THE STUDY OF CLINICAL VARIATIONS AND HISTOPATHOLOGICAL FINDINGS IN POLYMORPHOUS LIGHT ERUPTION

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ABSTRACT

BACKGROUND

Epstein. S in 1942 described PMLE under the name of Prurigo aestivalis. He first hypothesised that PMLE represents a form of delayed-type hypersensitivity response to an endogenous, cutaneous UV-induced antigen, because of the hours or days delay between sun exposure and manifestation of symptoms, and the histological appearance of lesional skin. Firm evidence; however, has been lacking and the responsible allergen has not been identified. PMLE a specific entity encompassing six clinical manifestations: Small erythematous papulovesicles, eczematous lesions, large papules, oedematous plaques, prurigo nodules and erythema multiforme-like lesions. The histological features of PMLE are characteristic, but not pathognomonic and vary with the different clinical presentation. A sincere effort has been put in this study to understand the clinical and histopathological features of polymorphic light lesions. The study is intended to help the practising physicians and dermatologists to diagnose the pathology on time and intervene before it develops into complications.

METHODS

Material for the present study consisted of 100 cases of clinically diagnosed untreated cases of polymorphous light eruption, who were attending the skin and STD and Leprosy Department, Kakatiya Medical College, Warangal, during January 2014 to Dec 2014 were selected for this study. The patients were selected randomly irrespective of age, sex, socioeconomic status.

RESULTS

Histopathology showed perivascular lymphocytic infiltration in the dermis in majority of our cases (93.33%) and clinico-histopathological correlation was observed in most cases; hence apart from clinical examination, histopathological examination plays an important role in diagnosing PMLE.

CONCLUSION

56 percent of the patients complained pruritus followed by burning. The study was similar to other studies in comparison.

KEYWORDS

Sunlight, UV rays, Clinical, Histopathological, Polymorphous Light Eruption.

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INTRODUCTION: The study of effect of non-ionising radiation on biological system is called as photobiology, human photobiology is concerned with its effect on two organ systems - skin and eye[1]. Epstein. S in 1942 described PMLE under the name of Prurigo aestivalis. He first hypothesised that PMLE represents a form of delayed-type hypersensitivity response to an endogenous, cutaneous UV-induced antigen, because of the hours or days delay between sun exposure and manifestation of symptoms, and the histological appearance of lesional skin. Firm evidence, however, has been lacking and the responsible allergen has not been identified.

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This abnormal immune response to sunlight influenced by:

Photobiologic: Sunlight is clearly primary factor. The eruption of PMLE typically occurs in spring or rarely in winter following ultraviolet radiation exposure reflected from snow[2]. Most PLE patients exhibit sensitivity to sunlight through window glass while travelling in cars, trains or planes[2]. This observation, along with the lack of protection from UVB-absorbing sunscreens in the majority of PLE cases, implicate the role of UVA in triggering the eruption[3]. Despite numerous efforts to define the most effective wavelengths in PLE induction, the action spectrum of the disease remains elusive. Studies of the minimal erythemal dose (MED), action spectrum, and energy doses required to reproduce the lesions have led to conflicting results because of variations in individual sensitivity, seasonal variations, geographical differences, and differences in artificial light sources, light testing protocols and result interpretations. Most patients with PLE have a normal MED value.[4,5] Although a significant number may exhibit a lowered MED value to UVA or UVB.[6]

Genetic Aspects: A family history of PLE has been reported in up to 50% of subjects in some series. The expression of the disease in these individuals is largely determined by a polygenic component with an additional environmental component, possibly exposure to sunlight. The data also supports the hypothesis that PLE and actinic prurigo share a common genetic background, with the latter possibly representing an HLA-restricted subset of PLE. More recently, a reverse link to a glutathione-s-transferase (GSTP1) allele was reported as the first genetic association in PMLE, supporting a potential role for reactive oxygen species in the pathogenesis.[7-8]

