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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/16012
DOI URL: http://dx.doi.org/10.21474/IJAR01/16012

RESEARCH ARTICLE

PASH SYNDROME CO-EXISTING WITH RHEUMATIC HEART DISEASE AND SEVERE MITRAL VALVE REGURGITATION: A RARE CASE REPORT

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Manuscript Info

Manuscript History
Received: 10 November 2022
Final Accepted: 14 December 2022
Published: January 2023

Key words:-
PASH, Pyodermagangrenosum, Hidradenitis Suppurativa, Rheumatic Heart Disease

Abstract

Pyoderma gangrenosum, Acne, and Hidradenitis Suppurativa (PASH) is a rare inflammatory syndrome, the exact etiology and pathogenesis of which remains unknown. PASH syndrome belongs to the spectrum of Interleukin-1 driven auto-inflammatory diseases.InPatientsdiagnosed with PASH syndrome, various ways of treatment are available with individual differences in efficacy. We reporta new case of PASH syndrome that highlights the features of this rare condition and is coexisting with rheumatic heart disease and severe mitral valve regurgitation.

Introduction:-

Pyoderma gangrenosum, Acne and Hidradenitis Suppurativa (PASH)is a rare auto-inflammatory dermatosis. Distinct genetic mutations and differences in clinical phenotypes distinguish it from other auto-inflammatory disorders.[1] Mutations causing defective inflammasome function have been reported with PASH syndrome that increase IL-1 to stimulate innate immunity and neutrophil recruitment.[2] The consistent feature of the syndrome is neutrophil-rich cutaneous infiltration without evidence of infection.Several treatments for PASH syndrome exist with varying degrees of efficacy, but none has been accepted as a standard therapy. Prolonged antibiotic therapyand systemic therapy with corticosteroids, IL-1 receptor antagonists, TNF-αinhibitors, dapsone, and others are often given due to the chronic and relapsing nature of the disease.[3]

Case Report:

A 26years old male presented with multiple, painful ulcers over both legs since 6 years that are recurrent and heals with scarring and the present episode was since 6 months. He alsoreported history of recurrent draining sinuses and abscess formation in both axillae, andgroins; and also severe nodular acne leading to scarring over face since puberty. On examination, patient was ill-built and ill-nourished, had multiple oval to circular shaped ulcers, the largest measuring 6x5 cm and the smallest measuring 3x2 cm, with violaceous, and undermined borders; the floor of the ulcers is covered with red granulation tissue and scanty serous discharge. A few cribriform scars were distributed over bilateral lower limbs.Twincomedones, papules, few pustules, and scarring were noted in the bilateralaxillae.A few papules and pustules with severescarring were noted on the face. Bilateral axillary and inguinal lymphadenopathy was noted. Patient hada history of rheumatic heart disease and on 2D echocardiography, severemitral valve regurgitation is seen. On histopathological examination, from the edge of the ulcer, dense neutrophilic infiltration in the epidermis with eosinophils, lympho-plasmocytic infiltrate, and histiocytes in dermis were seen. Based onclinical and histopathological findings, we made a diagnosis of PASH syndrome and the patient was started on oral corticosteroids and doxycycline capsules.

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Figure 1:- Pyoderma gangrenosum ulcers with violaceous borders over lower limbs and twin comedones, pustules and scarring over left axillae.

