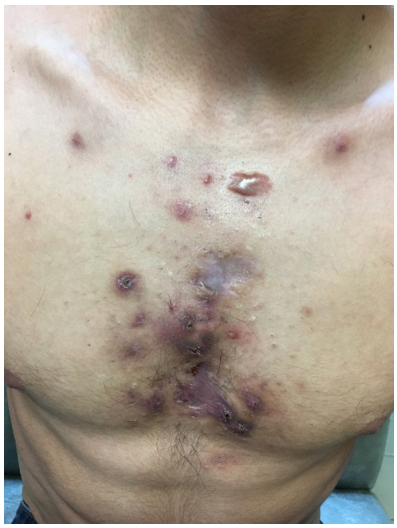

Figure 2: Severe acne in the presternal region.

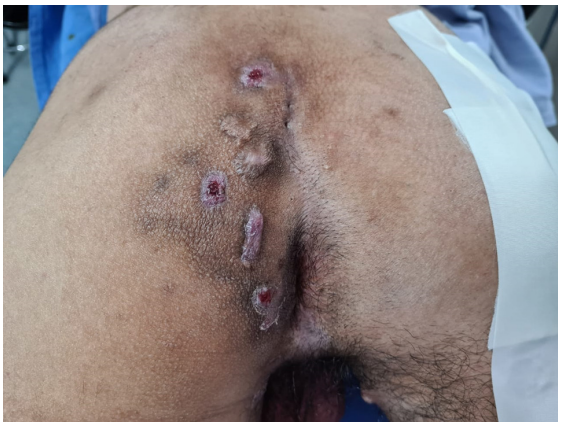

Figure 3: Ulcers and fistulous tracts in the intergluteal region.

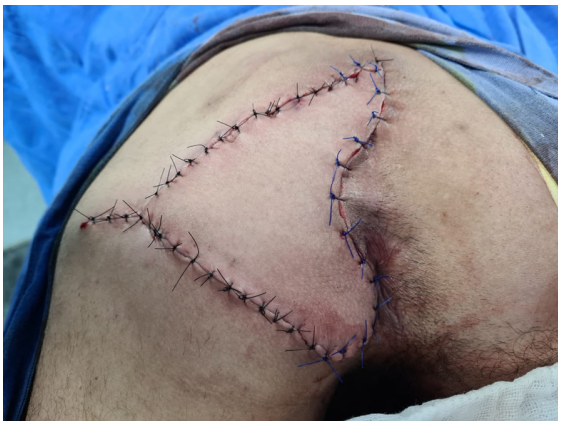

Figure 4: Disroofing of lesions and island flap (V-Y).

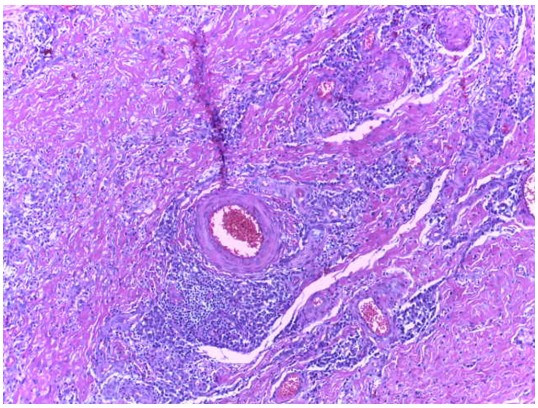

Figure 5: H&E 20X, neutrophil microabscesses, dilated and congested vessels consistent with pyoderma gangrenosum.

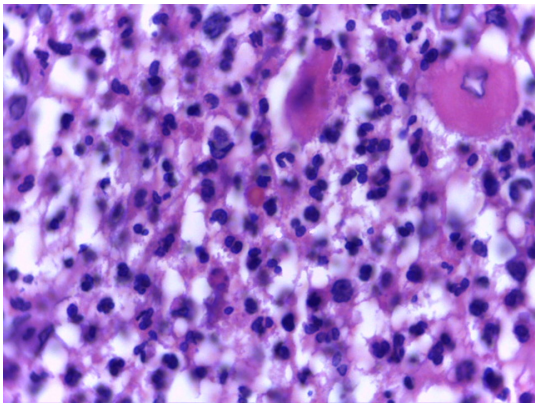

Figure 6: H&E 40X, close-up of the predominant polymorphonuclear cells.

Case 2

A 39-year-old male patient, originally from and residing in Temoaya, State of Mexico, with a history of grade III obesity (body mass index BMI 32). He presented with painful ulcers on both legs of two month's duration.

Physical examination revealed bilateral and symmetrical polymorphic dermatosis, disseminated to the head, trunk, and lower extremities, affecting the face, back, axillae, and legs. On the face and back, icepick and box scars were observed on seborrheic skin with some erythematous papules. In the axillae, two fistulous tracts with slight purulent discharge, hyperpigmentation and retractile scars were observed (Figure 7). On both sides of the legs, ulcers of 2 - 4 cm in diameter with poorly and violaceous borders were identified, some with slough and hemorrhagic crusts (Figure 8).

Our initial diagnoses were cicatricial acne, hidradenitis suppurativa in the axillae and groin (Hurley III), and probable pyoderma gangrenosum, ulcerative variant. The skin biopsy confirmed the diagnosis of pyoderma gangrenosum, including PASH syndrome. The patient received treatment with systemic corticosteroids, a short course of systemic antibiotics, and topical therapy, with a favorable clinical course.