Immunology: It was long thought that PLE was delayed type hypersensitivity reaction against UVR induced cutaneous antigen. Sequential skin biopsies following low dose solar irradiation experimentally demonstrates Perivascular infiltrate with predominance of CD_4+ T cells in lesions up to 72 hours and after 72 hours, later lesions an infiltrate dominated by CD_8+ cells. Increased number of dermal and epidermal Langerhans' cells and dermal macrophages were also present.[9]

Biochemical: Abnormalities of arachidonic acid metabolism and prostaglandins have been also reported in PLE. This observation may indicate that PLE comprises two different disease states, the most severe one associated with abnormal arachidonic acid metabolism in response to UVR. Dietary supplements of fish oil rich in ω3 polyunsaturated fatty acids, that compete with arachidonic acid and lead to the generation of less active prostanoids, have been shown to reduce the basal and UVB-generated prostaglandin levels in skin and increase the threshold for UVA-induced provocation of PLE.[10]

Environmental: The PLE eruption begins typically in the spring or early summer and tends to improve as the summer progresses. It rarely occurs in the winter unless the skin is exposed to UVR reflected by the snow. The period of sun exposure required to induce the reaction varies from subject to subject, but it usually lasts from 30 min. to several hours, and in cases of long sun abstinence, several days. The course of the eruption is one of the characteristic features of PLE that distinguishes it from the other photodermatoses. Epstein[11] attempted to define PMLE a specific entity encompassing six clinical manifestations: Small erythematous papulovesicles, eczematous lesions, large papules, oedematous plaques, prurigo nodules and erythema multiforme-like lesions. The skin lesions usually appear after a delay period of minutes to hours, but not less than 30 min., and in the absence of further sun exposure, resolve after 7–10 days, and occasionally longer, without scarring or other residual.[11] However, in an individual patient, lesions are monomorphic and tends always to affect the same skin sites, although its distribution may gradually spread or recede over some exposed sites, often those normally covered in winter such as the upper chest and arms. Associated systemic symptoms are rare, but chills, headache, fever, nausea, and a variety of other sensations are possible.

Discomfort in the form of pruritus or burning may be experienced by affected subjects prior to the onset of the eruption. The characteristic sequence of events is pruritus followed by patchy erythema and finally the onset of distinct lesions. Prominent and widespread skin lesions can be provoked after long periods of seasonal lack of sunshine.

During the course of the activity of the disease, some patients will show progressive worsening of their symptoms, while others will exhibit gradual improvement due to 'skin hardening'. This hardening phenomenon occurs after repetitive exposures to UVR during the summer months and results in complete tolerance even of intense sunshine towards the end of the summer period. The frequent sparing of the facial skin that is continuously exposed to some UVR throughout the year, the gradual diminution of the rash during the summer in some patients, and the preventive effect of phototherapy are all possibly related to the development of immune tolerance through skin hardening. The main mechanism by which hardening is achieved involves the suppression of the immune mechanisms of PLE, although the increased melanisation and thickening of the stratum corneum induced by UVR may also play a part. The histological features of PMLE are characteristic but not pathognomonic and vary with the different clinical presentations.

A sincere effort has been put in this study to understand the clinical and histopathological features of polymorphic light lesions. The study is intended to help the practising physicians and Dermatologists to diagnose the pathology on time and intervene before it develops into complications.

AIMS AND OBJECTIVES:

- To study the clinical variation of disease.
- To study the histopathological findings in relation to the clinical diagnosis made.

MATERIALS AND METHODS: The study consisted of 100 cases of clinically diagnosed untreated cases of polymorphous light eruption, who were attending the skin and STD and Leprosy Department, Kakatiya Medical College, Warangal, during January 2014 to December 2014 were selected for this study. The patients were selected at random irrespective of age, sex, socioeconomic status. A detailed history was taken.

A detailed general examination was carried out in all cases with particular reference to find out the distribution of skin lesions, type of skin lesions, any secondary changes. Local examination was carried out. All systems were carefully examined to find out any associated abnormalities in other systems.

