ACADEMIC JOURNAL OF HEALTH

CASE REPORT

A Case Report: Pyoderma Gangrenosum, Acne and Hidradenitis Suppurativa (PASH) Syndrome

ABSTRACT

PASH syndrome, consisting of the triad of pyoderma gangrenosum (PG), acne, and hidradenitis suppurativa (HS), is a rare chronic autoinflammatory disease. The etiology and pathogenesis of the syndrome are not yet clearly understood. However, certain genes, such as PSTPIP1, have been associated with this syndrome. In treating PASH syndrome, various options are available depending on the disease's severity, including recommendations for weight loss and smoking cessation. Furthermore, there is a rare possibility of cutaneous malignancy developing in HS, a component of PASH syndrome, which tends to worsen the prognosis for these patients. In HS patients, distinguishing between malignant transformation in chronic lesions and classical lesions is challenging, potentially leading to diagnostic delays. Therefore, in high-risk patients with long-standing atypical ulcerative lesions, malignancy should be suspected. This case report presents a 46-year-old male patient diagnosed with PASH syndrome.

Keywords: Acne, hidradenitis suppurativa, pyoderma gangrenosum, treatment

PASH syndrome, which consists of the triad of pyoderma gangrenosum (PG), acne, and hidradenitis suppurativa (HS), is a rare chronic autoinflammatory disease. Although its etiology and pathogenesis are not fully understood, certain genes such as PSTPIP1 are implicated. Subsequently, it has been found to share a cytokine profile with other autoinflammatory diseases (1). In managing PASH syndrome, recommendations include weight loss and smoking cessation, along with various treatment options depending on the disease's severity. For localized cases, topical tacrolimus and antiseptic preparations are advised; for generalized cases, systemic corticosteroids, immunosuppressives, immunomodulators, and biological agents are recommended (1, 2). Additionally, cutaneous malignancy, particularly in HS, a component of PASH syndrome, may rarely develop (3). This case report presents a 46-year-old male patient with PASH syndrome.

CASE REPORT

A 46-year-old male patient presented to our clinic with a 30-year history of hidradenitis suppurativa. He exhibited exudative, painful, malodorous lesions, along with sinus tract formation and scarring. These symptoms initially appeared in the axilla and later spread to the chest, groin, and hips (Figure 1). He initially responded well to intermittent treatment with doxycycline, rifampicin, and clindamycin, but later showed no improvement. His medical history included systemic isotretinoin use and surgical excision from the axilla, groin, and hips, yet the lesions recurred. According to reports from an external center, invasive squamous cell carcinoma (SCC) was diagnosed histopathologically following re-operation for a recurrence in the right axilla one year ago (Figure 2), with no detected metastasis.

Four years ago, following a leg trauma, he developed a painful, ulcerated lesion at the site of the injury. An external center made a clinicopathological diagnosis of pyoderma gangrenosum (Figure 3), and no new PG lesions have developed since he started systemic corticosteroid treatment.

The patient had no joint complaints but had acne and acne scars on his face and back. He had a 25 pack-year smoking history, underwent surgery for stomach cancer seven years ago, and received chemotherapy. We diagnosed him with PASH syndrome due to the pres-

Tuğba Atak

Selda Pelin Kartal

Department of Dermatology, Ankara Etlik City Hospital, Ankara, Türkiye

Corresponding author:
Tuğba Atak
✉ tugba.atak@dr.com

Received: November 03, 2023
Accepted: November 18, 2023

Cite this article as: Atak T, Kartal SP. A Case Report: Pyoderma Gangrenosum, Acne and Hidradenitis Suppurativa (PASH) Syndrome. Acad J Health 2023;1(2):47-49.

DOI: 10.14744/ajh.2023.18209

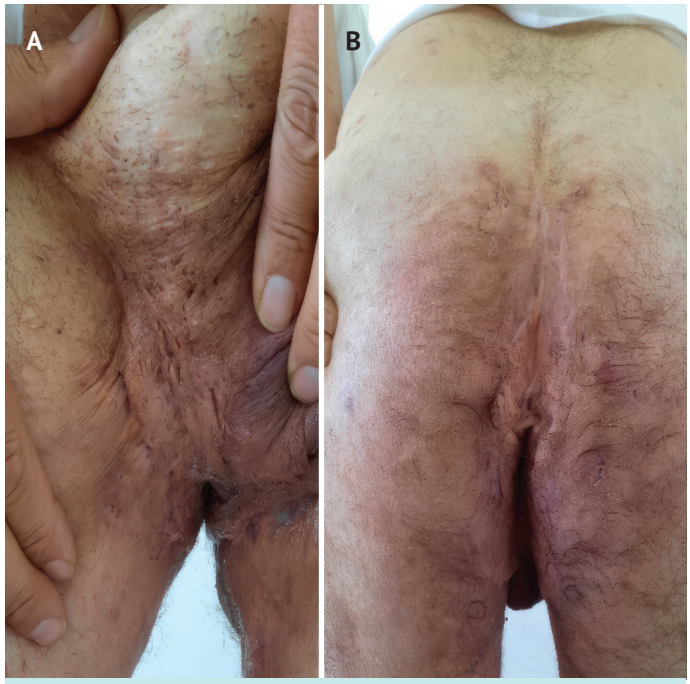

Figure 1: Inguinal (A) and hips (B) HS lesions with exudative, fistulized lesions, accompanied by sinus tract formation and scarring.

