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Original Research Paper

Dermatology

MUCHA-HABERMAN DISEASE : A RARE CLINICAL VIGNETTE

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ABSTRACT Pityriasis lichenoides is an uncommon condition, previously included in the parapsoriasis group, now belongs to the papulo-squamous group of disorders. It appears in two forms the acute variant also known as pityriasis lichenoides et varioliformis acuta (PLEVA); and the chronic variant or pityriasis lichenoides chronica (PLC). We report a case of a 25 year old male who presented with multiple, well defined, hyperpigmented, scaly papules and plaques present all over body except face, scalp and mucous membrane since 2 years. Histopathological examination revealed interface dermatitis, parakeratosis along with perivascular infiltrates and exocytosis of erythrocytes and lymphocytes indicating the diagnosis of Pityriasis lichenoides et Varioliformis acuta.

KEYWORDS : Pleva, Mucha Haberman Disease, Papulosquamous Disorder.

INTRODUCTION

Pityriasis lichenoides (PL) is a chronic recurrent inflammatory disorder belonging to the papulosquamous group, now considered to be lymphocytic vasculitis[1], is caused by an unknown aetiology. Lack of any definite aetiology and absence of vessel wall damage makes this condition to be included in any diagnostic classifications[2].

It is of two types, acute and chronic, where in the PLEVA (Pityriasis lichenoides et Varioliformis acuta) belongs to the acute type. Though the aetiology is postulated as unknown, many triggering factors have been found to initiate such as; an inflammatory reaction triggered by infectious agents(toxoplasma, cytomegalovirus, parvovirus, adenovirus and Epstein Barr virus[3]), an immune- complex mediated hypersensitivity, or an inflammatory response secondary to T-cell dyscrasia[4].

PLEVA, a male dominant condition is seen in all ages with more common occurrence in paediatric age group. Though the lesions are frequently self-healing; recalcitrance may occur and the resolving papules may misdiagnose. A differential diagnosis of PLC, Secondary syphilis, Pityriasis rosea and Guttate psoriasis should be considered in such cases and skin biopsy should be appreciated. Keeping the above facts in mind, we report a case of PLEVA which may be misdiagnosed.

Case Report

A 25 year old male, with congenital hearing and mute difficulties, was brought to the OPD by his sibling for the complaints of on and off dark lesions over the body for past 2 years. Lesions were associated with itching.

He had history of treatment with NBUVB for the similar complaints and was diagnosed as PLC clinically a year back, with no improvement. there was an episode of infection-Pharyngotonsillitis few days prior to the onset of the eruption. His personal and family history were non-contributory.

Dermatological examination revealed papules and plaques with fine scaling few with a central punctum and ulceration, with overlying crusts on the trunk, bilateral extremities and gluteal region. Few macules representing post inflammatory hyperpigmentation was present.

Mucosal membranes, palms, soles, hair, and nails were unremarkable. Differential diagnosis of PLEVA, Pityriasis rosea and Guttate psoriasis was arrived.

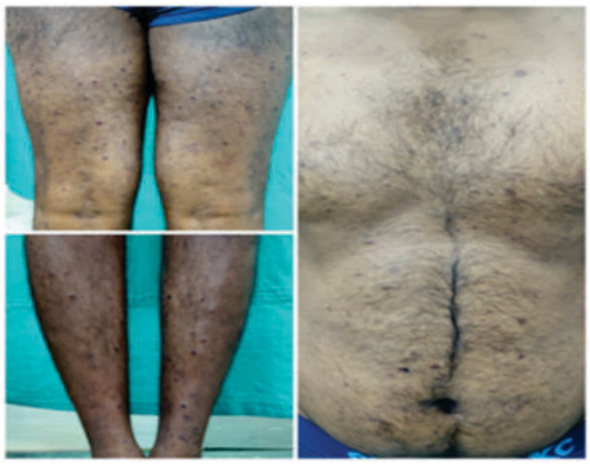

Figure 1 : Multiple papules with red-brown crusts and few scaling present over the anterior aspect of trunk and bilateral lower legs.

Histopathological examination revealed mild parakeratosis, regular acanthosis, interface dermatitis of lymphocytes admixed with some extravasated erythrocytes. With the clinical and pathological corelation final diagnosis of PLEVA was arrived. Patient was treated with topical and systemic steroids for 4 weeks, Doxycycline 200mg/day for 2 weeks and lesions showed regression after a month.