Skin biopsy of a classical skin lesion in 30 patients were taken. After thorough cleansing of the selected part with rectified spirit, the parts were infiltrated with 2% Xylocaine and bit of skin involving the whole thickness removed by using punch biopsy method from the edge of the involved skin and the specimens were fixed in aqueous solution of formalin 10.5% and submitted for histopathology studies at Pathology Department, Kakatiya Medical College, Warangal.

Inclusion Criteria:

1. Rash occurs following sun exposure mainly on photoexposed skin.
2. Rash is recurrent, pruritic & heals without scarring.
3. Lesions are papules and/or vesicles and/or plaques.

Exclusion Criteria:

1. Known coexistent photosensitivity disorder.
2. Cases which had been treated earlier.
3. Histology (if performed) incompatible with PLE.

RESULTS:

Symptoms	Number of patients	Percentage
Asymptomatic	18	18%
Pruritus	56	56%
Burning	14	14%
Both	12	12%
Total	100	100%
Table 1: Symptoms		

Itching was the commonest symptom in most of the patients (56%). It was alone present in 56 cases. Burning alone was noted in 14 patients. Both itching and burning was noted in 12 cases. 18 patients did not complain of any symptom.

Clinical Types	Male	Female	Total	Percentage
Papular	17	41	58	58%
Plaque	11	24	35	35%
Eczematous	3	4	7	7%
Total	**31**	**69**	**100**	**100%**
Table 2: Clinical types in Study Group				

Papular variant of PLE was the commonest clinical types in our study (58%) followed by plaque (35%) and eczematous (7%) types.

Site	Male	Female	Total	Percentage
Face	8	15	23	23%
Back & Sides of Neck	10	25	35	35%
Arm & Forearm	13	29	42	42%
Total	**31**	**69**	**100**	**100%**
Table 3: Distribution of Lesions				

Lesions are commonest on arm and forearm which are involved in 42 cases. Face was involved in 23 cases, back and sides of neck in 35 cases.

Fig. 1: Plaque variant of PMLE-I (Right)
Fig. 2: Plaque variant of PMLE - 2

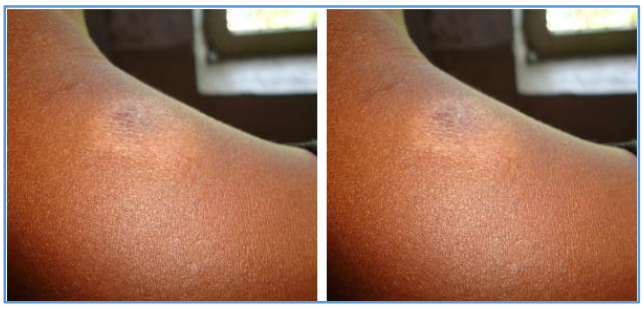

Fig. 3: Plaque variant of PMLE-II (Right)
Fig. 4: Plaque variant of PMLE-III (Left)

Fig. 5: Papular variant of PMLE – I
Fig. 6: Papular variant of PMLE – II

Fig. 7: Eczematous variant of PMLE – I
Fig. 8: Eczematous variant of PMLE - II

Epidermal/Dermal Changes	Total (n)	Percentage
Epidermis		
Hyperkeratosis	25	83.33%
Parakeratosis	10	33.33%
Spongiosis	14	46.66%
Acanthosis	18	60%
Hydropic degeneration of basal cells	12	40%
Increase in melanocytes	16	53.33%
Thinning of epidermis	4	13.33%
Subcorneal vesicles with exocytosis	2	6.67%
Dermis		
Perivascular lymphocytic Infiltration	28	93.33%
Oedema	15	50%
Table 4: Histopathological Findings		

