

Young Man with Acne and Recalcitrant Leg Ulcers

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TO THE EDITOR

A 28 years old man came to our dermatology clinic with chief complain of painful leg ulcers from 1 year ago. During the last year, he received different kinds of antibiotics and

topical creams but he did not get any improvement. In physical examination we found nodulocystic acne with hypertrophic scars on his face, chest and proximal upper extremity (**Figure 1& Figure 2**).



Figure 1: Inflammatory lesions of acne conglobata on the chest. The nodules associated with this recalcitrant acne variant are succulent, tender and dome shaped.

In addition to this, we found inflammatory nodules with chronic drainage in the axilla and groin compatible with hidradenitis suppurativa (**Figure 3**). There were three ulcers on the pretibial area of both legs. The ulcers had purulent base with irregular, undermined borders (**Figure 4 & Figure 5**).

We made a biopsy from edge of one of the ulcers and the pathology was compatible with pyoderma gangrenosum (**Figure 6 & Figure 7**).

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Figure 2: Multiple hypertrophic scars due to severe form of nodulocystic acne (acne conglobata).



Figure 3: Inflammatory nodule and sterile abscess develops in the axilla accompanied by chronic, malodorous drainage.

The family history of pyoderma gangrenosum, hidradenitis suppurativa, crohn's disease and ulcerative colitis was not relevant in the first-degree relatives of the patient. Routine and immunological investigations were normal or

negative, except for an increased C-reactive protein and anemia. In this setting we made a diagnosis of PASH syndrome.



Figure 4: Deep ulceration with irregular border that overhangs the ulcer bed. The edge of this ulcer on the shin is undermined with a violet-gray color.



Figure 5: Two deep ulcers with overhanging, gunmetal colored border and purulent base.

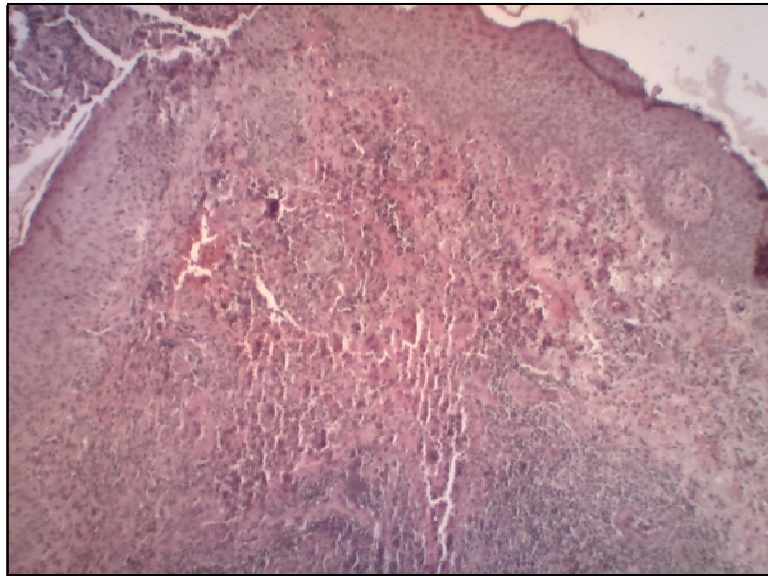


Figure 6 : The ulcer base is lined by an intense infiltrate of neutrophils.

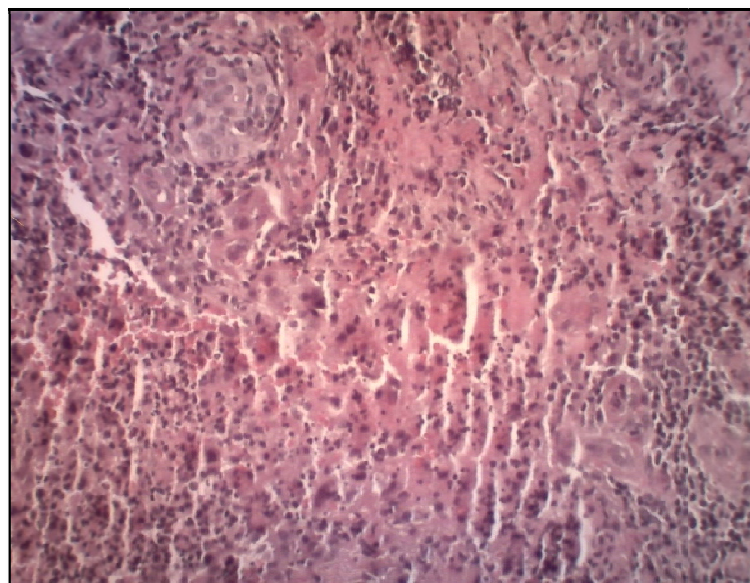


Figure 7: The center of the lesion shows a neutrophilic infiltrate with leukocytoclasia and dermolysis.

PASH syndrome is a rare

autoinflammatory skin disease, which has been recently described by Braun-Falco M. et al. [1]. This syndrome is clinically characterized by Pyoderma gangrenosum, acne, and suppurative hidradenitis, which is similar to PAPA syndrome but patients who suffer from PASH does not show arthritis and visceral involvement in contrast to patients who suffer from PAPA and aseptic abscesses syndromes. The only genetic explanation of the disease was considered as an increased number of CCTG microsatellite repeats in the *PSTPIP1* promoter region [1].

Treatment with intravenous infliximab in combination with cyclosporine and dapsone lead to sudden and prolonged improvement of the clinical symptoms [2]. One patient was treated with the interleukin (IL)-1 receptor antagonist anakinra and responded well, although without complete remission [1]. In this case, we recommended him to use Infliximab, but we lost him for follow up.

Conclusion

The term autoinflammatory skin disease comprises a range of disorders which all caused by or associated with mutations of genes regulating innate immunity. Adequate

management starts with an early diagnosis which requires recognition of rare entities such as PASH syndrome that we summarized here. Appropriate treatment may prevent undue medical expenses and minimize organ damage.

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