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Original Research Paper

Dermatology

PEMPHIGUS FOLIACEUS MASQUERADING AS PEMPHIGUS VEGETANS: A DIAGNOSTIC DILEMMA

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ABSTRACT Pemphigus is a group of autoimmune blistering disorders which is characterised by intraepidermal split. Pemphigus foliaceous being one of the most common and debilitating conditions second to pemphigus vulgaris. We report a patient presenting with atypical clinical manifestation and with uncommon distribution of this entity thus causing a diagnostic dilemma of flexural psoriasis and lichen simplex chronicus .. The histologic findings of biopsy showed features of Pemphigus foliaceus.

KEYWORDS :

INTRODUCTION

Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) are the two major forms of pemphigus accounting for more than 90% among the pemphigus group of disorders in the clinical practice. In which PF is a superficial type of pemphigus with crusting, erosions and scaling predominantly over seborrheic areas of body with histology showing subcorneal acantholysis. Whereas inverse psoriasis also known as intertriginous or skin fold psoriasis is a rare type of chronic inflammatory skin disease involving epidermis presenting as erythematous plaques with poor or non desquamation in flexural areas . Lichen simplex chronicus being a chronic neurodermatitis presenting as dry, patchy areas of thick scaly skin usually affecting self accessible areas such as scalp, head, neck, hands and genitals.

Case Report

A 54-year-old male presented with complaints of raised skin lesion over the groin for past 3 years associated with itching for which he has been treated with topical, oral and natural medications with less response. On examination multiple well to ill-defined hyperpigmented plaques over groin. The differentials were pemphigus foliaceus, flexural psoriasis, and lichen simplex chronicus. On investigation KOH mount was found to be negative ruling out any fungal infection. Punch biopsy of 5mm was taken and was sent to histopathological examination which showed bulla in the superficial part of epidermis, the granular layer forming the roof of the bulla containing acantholytic cells, eosinophils and neutrophils. The epidermis underlying the bulla shows acanthosis with irregular elongation of rete ridges. The upper dermis is infiltrated by numerous eosinophils and neutrophils. The histopathological features are more in favour of pemphigus foliaceaus. Patient was started on systemic corticosteroids for which he responded.

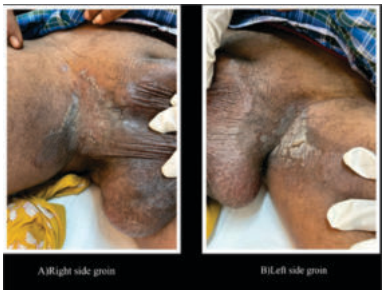

Figure: 1 – Clinical Picture

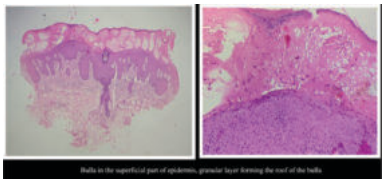

Figure 2 : Histopathological Picture

DISSCUSSION

Pemphigus foliaceus (PF), or superficial pemphigus,' rare immunobullous disease a pemphigus group of blistering disorders which usually affects persons in their fifth to seventh decade. It is characterized by superficial blisters that rupture and leave shallow erosions. PF initially present as moist papules or superficial bullae which readily rupture and patient usually present with moist erosions with thin delicate crusting termed as bran like/ cornflakes like. They may also present with small vesicles over the borders of lesions(Malik et al., 2021). They usually involve seborrheic areas including face, scalp, upper trunk but can also involve beyond standard sebrroheic areas to diffusely involve larger areas of skin in severe cases(Griffiths et al., 2023). PF rarely develop into erythroderma(Galambrun et al., 1997). They have skin fragility with nikolsky sign being positive. Histologically there is acantholysis with occasional dyskeratosis within the granular cell layer. The Direct immunoflurocence being the confirmatory test shows IgG and often C3 deposited on the surface of keratinocytes throughout the epidermis in perilesional skin . IgG1 and IgG4 are the most common subclasses; IgM and IgA are present less frequently than IgG.. Pemphigus foliaceus rarely affects mucosa. Pemphigus foliaceous has three sub types namely: 1) classic or sporadic, 2) pemphigus erythematosus, 3) endemic PF (fovo selvagem). These three entities has similar histological features(Ramam et al., 2023).

The treatment approach for pemphigus mainly relies on immune system suppression to prevent new lesion formation and heal existing bullous skin or mucous lesions, while minimizing serious treatment side effects. For all forms of pemphigus, the literature supports corticosteroids as a first-line therapy, with or without adjuvant therapies. Ultimately, for all forms, the most important factor is treating quickly and effectively and to taper corticosteroids to avoid recurrence (Malik et al., 2021).

Inverse psoriasis (intertriginous psoriasis) present usually in

self accessible areas such as genitalia. It usually presents as erythematous, irregular, well demarcated thin symmetrical plaques with poor or no scaling with few fissures and rhagades. It also produces more plaques through the Koebner phenomenon and local lichenification . The histopathological features shows Kogoj's and Munro-Sabouraud's collections of neutrophils, thickening of the malpighian layer, hypogranulosis, hyperkeratosis, parakeratosis, and elongation of the papillae similar to psoriasis.(Guglielmetti et al., 2012).

Lichen simplex chronicus usually present as hypertrophic epidermis due to habitual scratching or rubbing of a specific area of the skin often due to itch scratch cycle .On histopathology it shows features of hyperkeratotic plaque with foci of parakeratosis, prominent granular cell layer, elongated and irregularly thickened epidermal rete, acanthosis, pseudoepitheliomatous hyperplasia, papillary dermal fibrosis, mild spongiosis, and perivascular as well as interstitial inflammation with histiocytes, lymphocytes, and occasional eosinophils in the superficial dermis(Charifa et al., 2024).

CONCLUSION

Early diagnosis and treatment of Pemphigus foliaceous is essential to prevent morbid complications. Hence it is necessary to arrive at diagnosis even in the milieu of atypical presentation.

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