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Pityriasis lichenoides et varioliformis acuta in 35 years old male – A rare case report

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ABSTRACT

Pityriasis lichenoides (PC) is a papulo-squamous disorder of unknown etiology with acute and chronic presentation. Pityriasis lichenoides et varioliformis acuta (PLEVA) is the acute form of the disease. In many cases of the PLEVA, the diagnosis is made after clinico-pathologic correlation. Histopathological examination helps greatly in the diagnosis of this condition. Here, we are presenting a case of 35 years old male, hospitalized for multiple excoriated macula-papular skin lesions varying from 2 mm to 14 mm all over the body. Here, we want to remind of this rare entity which may present difficulties in diagnosis.

Keywords: Pityriasis lichenoides, PLEVA, Maculo-papular skin lesions.

1. INTRODUCTION

Pityriasis lichenoides (PC) is a papulo-squamous disorder of unknown etiology with acute and chronic presentation (Habermann, 1925). Pityriasis lichenoides chronica (PLC) is considered as mild and chronic form of the disease while Pityriasis lichenoides et varioliformis acuta (PLEVA) is the acute form of the disease (Willemze et al., 1985). PLEVA, otherwise known as Mucha-Habermann disease, is an uncommon cutaneous inflammatory disorder. The precise

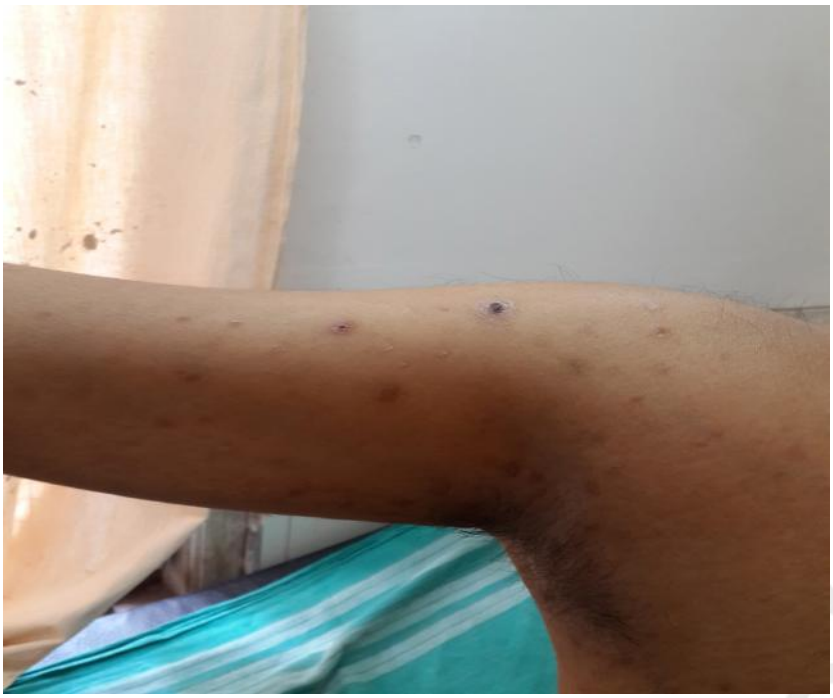

Figure 1

Multiple excoriated papules along with presence of post inflammatory hyper pigmentation