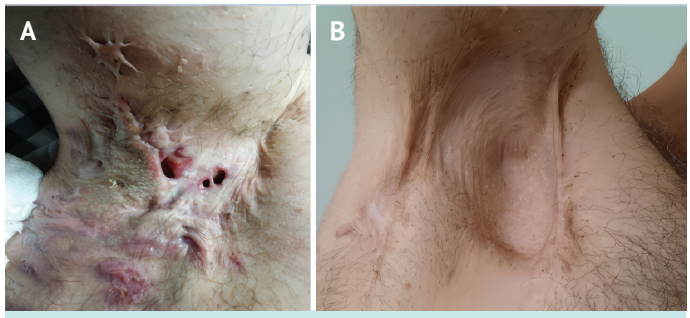

Figure 2: (A) SCC developing area in the right axilla. (B) After surgical treatment.

ence of HS, PG, and acne. Dapsone, a recommended agent for PASH syndrome, was initiated to taper off the systemic corticosteroid due to its side effects. Additionally, low-dose acitretin was added to the treatment for its antitumoral effect.

DISCUSSION

PASH syndrome, first described in 2012, is a rare autoinflammatory disease that includes PG, mild-to-severe acne, and HS. Unlike other autoinflammatory diseases, PASH syndrome is known to exclusively affect the skin and can be diagnosed based on typical clinical findings. Both PG and HS are categorized as neutrophilic dermatoses and can be either idiopathic or syndromic (2, 4). Although the etiology and pathogenesis of this syndrome have not been fully elucidated, certain genes such as PSTPIP1 are implicated, and it has been found to share a cytokine profile with other autoinflammatory diseases (1, 4). While neutrophilic dermatoses may be associated with inflammatory bowel disease or malignancy, cases of PASH syndrome coexisting with these diseases have been reported (2).

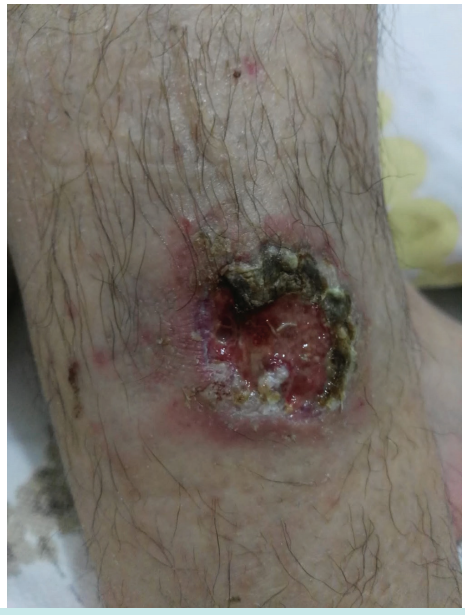

Figure 3: Pyoderma gangrenosum.

Our patient also developed cutaneous SCC and gastric carcinoma in conjunction with this syndrome.

Patients with long-standing HS are at a rare but increased risk of developing cutaneous SCC, and the prognosis in these patients is generally poorer. The development of cutaneous malignancy is thought to be secondary to chronic inflammation and epidermal hyperproliferation in the affected areas. In HS patients, distinguishing malignant transformation in chronic lesions from classical lesions can be challenging, potentially leading to a delay in diagnosis. Therefore, malignancy should be suspected in long-standing, atypical, ulcerative lesions in high-risk patients (3, 5). In our patient, invasive SCC was detected in histopathology following surgical excision for axillary HS, and subsequent metastasis studies were conducted.

Since PASH syndrome is a rare condition, there is no specific treatment protocol. Generally, treatments recommended for PG and HS are applied. Lifestyle modifications such as weight loss and smoking cessation can be beneficial. Recommended treatments include wound care, topical and intralesional treatments (corticosteroids, tacrolimus, photodynamic therapy), oral antibiotics (doxycycline, rifampin, metronidazole, amoxicillin, etc.), immunosuppressives (corticosteroids, cyclosporine, azathioprine, etc.), immunomodulators (thalidomide, dapsone, colchicine), biologics (anti-TNF, anti-IL-1, anti-IL-17, anti-IL-23), and surgical methods (1, 2). Biological treatment was not recommended for our patient due to his history of malignancy. Instead, dapsone was initiated as an immunomodulator, and acitretin was added for its antitumor effect.

In conclusion, this case is presented to highlight the importance of considering SCC in non-healing, atypical ulcers of HS patients and to emphasize the rarity and complexity of PASH syndrome.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – T.A.; Design – T.A.; Supervision – T.A.; Literature Search – T.A.; Writing – T.A.; Critical Review – T.A., P.K.

Conflict of Interest: The authors have no conflict of interest to declare.

Funding: The authors declared that this study has received no financial support.

REFERENCES

1. Cugno M, Borghi A, Marzano AV. PAPA, PASH and PAPASH Syndromes: Pathophysiology, presentation and treatment. Am J Clin Dermatol 2017;18(4):555-62. [CrossRef]
2. Huang J, Tsang LS, Shi W, Li J. Pyoderma gangrenosum, acne, and hidradenitis suppurativa syndrome: A case report and literature review. Front Med Lausanne 2022;9(1):856786. [CrossRef]
3. Lee SJ, Lim JM, Lee SH, Chung KY, Roh MR. Invasive cutaneous squamous cell carcinoma arising from chronic hidradenitis suppurativa: A case report of treatment by slow mohs micrographic surgery. Ann Dermatol 2021;33(1):68-72. [CrossRef]
4. Garcovich S, Genovese G, Moltrasio C, Malvaso D, Marzano AV. PASH, PAPASH, PsAPASH, and PASS: The autoinflammatory syndromes of hidradenitis suppurativa. Clin Dermatol 2021;39(2):240-7. [CrossRef]
5. 5-Roy CF, Roy SF, Ghazawi FM, Patocskai E, Bélisle A, Dépeault A. Cutaneous squamous cell carcinoma arising in hidradenitis suppurativa: A case report. SAGE Open Med Case Rep 2019;7(1):2050313X19847359. [CrossRef]