Epidermis	Papular	Plaque	Eczematous
Hyperkeratosis	17	8	0
Parakeratosis	6	4	0
Spongiosis	7	3	4
Acanthosis	10	7	1
Hydropic degeneration of basal cells	6	3	3
Increase in melanocytes	11	4	1
Thinning of epidermis	3	1	0
Subcorneal vesicles with exocytosis	0	0	2
Dermis			
Perivascular lymphocytic infiltration	15	9	4
Oedema	5	7	3

Table 5: Histopathological Changes According to Clinical Types

Epidermal Changes: Hyperkeratosis was observed in the biopsy specimens of 25 cases, whereas parakeratosis was observed in 10 cases. Parakeratosis was only focal in all 10 cases. Spongiosis was observed in 14 cases only and in all biopsy specimen spongiosis was focal. Acanthosis was observed in 18 biopsy specimens and it was focal in 3 biopsy specimens. The hydropic degeneration of the basal cell layer was observed in 12 biopsy specimens and in all these 12 biopsy specimens, it was only focal degeneration. The melanocytes were increased in 16 biopsy specimens studied. Thinning of the epidermis was observed in 4 biopsy specimens. There was subcorneal vesicles with exocytosis in 2 biopsy specimen studied.

Dermal Changes: Perivascular lymphocytic infiltration was observed in all of 28 specimens studied. The infiltration was mild in 10 cases, moderate in 13 cases and was dense in 5 case Dermal Oedema was noted in 15 cases.

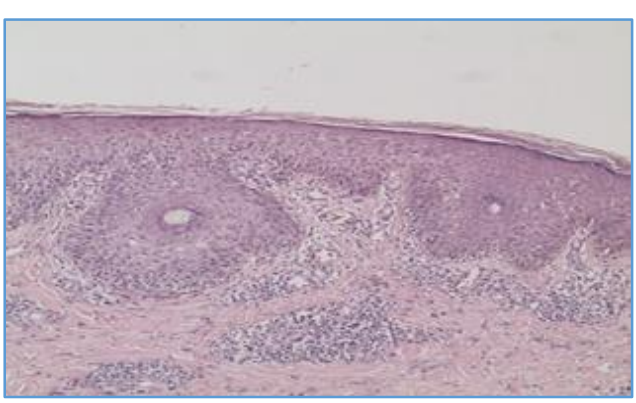

Fig. 9: Histopathology of PMLE - I

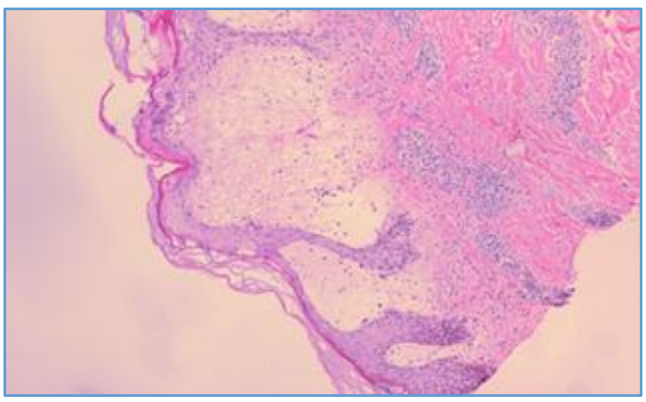

Fig. 10: Histopathology of PMLE - II

DISCUSSION: Polymorphic light eruption is a common form of primary photosensitivity that mainly occurs in young adult women in temperate climates during spring and summer.

PLE generally affects adult females aged 20–40, although it sometimes affects children and males. It is particularly common in places where sun exposure is uncommon, such as Northern Europe, where it is said to affect 10–20% of women holidaying in the Mediterranean area. It is less common in Australasia. It has also been reported to be relatively common at higher altitudes compared to sea level.

PLE can occur in all races and skin photo types and may be more prevalent in skin of colour than in white skin. There is a genetic tendency to PLE, and it is sometimes associated with or confused with photosensitivity due to lupus erythematosus.