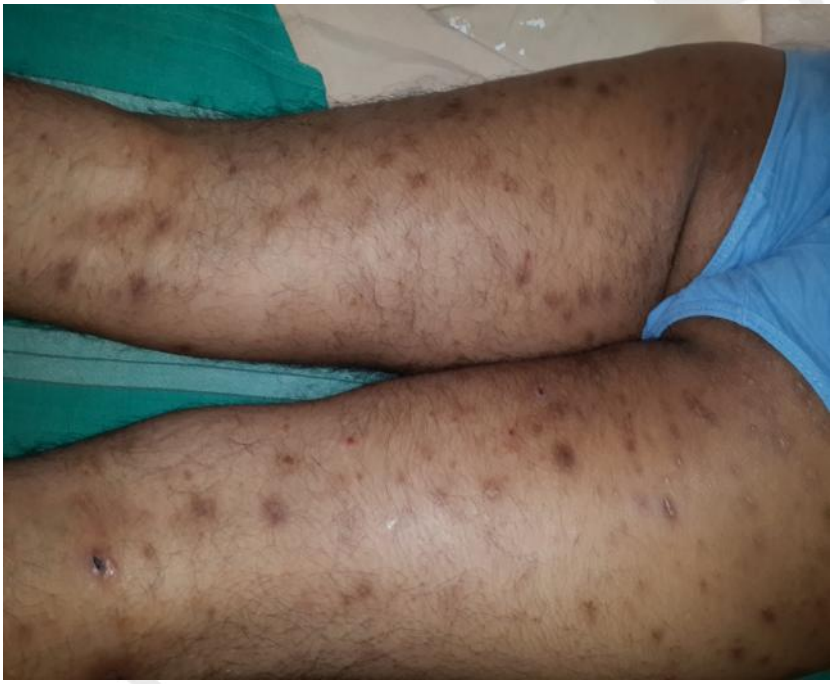

Figure 2

Multiple papules with central necrotic skin and crusting on thigh

incidence and prevalence are not known. PLEVA may occur at any age (including infants) (Hoshina et al., 2007), but most frequently occurs in children and young adults. The cutaneous lesions in PLEVA are usually asymptomatic, but may be pruritic and may heal with scarring. These patients rarely have systemic signs. In many cases of the PLEVA, the diagnosis is made after clinic-pathologic correlation.

2. CASE REPORT

A 35 years old male patient came to skin OPD with a history of skin lesions all over body since 10-11 years. There was history of progressive increase in the number of lesions. On examination, patient was having multiple excoriated papules of size varying from 2 mm to 14 mm all over the body. Many papules were with central necrotic skin and crusting on thigh and back region. Post inflammatory hyper pigmentation is also present (Figures 1 & 2). Patient denied of taking any medications and there was no apparent episode of any infection before the onset of the eruption. No history of any joint pain or fever. All the hematological and serological investigations in the form of full blood cell count with differential, liver, and renal studies, serum protein electrophoresis, serology for hepatitis B virus, hepatitis C virus, HIV were negative/ normal. Punch biopsy of the lesion was taken and sent to the histopathology department. Microscopic examination showed a perivascular and dense, band like, predominantly lymphocytic infiltrate in the papillary dermis that extends into the reticular dermis in a wedge-shaped pattern (Figure 3). There was also presence of intercellular and intracellular edema leading to variable degree of epidermal necrosis. The overlying cornified layer shows parakeratosis and a scaly crust with neutrophils (Figure 4). There was also presence of papillary dermal edema, endothelial swelling, and extra-vasated erythrocytes along with lymphocytic vasculitis (Figure 5). The overall features suggestive of Pityriasis lichenoides et varioliformis acuta. Oral erythromycin (1 g per day), topical corticosteroids, and antihistamines were prescribed. Patient came for follow up after 4 months with only residual macular pigmented lesions.

3. DISCUSSION

In 1916, Mucha and in 1925, Habermann reported an acute form of pityriasis lichenoides characterized by the abrupt onset of papulo vesicular eruptions and described it as pityriasis lichenoides et varioliformis acuta (PLEVA) or Mucha-Habermann disease. The etiology of PLEVA remains unknown. Several hypotheses have been proposed to explain this dermatitis: an inflammatory reaction triggered by infectious agents, an inflammatory response secondary to T-cell dyscrasia, or an immune complex-mediated hypersensitivity (Fernandes et al., 2010). The lesions of PLEVA consist of a fairly extensive eruption, present mainly on the trunk and proximal extremities. It is characterized by erythematous papules that develop into papulo necrotic, occasionally hemorrhagic or vesiculo-pustular lesions that resolve within a few weeks,

