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(CASE REPORT)

A Rare Association of Pyoderma Gangrenosum and Hyper-IgE Syndrome (Job-Buckley) in a Child: A case report

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Abstract

Background: Hyper-IgE syndrome (Job-Buckley syndrome) is a rare primary immunodeficiency caused by mutations in the STAT3 gene. It is characterized by recurrent severe infections, chronic eczema-like skin lesion, and markedly elevated serum IgE levels. Pyoderma gangrenosum (PG) is an inflammatory neutrophilic dermatosis that is rare in children.

Case presentation: We report the case of a 9-year-old boy, followed for dilated cardiomyopathy and diagnosed with Hyper-IgE syndrome since 2021, treated with sulfamethoxazole-trimethoprim. He presented with painful chronic ulcers on both lower limbs, evolving over the course of one year. Clinical examination revealed multiple deep, painful ulcers over the knees and ankles with violaceous raised borders and granulating bases, associated with erythematous, scaly papules resembling atopic dermatitis. Microbiological and laboratory investigations were negative. Histopathological analysis revealed a neutrophilic dermatosis. A diagnosis of pyoderma gangrenosum was made, and treatment with systemic corticosteroids (1 mg/kg/day) was initiated, resulting in favorable clinical evolution without relapse.

Conclusion: To our knowledge, this is the first reported case of pyoderma gangrenosum associated with Job-Buckley syndrome. This case highlights the importance of considering PG in immunocompromised children presenting with chronic ulcers.

Keywords: Hyper-Ige Syndrome; Job-Buckley Syndrome; Pyoderma Gangrenosum; Neutrophilic Dermatosis; Pediatric Dermatology.

1. Introduction

Hyper-IgE syndrome (Job-Buckley) is a rare primary immunodeficiency estimated to affect approximately 1 in 1 million individuals annually[1]. It presents as a triad of recurrent severe infections, eczema-like chronic dermatitis, and markedly elevated serum IgE levels (>2000 IU/mL), often accompanied by eosinophilia and normal levels of other immunoglobulins[2]. Cutaneous features often resemble atopic dermatitis and may appear during infancy, adolescence, or adulthood[3][4].

Pyoderma gangrenosum (PG) is an inflammatory neutrophilic dermatosis that is rarely observed in pediatric populations. It typically manifests as painful, rapidly progressing cutaneous ulcers often mistaken for infectious or ischemic lesions[5]. PG has been reported in association with inflammatory bowel disease, hematological disorders, vasculitis, immunodeficiencies, and autoinflammatory syndromes[6]. However, no cases of PG associated with Hyper-IgE syndrome have been previously reported.

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2. Case Report

A 9-year-old boy with a known history of dilated cardiomyopathy since the age of one and diagnosed with Hyper-IgE syndrome in 2021, under prophylactic treatment with sulfamethoxazole-trimethoprim, presented with painful chronic ulcers of both lower limbs evolving for the past year.

2.1. Clinical findings

Examination revealed multiple deep ulcers overlying the knees and ankles, with violaceous, raised borders and red granulating bases. The lesions were painful and grossly rounded. Additionally, the patient exhibited erythematous, scaly papules and plaques involving the face, skin folds, and trunk—consistent with an atopic-like dermatitis.

2.2. Investigations

Bacteriological cultures and blood tests, including inflammatory markers, were negative. Skin biopsy revealed a neutrophilic dermatosis without vasculitis or infectious agents, consistent with pyoderma gangrenosum.

2.3. Treatment and Outcome

Systemic corticosteroid therapy was initiated at 1 mg/kg/day of prednisone. Treatment lasted 5 months with gradual tapering. The patient showed significant clinical improvement with complete ulcer healing and no relapse reported upon tapering and cessation of corticosteroids.

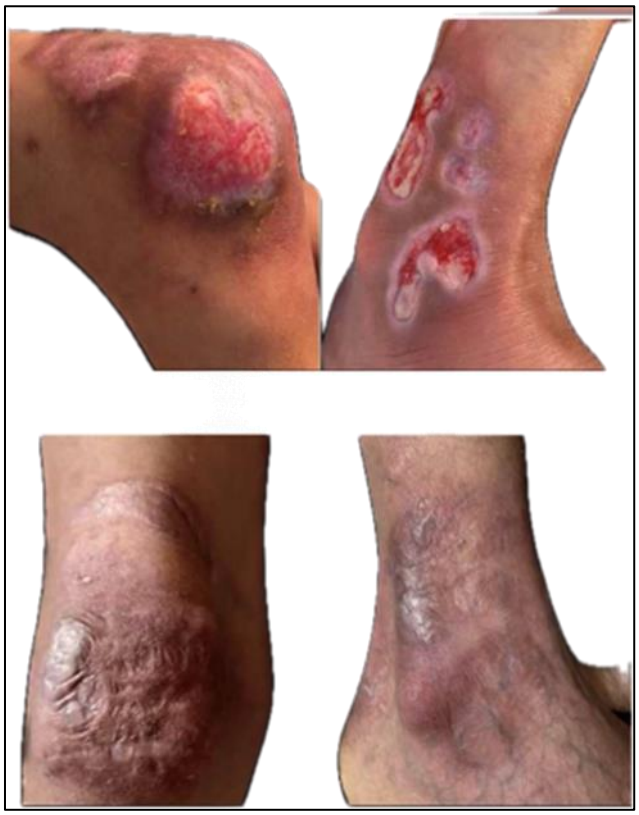

Figure 1 Multiple deep ulcers overlying the knees and ankles

3. Discussion

Hyper-IgE syndrome involves immune dysregulation due to defective STAT3 signaling, impairing Th17 cell function and neutrophil recruitment. This predisposes patients to unusual infections and inflammatory responses. Although PG has been documented in association with various immune defects such as LAD-1 deficiency, RAG1 mutations, and CVID, no previous association with Job-Buckley syndrome has been published to our knowledge [7].

This case suggests a possible link between immune dysregulation in Hyper-IgE syndrome and the emergence of severe inflammatory dermatoses such as PG. Accurate diagnosis of PG is crucial to avoid inappropriate antibiotic use or surgical interventions, and systemic corticosteroids remain the cornerstone of effective treatment in pediatric cases.

4. Conclusion

This case represents the first documented association of pyoderma gangrenosum with Hyper-IgE syndrome. It underlines the need to consider PG in children with immunodeficiencies presenting with chronic, non-healing ulcers, and it confirms the efficacy of systemic corticosteroids in achieving remission.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare no conflicts of interest.

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Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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