Some patients experience PLE during phototherapy which is used to treat skin conditions such as psoriasis and dermatitis.

PLE is caused by a delayed hypersensitivity reaction to a compound in the skin that is altered by exposure to ultraviolet radiation (UVR). UVR leads to impaired T cell function and altered production of cytokines in affected individuals. There is a reduction in the normal UV-induced immune suppression in the skin. This has been suggested to be either due to oestrogen or deficiency of vitamin D.

PLE may be a rare occurrence in the individual concerned or may occur every time the skin is exposed to sunlight. The commonest variety of PLE presents as crops of 2–5 mm pink or red papules. The arms, the back of the hands, the V of the neck, the chest and lower legs/feet may be affected, but the face is usually spared. A few people complain of ocular and/or lip lesions. Juvenile Spring Eruption is a variant of PLE that is confined to the ears of children (usually boys). It persists for several days, and often longer if the affected skin is exposed to more sunlight. It resolves without scarring.

PLE usually causes a burning sensation or itch. A few individuals also report fever and malaise following sun exposure.

Skin biopsy is necessary to make a diagnosis. PLE has characteristic histopathological features, with upper dermal oedema and a dense perivascular lymphocytic infiltrate.

Eczematous changes may be present. Direct immune fluorescence is negative, unless the patient has cutaneous lupus erythematosus.

It is usual to have a blood count and a check for circulating antinuclear antibodies (ANA) and extractable nuclear antigens (ENA) in case of photosensitive cutaneous lupus erythematosus.

Photo testing is not usually carried out, but provocation tests of exposure to UVA daily for 3 days to a small area of skin can confirm the diagnosis.

Many people can avoid developing a rash by using effective sun protection during the middle hours of the day during summer. Cover affected areas with densely woven sun protective clothing, stay in the shade.

Many patients find antioxidant nutritional supplements somewhat helpful. It has been noted that PLE appears to be less frequent and severe in women after the menopause. Topical corticosteroid creams to relieve symptoms, short course of oral steroids and phototherapy are the treatment of choice.

Pruritus was one of the symptoms observed in 56 patients out of 100 patients. It was of mild to moderate degree in 45 patients and was of severe degree in 11 patients. Burning was present in 14 patients. Both itching and burning were present in 12 patients. According to JH Epstein, [12] rash (1980) usually starts with a burning, stinging sensation followed by erythema and oedema. In a study, HE Boonstra[13] (2000) found that most patients suffered from itch, but also burning and painful sensations were complained. As mentioned in the above studies, pruritus is the predominant finding which is also the major symptom in our study.

In our series, papular type occurred in 58 patients, plaque type in 35 patients and eczematous type in 7 patients. AJ Stratigos, C Antoniou (2002)[14] described clinical types as Papular, Papulovesicular, Plaque-like, Urticarial, Vesiculobullous, Haemorrhagic, Eczematous, Erythema multiforme-like, Insect bite-like, Prurigo-like. Our findings are consistent with the other studies.

In our study, we noted face involvement in 23%, neck involvement in 35%, arm & forearm in 42%, in both sexes. According to Percy Lehmann (2006),[15] most common sites noted were V of the neck, lateral aspects of the upper arms, dorsal aspects of hands, thighs and lateral aspects of the face. Our study is consistent with few of the above studies and differences in the site are due to distribution in clothing. 30 biopsy specimens were studied in this series. Hyperkeratosis was observed in 25 biopsy specimens, parakeratosis was observed in 10 out of 30 biopsy specimens and acanthosis in 18 specimens, spongiosis in 14 cases. There was a focal basal cell layer hydropic degeneration in 12 biopsy specimens, melanocytes were increased in 16 specimens. There was thinning of the epidermis in 4 specimens. Perivascular lymphocytic infiltration at the mid and upper dermis was observed in 28 biopsy specimens. Dermal oedema was noted in 15 cases. Lamb et al.[16] described 28 patients with PMLE, microscopic sections from these biopsies showed parakeratosis (4/15 cases), atrophy (12/15 cases), follicular plugging (number of cases not specified), either intercellular or intracellular oedema (10/15 cases), vacuolar alteration (5/15 cases), papillary dermal oedema (14/15 cases), dilated blood vessels and a dense perivascular infiltrate. Wright et al[17]. described histopathology from 14 patients who had PMLE and reported that there was parakeratosis, hyperkeratosis, focal acanthosis, follicular plugging, slight vacuolar alteration (occasionally), subepidermal oedema, vascular dilatation and a perivascular infiltrate of lymphocytes, occasional neutrophils. Histopathological findings noted in our study were consistent with the above-mentioned studies.