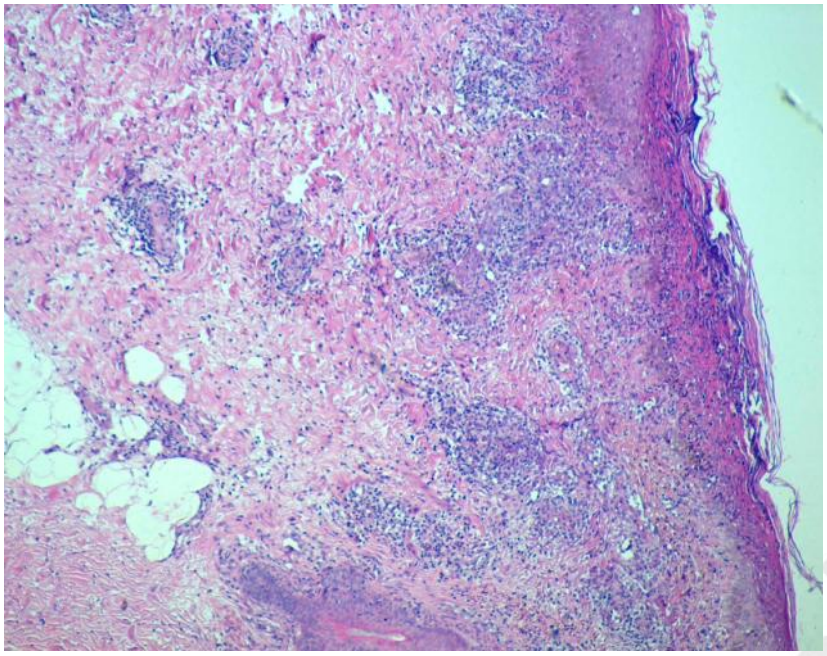

Figure 3

Microphotograph shows lymphocytic infiltrate in the papillary dermis that extends into the reticular dermis in a wedge-shaped pattern. [H & E, 4 X]

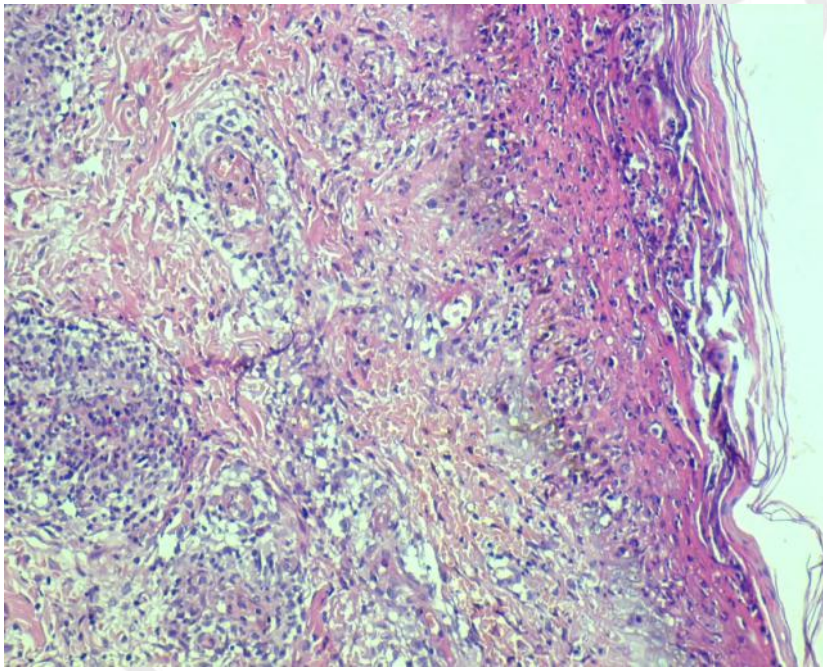

Figure 4

Microphotograph shows the cornified layer parakeratosis and a scaly crust with neutrophils. [H & E, 10 X]

usually with little or no scarring. In occasional patients, some lesions increase in size to necrotic ulcers of 1 to 2 cm in diameter healing with an atrophic or varioliform scar (Lever's Histopathology). Very rarely, patients with PLEVA have a sudden, severe flare-up of their disease, characterized by innumerable coalescent necrotic ulcerations associated with high fever and systemic manifestations (Maekawa et al., 1994; De cuyper et al., 1994; Lopez-Estebaranz et al., 1993).