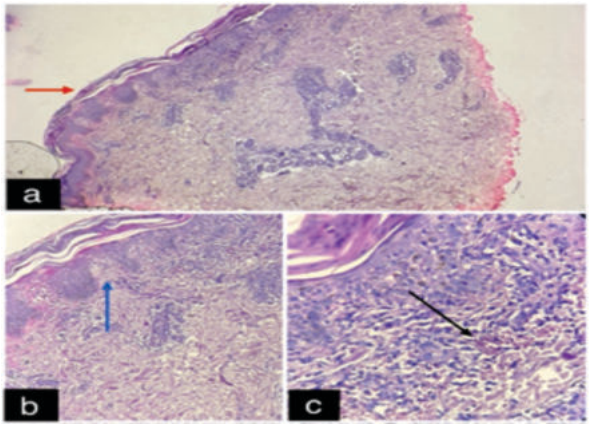

Figure 2 :
Sub Figure a and b : 4x and 10 x view respectively shows parakeratosis (RED arrow), perivascular and dense,

predominantly lymphocytic infiltrate in the papillary dermis which extends into the reticular dermis in a wedge shaped pattern (BLUE arrow).

Sub figure c : scattered necrotic keratinocytes are seen in addition to parakeratosis. Black arrow indicates exocytosis of erythrocytes.

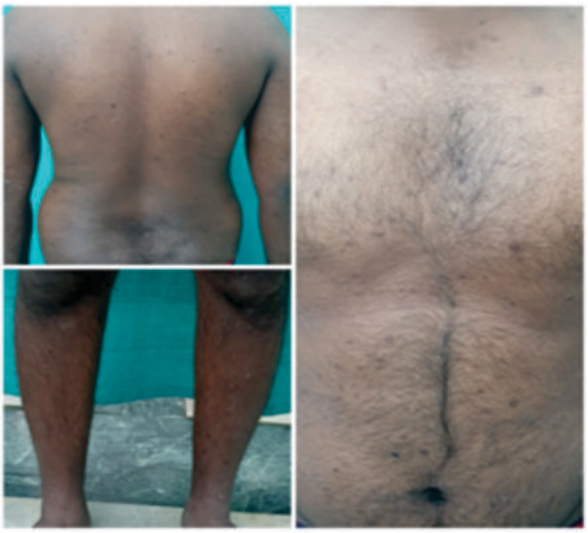

Figure 3: Lesions resolving with post inflammatory hyperpigmentation after 2 weeks of treatment.

DISCUSSION

Mucha-haberman disease also known as, PLEVA was first named by Haberman.R in 1925. It is characterized by cutaneous inflammatory rash which is the presence of diffuse red-brown papules in various stages with a mica-like scale on more established lesions.

PL is more common in paediatric age group but can also be seen in all age groups with a male dominance. PLEVA presents with acute to subacute, erythematous macular-papular rash which progresses rapidly to polymorphic lesions of different stages.

Though the condition is self-limiting, the uncertainty in aetiology leads to difficulty in management. Besides infectious agents it is also triggered by drugs which include tetracycline, erythromycin, methotrexate, calciferol, chinoline and acridine derivates, cyclosporine, intravenous gamma globulin, and retinoids[5].

It is typically characterized by erythematous macules that quickly evolve into papules with fine scales. When the scale thickens, it may become free at the periphery but centrally attached. The papule often has a central punctum which becomes vesiculopustular, undergoes haemorrhagic necrosis, and becomes ulcerated, with overlying red-brown crusts. Varioliform scars and post inflammatory hyper- and hypopigmentation may occur[6]. They occur predominantly on the trunk followed by extremities with usual sparing of mucous membranes[7]. PLEVA Symptoms include burning and pruritus. Histopathology in PLEVA features with parakeratosis, perivascular and dense, predominantly lymphocytic infiltrate in the papillary dermis which extends into the reticular dermis in a wedge shaped pattern. The infiltrate may obscure the DEJ with vacuolar alteration of the basal layer, marked exocytosis of lymphocytes and erythrocytes resulting in a variable degree of epidermal necrosis[8].

The differential diagnosis for this condition includes Lymphomatoid papulosis, Pityriasis rosea, insect bites, subcutaneous eczematous dermatitis, transient acantholytic dermatoses, and dermatitis herpetiformis[7]. These are mainly differentiated by Clinical features such as Papules with fine scales and central punctum evolving to ulceration resulting in red-brown crusts and histopathological findings of Lymphocytic infiltrate extending as wedge shaped pattern from papillary to reticular dermis and marked exocytosis of erythrocytes and lymphocytes. Further confirmation can be done with immunohistochemistry markers, showing CD8+/CD30– phenotype.

Treatment with topical corticosteroids/tacrolimus, systemic steroids and antihistamines provide symptomatic relief, but they do not alter the course of the disease. There are recent reports of the efficacy of etanercept, oral bromelin, and photodynamic therapy. In our case we did the earlier option and with systemic steroids with keeping the routine investigations a check. We found a remarkable improvement and the lesions healed with post inflammatory hyper pigmentation.

CONCLUSION

We conclude the article by emphasizing the rarity of this disease which poses difficulties such as misdiagnosis and mistreatment resulting in the necessity of confirmatory tests such as skin biopsy. Early diagnosis of this condition with prompt treatment results in the improvement of quality of life of the patient along with cosmetic betterment by avoiding post inflammatory changes.

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