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Case Report

Pityriasis Lichenoides-Et Varioliformis Acuta (PLEVA): A Rare Case Report

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ABSTRACT

We present a case of a 70 year old male who presented with multiple, well defined, macular -papular erythematous patches present all over body except face and mucous membrane since 25 days. Histopathology showed interface dermatitis, epidermotropism, parakeratosis along with perivascular infiltrates chiefly comprising of lymphocytes and extravasated red blood cells indicating the diagnosis of Pityriasis lichenoides et Varioliformis acuta.

Key words: Pityriasis lichenoides; papules; PLEVA.

INTRODUCTION

Pityriasis lichenoides (PL) is a papulosquamous disorder of unknown etiology. Pityriasis lichenoides (PL) is of two clinical forms acute and chronic. [1] Pityriasis lichenoides et Varioliformis acuta (PLEVA) is the acute form of the disease. Histopathological examination helps in clinico-pathological correlation of this condition. Pityriasis lichenoides is more prevalent in the pediatric population, but it can affect patients in all age groups, races and geographic regions and there is a male predominance, adults are rarely involved. [2]

The etiology of Pityriasis lichenoides is unknown, but it has been postulated to be a response to an inflammatory reaction triggered by infectious agents, an immune- complex mediated hypersensitivity, or an inflammatory response secondary to T- cell dyscrasia. [3]

Therefore concept of pityriasis lichenoides as a T-cell lymphoproliferative disorder may help to explain its occasional association with other lymphoproliferative disorders such as cutaneous T-cell lymphoma, Hodgkin's disease, other lymphomas, HIV, medications (e.g. estrogen-progesterone, TNF-α inhibitors [infliximab, adalimumab], and radiocontrast dye. There is CD8 +T cell predominance in PLEVA and expression of HLA DR on the surrounding keratinocytes. [4] Now Pityriasis lichenoids is considered a lymphocytic vasculitis and it has been removed from the parapsoriasis group of disorders. [5]

CASE REPORT

A 70 year old male presented with widespread polymorphous, well defined, generalized macular-papular erythematous patches all over body (Figure 1) sparing the face (Figure 4) and mucous membrane since 25 days. The lesions started on bilateral legs, than bilateral arms followed by trunk (Figure 2 & 3). Lesions were not associated with itching, burning sensation or pain. His

personal and family history was non-contributory. He was not receiving any medications and there was no apparent episode of any infection before the onset of the eruption.

Histopathology of skin showed interface dermatitis, epidermotropism, parakeratosis along with perivascular infiltrates chiefly comprising of lymphocytes and extravasated red blood cells leading to the diagnosis of Pityriasis lichenoids et Varioliformis acuta.

He was treated with oral dapsone and after a month of treatment, there was regression of papular lesion.