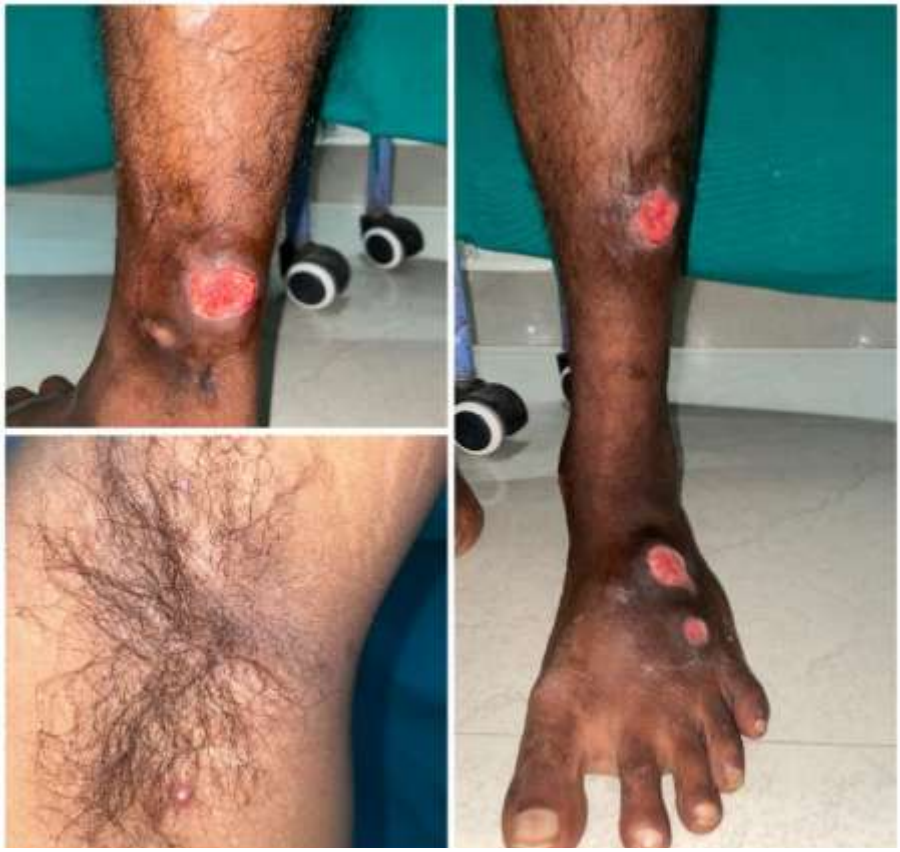
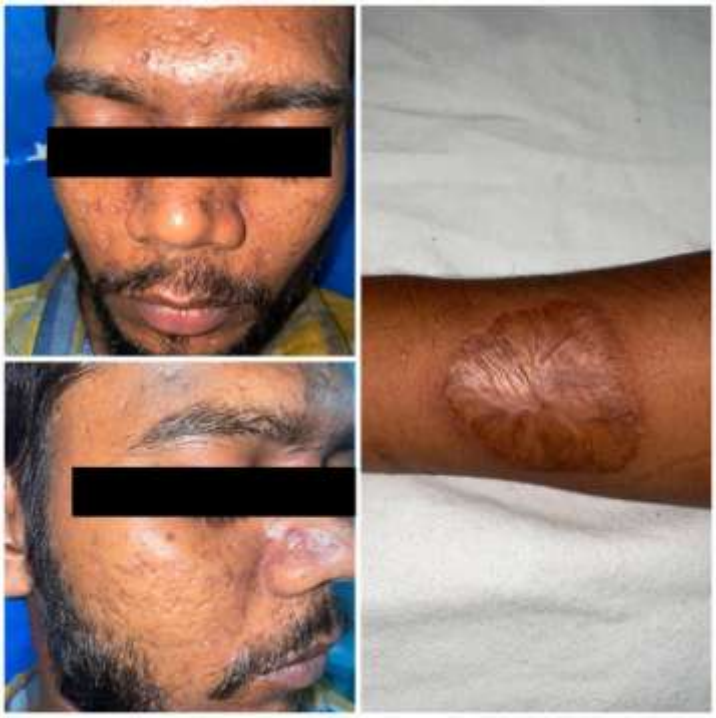


Figure 2:- Patient with severe scarring over the face and cribriformscarring over left forearm.



Discussion:-

PASH syndromeis a new entity, first described in 2012. However, Hsiao et al. had previously describedthis clinical phenotype.[4]PASH syndrome is a heterogeneous disease whose genetic background is still unclear, but mutations in PSTPIP1,PSENEN and NCSTN genes have been identified in a portion of patients with PASH.In the above case report, the patient had PASH syndrome coexisting with rheumatic heart disease and severe mitral valve regurgitation; the exact association between them cannot be explained. The probablerelation was attributed to recurrent secondary bacterial infections of pyoderma gangrenosum ulcers that ultimately led to rheumatic heart disease with severe mitral valve regurgitation.As thereported cases are limited, there are no standard treatment protocols available. Often, treatment is directed at the management of pyoderma gangrenosum and hidradenitis suppurativa, with corticosteroids, tacrolimus, oral antibiotics, immunosuppressants like cyclosporine, and other drugs like thalidomide, dapsone, biologicals like TNF-α inhibitors, IL-1 inhibitors and IL-17 inhibitors, and various surgical procedures.[5] In the above case, treatment was started with oral corticosteroids at a dose of1mg/kg of prednisolone, with good improvement of lesions along with wound care, andcap.doxycycline100mg OD.The patient was advised intensive lifestyle modifications focusing on stress reduction and increase in physical activity.

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