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A Case of Relapsing PASH Syndrome After Prior Remission With Humira

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Background: Pyoderma Gangrenosum, acne, and hidradenitis suppurative syndrome is a rare inherited disease syndrome that encompasses features of pyoderma gangrenosum (PG), nodulocystic acne, and hidradenitis suppurativa (HS). These three diseases share common mechanisms consisting of overactivation of the innate immune systems, resulting in 'sterile' neutrophilic dermatoses. Due to its pronounced cutaneous presence, PASH significantly alters quality of life. Currently, treatment options are limited and aimed at targeting the inflammatory cytokines propagating the disease.

Case: A 28-year-old female with a past medical history significant for PG, HS, and tobacco use presented to the emergency department for evaluation of worsening lower extremity wounds and draining abscesses of the left breast and axilla (Figure 1). The patient's HS had previously been well controlled on Humira until discontinuing 1 year ago after resolution of her symptoms. The patient was admitted to the hospital due to concern for necrosis and super infection. The inpatient dermatology team was consulted to evaluate for recurrence of PG. The team's assessment of the patient's presentation was consistent with PASH syndrome. An appropriate treatment plan was recommended, and the patient was discharged in stable condition with plans to follow-up with outpatient dermatology to reinstate Humira. Patient consent was obtained to use this case for educational purposes.

Conclusion: This case highlights how biologic drugs targeting key inflammatory cytokines in the innate immunity pathway can result in disease remission of PASH syndrome, while lack of such agents can result in relapse and exacerbation of the disease state. In addition to this case, we will review the overlapping neutrophilic dermatoses, PASH, PAPA, and PAPASH, as well as their reported treatment options. ■

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Figure 1. The patient demonstrated the three disease features that comprise PASH syndrome: Nodulocystic facial acne (upper left), hidradenitis suppurativa of the axilla (lower left), and pyoderma gangrenosum of the left lower extremity and left breast (upper and lower right, respectively).