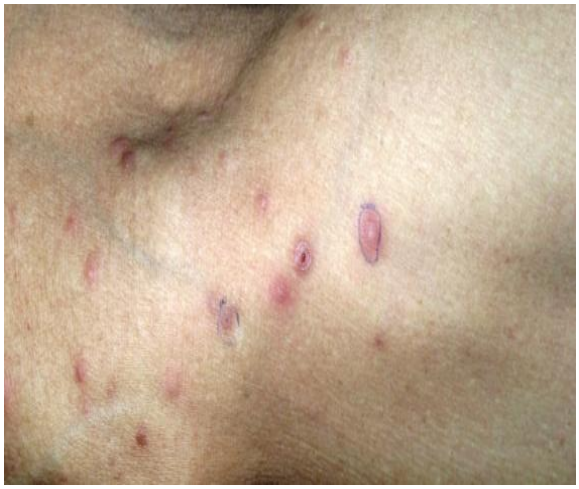

Figure 1: Erythematous macular patch on arm

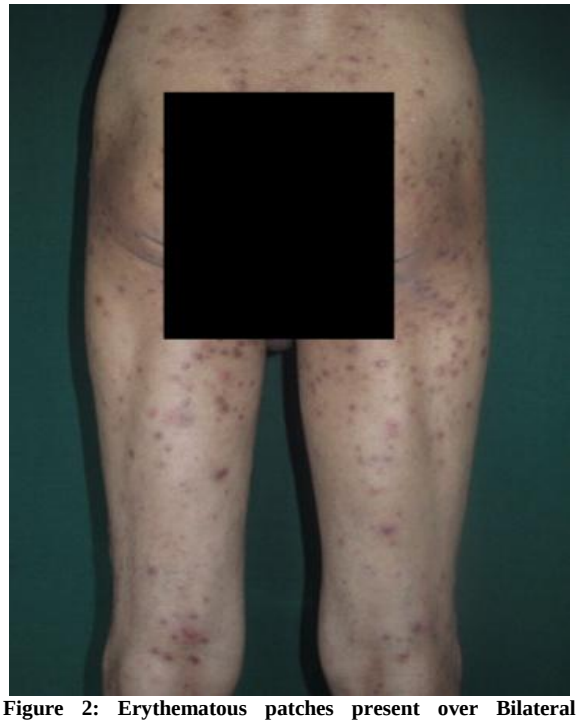

Figure 2: Erythematous patches present over Bilateral Extremities

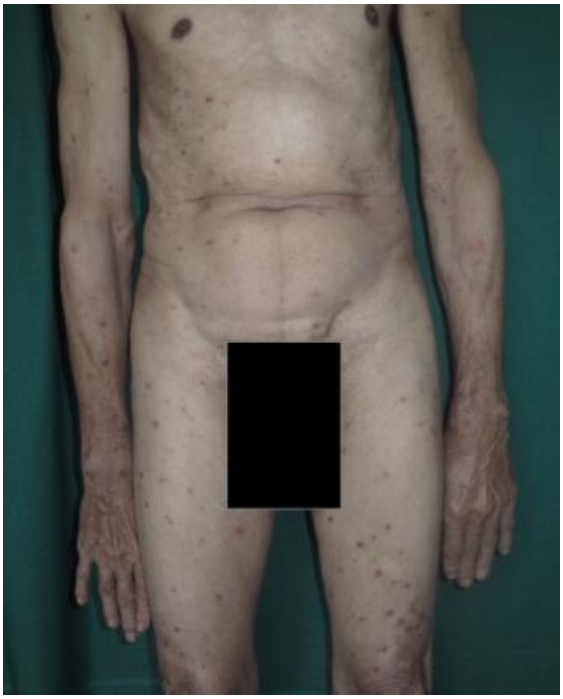

Figure 3: Trunk is involved

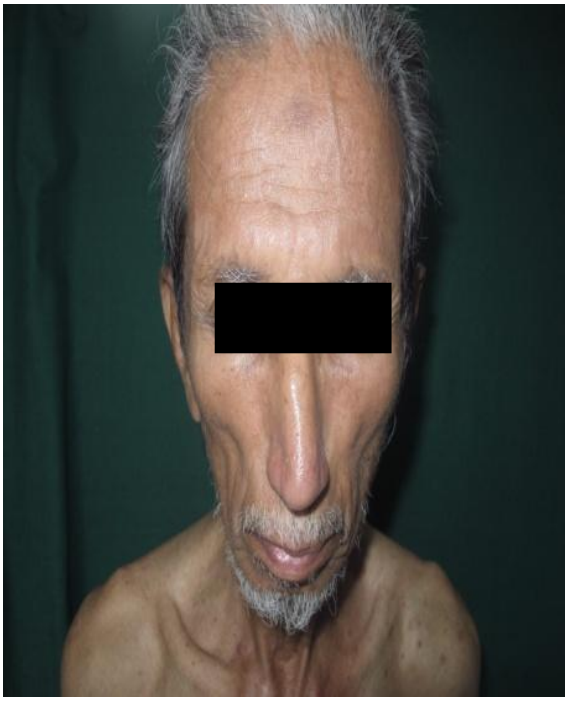

Figure 4: No face involvement

DISCUSSION

In 1916, Viktor Mucha first published a case of acute papular- squamous eruption named Parakeratosis Variegata (Unna) or Pityriasis Lichenoides Chronica. [5] In 1925, R. Haberman named it as Pityriasis lichenoides et Varioliformis acuta (PLEVA) [5] also known as Mucha - Haberman disease. Patients of PLEVA usually present with recurrent crops of

erythematous papules that develop into crusts, pustules or erosions before spontaneously regressing within a matter of weeks. [6] Patients can also develop scaly, red-brown papules that are consistent with Pityriasis lichenoides chronica (PLC) during the course of PLEVA, and occasionally, patients may completely transition from PLEVA to PLC. [7] These observations support the concept that PLEVA and PLC represent a spectrum of a single disease and can occur side by side.

In PLEVA there is a perivascular and dense, predominantly lymphocytic infiltrate in the papillary dermis that extends into the reticular dermis in a wedge shaped pattern [4] (Figure 5). The infiltrate may obscure the dermal- epidermal junction with vacuolar alteration of the basal layer, marked exocytosis of lymphocytes and erythrocytes leading to a variable degree of epidermal necrosis (Figure 6). The overlying cornified layer shows parakeratosis (Figure 7).

The differential diagnoses of PLEVA are Pityriasis rosea, vesicular insect bites and subcutaneous eczematous dermatitis. The distinguishing features are presence of deeper inflammatoryinfiltrate, extensive epidermal necrosis and the absence of intraepidermal spongiotic micro vesicles in PLEVA may from Pityriasisrosea and subcutaneous eczematous dermatitis.