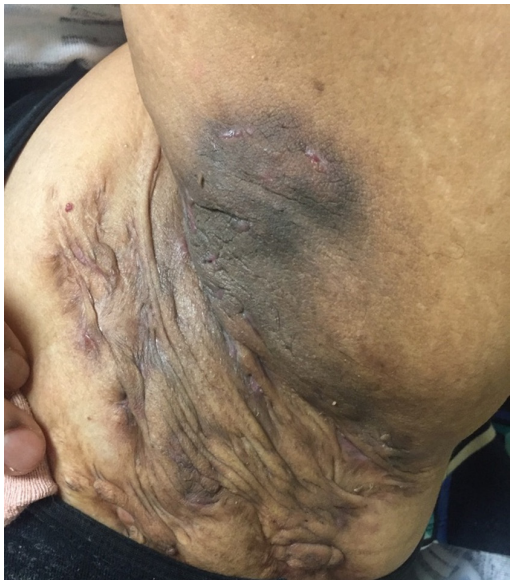

Figure 7: Axillary hidradenitis.

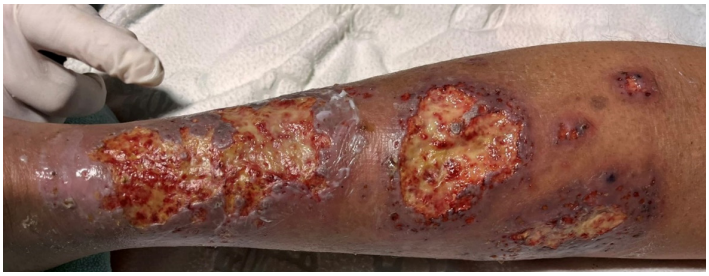

Figure 8: Circular ulcers with violaceous borders and a dirty-looking surface (slough).

Discussion

PASH syndrome is a rare and likely underrecognized autoinflammatory skin disorder, attributable to its heterogeneous clinical presentation and significant overlap with other neutrophilic dermatoses. The coexistence of pyoderma gangrenosum, severe acne, and hidradenitis suppurativa poses a significant diagnostic challenge, particularly when these manifestations occur asynchronously or are evaluated in isolation [1].

Most reported cases involve young adult patients, predominantlymale, with moderate to severe hidradenitis suppurativa [2,3].

From a genetic standpoint, variants in the PSTPIP1 gene have been identified in patients with autoinflammatory syndromes associated with neutrophilic dermatoses, supporting the concept of a shared inflammatory spectrum that includes related entities such as PAPA (pyoderma gangrenosum, pyogenic arthritis, and acne) and PAPASH (pyoderma gangrenosum, acne, hidradenitis suppurativa, and sterile septic arthritis). PSTPIP1 (proline-serine-threonine phosphatase–interacting protein 1) is a key regulator in these conditions, as its variants disrupt inflammasome function and lead to increased IL-1β production.

However, the absence of genetic alterations in all cases suggests a multifactorial origin, involving immunological, and environmental factors [4,5].

The pathophysiology involves abnormal activation of the innate immune system, with excessive release of proinflammatory cytokines such as IL-1β, IL-17, and IL-23, along with intense dermal neutrophilic infiltration. Inflammasome dysfunction and activation of IL-36–mediated pathways have also been implicated. These mechanisms result in chronic sterile inflammation and tissue damage characteristic of pyoderma gangrenosum, acne, and hidradenitis suppurativa.

Clinically, pyoderma gangrenosum is typically the most aggressive manifestation, characterized by rapidly progressive, painful ulcers. In contrast, severe acne and hidradenitis suppurativa contribute significantly to the overall inflammatory burden and negatively impact the patient's quality of life.

Pyoderma gangrenosum presents with rapidly progressive, painful ulcers with undermined, violaceous borders and pathergy. Acne is usually of the conglobate or severe nodulocystic type, affecting the face, chest, and back. Hidradenitis suppurativa is characterized by nodules, abscesses, and fistulous tracts in intertriginous areas. The coexistence of these three entities strongly supports the clinical diagnosis of PASH syndrome. Systemic manifestations, such as arthralgia or inflammatory colitis, may be present in some cases [6,7].

The diagnosis of PASH syndrome is primarily clinical and based on exclusion, supported by nonspecific histopathological findings, including dermal neutrophilic infiltration. Exclusion of infectious, vasculitic, and neoplastic conditions is essential to avoid misdiagnosis and inappropriate treatment.

Management of PASH syndrome requires a multidisciplinary and individualized approach. Treatment of active pyoderma gangrenosum lesions includes systemic corticosteroids and immunosuppressive agents such as cyclosporine, methotrexate, or mycophenolate mofetil. In refractory cases, biologic therapies targeting TNF-α (adalimumab, infliximab), IL-1 (anakinra, canakinumab), or IL-17 (secukinumab) may be considered. In hidradenitis suppurativa, surgical interventions such as deroofing or wide excision may be necessary. Treatment should be tailored according to disease severity and prior therapeutic response [8].

Conclusions

PASH syndrome represents a model of neutrophil-mediated autoinflammatory skin disease. Its recognition enables a more rational therapeutic approach by targeting key inflammatory pathways. Increasing knowledge of its pathophysiology and associated genetic variants may facilitate the development of more specific and effective biologic therapies.

The patients presented here exhibited different clinical manifestations. In case 1, the ulcers and nodular lesions were initially diagnosed as hidradenitis suppurativa. However, histopathological findings consistent with pyoderma gangrenosum led to the suspicion of pathergy. Long-term follow-up has not demonstrated recurrence. In case 2, the bilateral lower-limb ulcers confirmed the initial clinical suspicion, allowing fulfillment of diagnostic criteria for PASH syndrome. Reporting additional cases contributes to expanding current clinical and therapeutic knowledge, as well as generating hypotheses for future research.

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