Case Study

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A Case Study on Childhood Pemphigus Vulgaris

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Abstract

Pemphigus vulgaris is a rare autoimmune blistering disorder characterized by the formation of painful, fluid-filled blisters on the skin and mucous membranes. This condition is caused by the production of autoantibodies that target desmoglein's, which are proteins responsible for maintaining the adhesion between skin cells. The breakdown of cell adhesion leads to the formation of blisters and erosions, causing significant morbidity. Pemphigus vulgaris often presents with oral lesions, but it can affect other mucosal surfaces and the skin. Diagnosis involves clinical examination, histopathology, and immunofluorescence studies. Management typically includes systemic corticosteroids and immunosuppressive agents, although treatment strategies may vary based on the severity of the disease. Despite advancements in understanding the pathophysiology of pemphigus vulgaris, challenges remain in achieving long-term remission and minimizing treatment-related side effects. Ongoing research aims to uncover novel therapeutic approaches and improve the overall quality of life for individuals affected by this autoimmune blistering disorder.

***Keywords*:** pemphigus vulgaris, auto immune blistering disorder, desmoglein's, immunosuppressive agents.

Introduction

Pemphigus vulgaris (PV) stands as a formidable challenge in the realm of autoimmune dermatological disorders, casting a shadow over the delicate equilibrium that maintains the integrity of the skin and mucous membranes. Characterized by the relentless formation of painful blisters and erosions, PV is a rare yet debilitating condition that arises from the immune system's misguided assault on the intricate network of proteins responsible for cell adhesion. The disorder targets desmoglein's, key components in the structural integrity of skin cells, leading to the breakdown of cell-to-cell connections and the subsequent manifestation of mucocutaneous lesions.

As an autoimmune blistering disease, PV poses significant clinical complexities, often surfacing with oral involvement as a hallmark presentation. Beyond its impact on oral mucosa, PV can extend its reach to affect various mucosal surfaces and the skin, causing substantial morbidity and impairing the patient's quality of life. The diagnosis of PV necessitates a comprehensive approach, integrating clinical examination, histopathology, and immunofluorescence studies to unravel the intricate immunological mechanisms at play.

Epidemiology

the prevalence of PV in the general population is estimated to be around 1 to 16 cases per million people per year. PV typically manifests in adults, with an average age of onset ranging from 40 to 60 years. Paediatric cases represent a small fraction of the overall incidence of PV. Pemphigus Vulgaris (PV) is an extremely rare autoimmune blistering disorder, and childhood-onset cases are even more uncommon.

Clinical Presentations

- painful oral ulcers
- blisters on the skin
- Mucous Membrane Involvement

- Pain and Discomfort
- Pruritus (Itching)
- Nail Involvement
- Ocular Involvement
- Scalp Involvement.

Diagnostic Challenges

Diagnosing childhood PV requires a combination of clinical, histopathological, and immunological assessments. Skin biopsies are essential for confirming the diagnosis, typically revealing intraepidermal acantholysis. Immunofluorescence studies often show the deposition of immunoglobulins within the epidermis. However, obtaining accurate biopsies from children, especially from oral mucosa, can be challenging due to their limited cooperation and the discomfort associated with the procedure.

Management

The management of childhood PV involves a multidisciplinary approach, including dermatologists, paediatricians, and, in severe cases, immunologists and rheumatologists. Systemic corticosteroids remain the primary treatment, often in conjunction with immunosuppressive agents such as azathioprine or mycophenolate mofetil. The challenge lies in balancing the need for effective control of the disease with minimizing the potential long-term side effects of immunosuppressive therapy in a developing child.

Case Study

A 9-year-old male patient admitted in dermatology department with the chief complaints of fluid lesions all over the body for 10 days. Patient initially developed oral erosions and followed by extensive raw areas all over the body.

Table: 01 Complete Blood Picture:

Haemoglobin	12.4g/dl
Pcv	3.8
Plt	5.03
Tc	26,800
ESR	20 mm/hr

Table: 02 Complete Urine Picture

Albumin cells	2+
Pus cells	6-8
Epithelial cells	8-10

Treatment

Patient was given with medication injection ceftriaxone (500mg), Tablet dapsone (25mg), tablet cetirizine (10mg), 2%fusidic acid ointment and 0.1% betamethasone ointment.

Conclusion

Although childhood Pemphigus Vulgaris is a rare condition, it poses significant challenges in diagnosis and management. A collaborative and multidisciplinary approach involving dermatologists, paediatricians, and other specialists is essential for optimal care. Further research is needed to better understand the unique aspects of childhood-onset PV, paving the way for improved diagnostic tools and targeted therapeutic interventions.

Discussion

Pemphigus vulgaris (PV) stands as a challenging autoimmune blistering disorder, necessitating a comprehensive understanding of its pathophysiology and an evolution in therapeutic approaches. The disease's complexity lies in its intricate interplay of immunological factors, genetic predispositions, and environmental triggers. One key aspect to consider is the pivotal role of autoantibodies targeting desmoglein's, integral proteins responsible for maintaining cell adhesion in the epidermis. The breakdown of this adhesion results in the formation of painful blisters and erosions on the skin and mucous membranes. However, the mechanisms triggering the production of these autoantibodies remain an area of active research. The clinical presentation of PV, often marked by oral lesions, underlines the importance of early diagnosis. Clinicians must navigate the challenges of distinguishing PV from other autoimmune blistering diseases,

emphasizing the need for a multi-faceted diagnostic approach, including clinical examination, histopathology, and immunofluorescence studies.

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