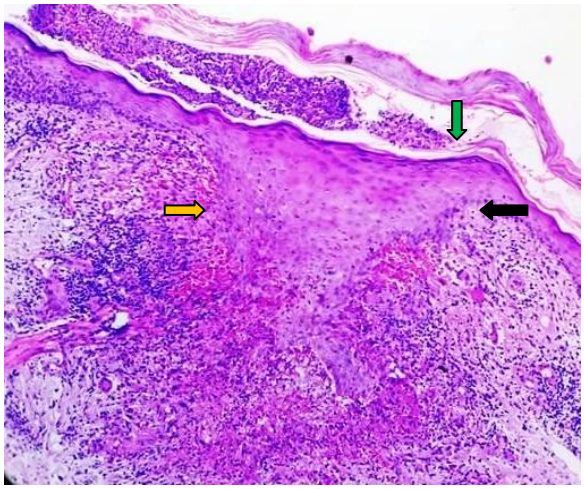

Figure 5: Histomorphological features show lymphocytic infiltrate in the papillary dermis (⬅) extends into the reticular dermis in a wedge-shaped pattern (⇨) and parakeratosis (⇩)

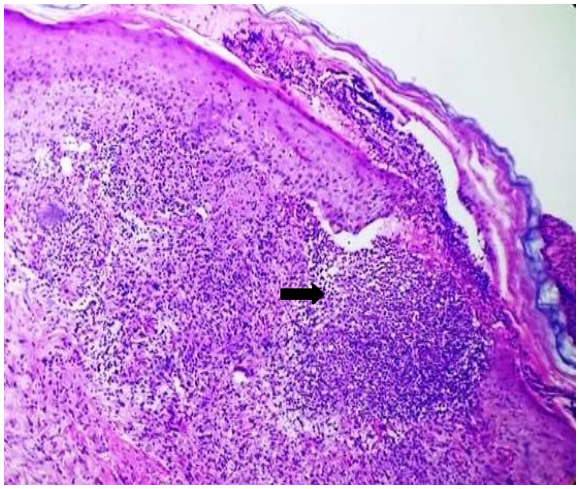

Figure 6: Histopathologyshowing Lymphocytic infiltrate (➡)

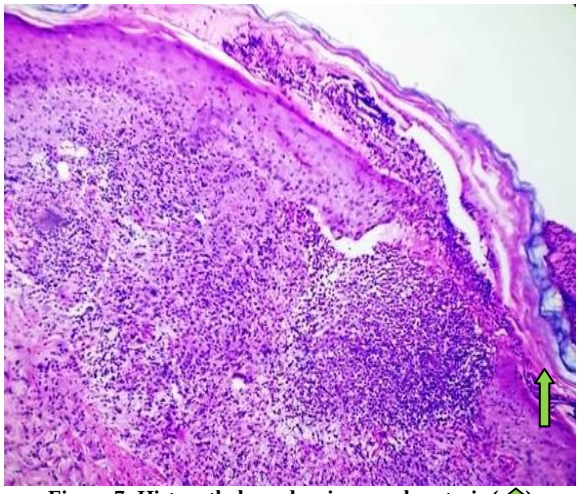

Figure 7: Histopathology showing parakeratosis (⇧)

CONCLUSION

In conclusion PLEVA is a rare entity in adults which may present difficulties in diagnosis and therapy. 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. Erythromycin and dapsone was effective in this patient. However one cannot rule out the possibility of spontaneous resolution.

REFERENCES

1. Bowers S, Warshaw EM. Pityriasis lichenoids and its subtypes. J Am Acad Dermatol 2006; 55:557-72.
2. Jean L Bolognia, Joseph L Jorizzo, Julie V Schaffer Chapter 9 other papulosquamous disorders, page 85.
3. Fernandes NF, RozdebaPj, Schwartz

RA, kichiczak G, Lambert WC. Pityriasis lichenoids et varioliformis acuta: A disease spectrum. Int J Dermatol 2010; 49:257-61.
4. Elder, David E.; Elenitsas, Rosalie; Johnson, Bernett L.; Murphy, George F. Lever's Histopathology of the Skin, 9th Edition. Philadelphia: Wolters Kluwer/Lippincott Williams & Williams, 2008.
5. Yadav N, Kar S, Gangane N, Madke B. Pityriasis lichenoides et varioliformis acuta (PLEVA). J Mahatma Gandhi Inst Med Sci 2015; 20:79-81.
6. Fitzpatcrik's Dermatology in General medicine, 7th edition, chapter 25, parapsoriasis and Pityriasis lichenoids, page 236.
7. Pityriasis lichenoides et varioliformis acuta in 35 years old male -A rare case report, Medical Science, 2014, 9(34), 47-50.

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