During the course of PLEVA, patients can develop scaly, red-brown papules that are consistent with pityriasis lichenoides chronica (PLC) alongside lesions consistent with PLEVA, and occasionally, patients completely transition from PLEVA to PLC (Fernandes et al., 2010). These observations have been used to support the concept that PLEVA and PLC represent a spectrum of a single disease.

Histologically, there is a perivascular and dense band-like, predominantly lymphocytic infiltrate in the papillary dermis that extends into the reticular dermis in a wedge-shaped pattern. The infiltrate obscures the dermal-epidermal junction with pronounced vacuolar alteration of the basal layer, marked exocytosis of lymphocytes and erythrocytes, and intercellular and intracellular edema leading to variable degrees of epidermal necrosis. Ultimately, erosion or even ulceration may occur. The overlying cornified layer shows parakeratosis and a scaly crust with neutrophils in the more severe cases (Willemze et al., 1985; Muhlbauer et al., 1984).

Occasionally, the histologic picture of PLEVA can be mimicked by other diseases such as lymphomatoid papulosis (the condition most commonly mistaken for PLEVA), arthropod bite reactions, varicella, Gianotti-Crosti syndrome, erythema multiforme, pityriasis rosea, guttate psoriasis, vasculitis, and secondary syphilis (Bowers et al., 2006).

The presence of a deeper inflammatory infiltrate, extensive epidermal necrosis, and absence of intra epidermal spongiotic microvesicles may help distinguish PLEVA from pityriasis rosea and subacute eczematous dermatitis. Numerous eosinophils in a vertically oriented dermal infiltrate are more commonly seen in insect bites (Hood et al., 1982). Thorough histopathological examination is quite helpful for ruling out all the differential diagnosis of PLEVA.

4. CONCLUSION

In conclusion, we want to remind of this rare entity which may present difficulties in diagnosis. Various diagnostic modalities are helpful for the diagnosis although it should be confirmed by histopathological examination of biopsy specimen because it is the gold standard for final diagnosis.

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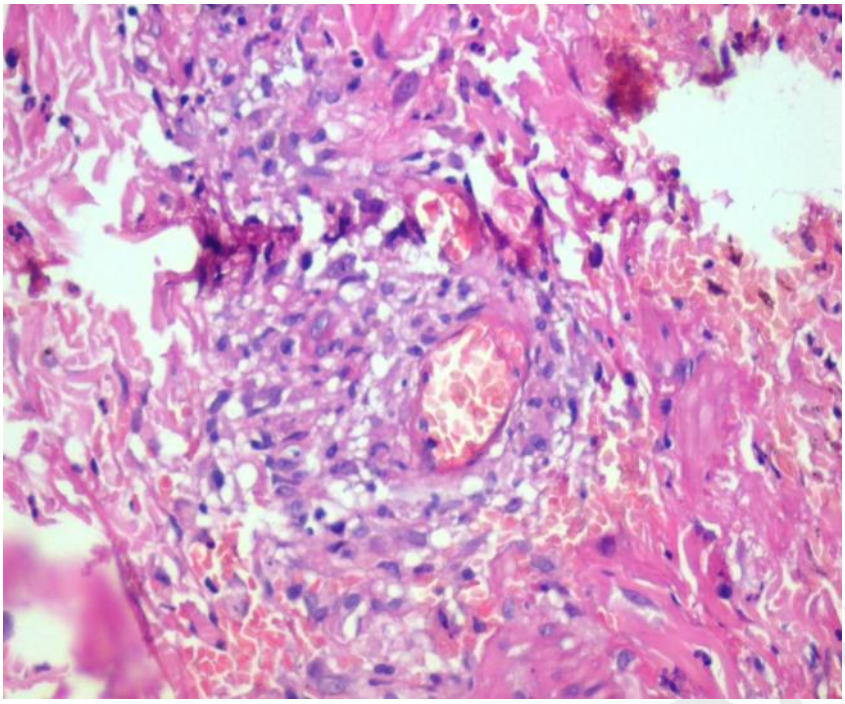

Figure 5

Microphotograph extravasated erythrocytes along with lymphocytic vasculitis. [H & E, 40 X]

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