CONCLUSION: Majority of patients complained of pruritus (56%) followed by burning (14%) and 18% of them are asymptomatic. Papular form constituted the major clinical type noted in 58% followed by plaque variant (35%). Arm and forearm were more commonly involved in both sexes (42%). Histopathology showed perivascular lymphocytic infiltration in the dermis in majority of our cases (93.33%) and clinicohistopathological correlation was observed in most cases; hence, apart from clinical examination, histopathological examination plays an important role in diagnosing PMLE.

REFERENCES

1. Deleo VA, Masu MJ. Photosensitivity Moschella. 3rd edn. Vol. 1 Biological Photographic Association 1978:p.508.
2. Honigsmann H. Polymorphous light eruption. In: Lim HW, Soter NA, editors. Clinical photomedicine. Marcel Dekker Inc. New York 1993:167–180.
3. Diffey BL, Farr PM. An evaluation of sunscreens in patients with broad spectrum photosensitivity. Br J Dermatol 1985;112(1):83–86.
4. Diffey BI, Farr PM. The erythemal response to ultraviolet radiation in subjects with polymorphic light eruption. Br J Dermatol 1986;114(1):103–108.
5. Jansen CT. Erythemal and pigmentary photo test reactions in polymorphous light eruption. Acta Derm Venereol (Stockh) 1979;59:499-503.
6. Rottier PB. Testing of the skin for reactions to ultraviolet irradiation. Dermatologica 1963;127:260-266.
7. Norris PG, Hawle JLM. The idiopathic photodermatoses; polymorphic light eruption, actinic prutig and hydroavacciniforme. In: Hawk JLM, ed. Photodermatology. London: Arnold 1999:178-190.
8. Hawk JLM. Polymorphic light eruption. Rooks Textbook of Dermatology. 7th edn. Vol. 2. McGraw Hill Professional 2007:10-24.
9. Norris PG, Morris J, McGibbon DM, et al. Polymorphic light eruption: an Immunopathological study of evolving lesions. Br J Dermatol 1989;120(2):173-183.

10. Rhodes L, Durham BH, Fraser WD, et al. Dietary fish oil reduces basal and ultraviolet B-generated PGE2 levels in skin and increases the threshold to provocation of polymorphic light eruption. J Invest Dermatol 1995;105(4):532-535.
11. Epstein JH. Polymorphous light eruption. Ann Allergy 1966;24(8):397-405.
12. Epstein JH. Polymorphous light eruption. J Am Acad Derm 1980;3(9):329-343.
13. Boonstra HE, van Weelden H, Toonstra J, et al. Polymorphous light eruption: a clinical, photobiologic and follow-up study of 110 patients. J Am Acad Dermatol 2000;42(2 pt 1):199-207.
14. Stratigos AJ, Antoniou C, Katsambas AD. Polymorphous light eruption. JEADV 2002:16(3):193-206.
15. Lehmann P. Diagnostic approach to photodermatoses. JDDG 2006:4(11):965-975.
16. Lamb JH, Jones PE, Maxwel TB. Solar Dermatitis. Arch Derm 1957;75(2):171-180.
17. Wright ET, Winer LH. Histopathology of allergic solar dermatosis. J Inv Derm 1960;34:103-106.