Polymorphous Light Eruption- An Indian Scenario

Abstract

Polymorphous light eruption (PMLE) is the most common, idiopathic, acquired photodermatosis, characterized by abnormal, recurrent, and delayed reaction to sunlight. Polymorphous light eruption is common worldwide but the morphology, distribution, and pigmentary changes are unique in Indian skin which is discussed in this review. The prevalence of PMLE is around 10-20% in the general population. It commonly occurs in females between 20and 30 years of age. It is the most common photodermatosis in school-going children. Visible light sensitivity is an important phenomenon in PMLE. It typically presents as recurrent and chronic lesions over photoexposed sites. Initially, patchy erythema occurs with pruritus. Most of the Indians belong to type IV to type VI skin and pigmentary changes are commonly seen. The unique feature of PMLE in Indian skin is the pigmentary change which varies from hypopigmented to hyperpigmented lesions. These pigmentary changes may occur alone or in combination with erythematous or skin-colored lesions. The pigmentary lesions are seen in more than 50% of lesions. The histopathology of PMLE is characterized by the presence of hyperkeratosis, spongiosis with or without the presence of liquefactive degeneration in the epidermis. Dermal changes in the upper and mid dermis include the presence of dense perivascular lymphocytic infiltrate. The management of PMLE includes both preventive measures and medical management. Topical sunscreens, topical steroids, hydroxychloroquine and antioxidants play a very important role.

Keywords: Hydroxychloroquine, Indian, pigmentary changes, polymorphous light eruption

Polymorphous light eruption- an Indian Scenario

Polymorphous light eruption (PMLE) is the most common, idiopathic, acquired photodermatosis, characterized by abnormal, recurrent, and delayed reaction to sunlight. It is an immunologically mediated disease that occurs due to delayed hypersensitivity reactions.[1-4] It is commonly known as "sunrash" and referred to as "sunallergy" by the patients even though there is no real allergy associated with its pathogenesis.^[5] It was first described by Robert Willanin 1817 as"eczema solare." The term "polymorphous light eruption" was coined by Carl Raschin 1990 and was again described as a common term for prurigo aestivalis and eczema solare by Haxthausenin1919.^[6] It is also referred to as dermatographia photogenica, erythema perstans solare, and prurigo aestivalis.^[7] Polymorphous light eruption is common worldwide but the morphology, distribution, and pigmentary changes are unique in Indian skin which is discussed

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in this review. Further, the treatment modalities and the role of sunscreens in Indian context is also discussed.

Epidemiology

The prevalence of PMLE is around10-20% in the general population^[8] with females between 20 and 30 years of age^[6] and children school-going affected more commonly. PMLE frequently occurs in temperate climates due to a greater proportion of UVA to UVB in these regions. Though the disease is said to be more common in temperate regions, the prevalence of PMLE in India is similar to that reported in the world.^[9] The proportion of cases varies between 2% and13.5% across different areas in India.[10-17] Most of these studies are hospital-based and may not represent the community prevalence. This probably is an underestimate of the real prevalence.

In Indian studies, a female preponderance was noted. The disease is seen in people who indulge in outdoor activities such as farmers and laborers. In certain studies,

How to cite this article: Karthikeyan K, Aishwarya M. Polymorphous light eruption- An Indian scenario. Indian Dermatol Online 2021;12:211-9.

Received: 20-Aug-2020. Revised: 20-Sep-2020 Accepted: 20-Oct-2020. Published: 02-Mar-2021.

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high incidence was noted in housewives probably because of household activities.^[10,11,18]

Seasonal Variation

The wide climatic and geographic variations in India lead to a range of seasonal variation. The PMLE peaks in the month of March and continues into early summer. These are the days when sunshine is longer. Some cases also occur during later winter from January onwards in Northern India probably because of the habit of sunbathing. The second peak was noted in September.^[10,11,14,17]

Genetic Predisposition

Polymorphous light eruption occurs in family members in 12–46% of PMLE patients.^[8] "Familial clustering" in PMLE suggests a genetic etiology.^[10] In India family clustering was not observed.

Pathogenesis of PMLE

PMLE occurs due to the interplay of genetic and environmental factors.^[19-22] The possible mechanisms involved in pathogenesis are given in the form of a chart [Figure 1] which incorporates various factors involved.^[23-27]

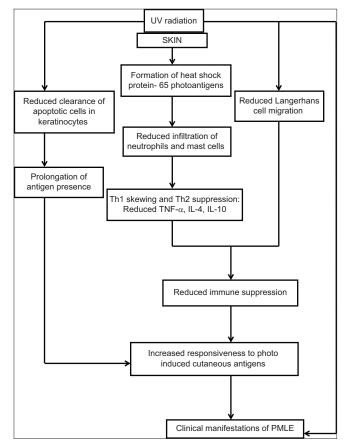


Figure 1: Pathogenesis of PMLE

Clinical Features

Photosensitivity

Visible light sensitivity is an important phenomenon in PMLE.^[27] It typically presents over photo exposed sites within a few minutes of exposure to sunlight. Photosensitivity is the most common symptom that occurs in Indian patients but some experienced a burning sensation on exposure to the sun.

Morphological lesions

Initially, patchy erythema occurs with pruritus. The primary sites involved are upper chest, "V" area of the neck, upper arms, forearms, back of hands, thighs, and the sides of the face.^[28-34] Improvement of lesions is noticed during the latter half of the summer season, which is called "hardening."^[32,34] Due to repeated daily sun exposure, the face and hands undergo hardening.^[35]

The action spectrum involves UVA, UVB, and visible light.^[25,36] There is also an interesting observation of PMLE preferentially involving the sites of hypopigmented scars.^[37]

PMLE is typically characterized by transient, intermittent, and a delayed response at 30 minutes to several hours after UV light exposure resulting in an abnormal cutaneous response.^[10,15,16] The eruption usually takes up to two weeks to resolve in the absence of further ultraviolet radiation.^[17] A typical lesion of PMLE is a hyperpigmented plaque with a hypopigmented rim. Morphologically many variants have been described, hence the name "polymorphous."^[28] But one morphology usually predominates in an individual i.e., monomorphous.^[38] The lesions usually resolve in about 2 weeks in the absence of UV radiation.

The clinical features of PMLE usually follow a characteristic sequence following sun exposure. It starts with itching followed by the appearance of a patchy erythema. Following this, distinct lesions appear which are sparsely distributed initially, which then coalesce to form densely aggregated lesions. Lichenification, scaling, and scarring usually occur as secondary lesions as a result of scratching and rubbing.

The morphological types of lesions noted in the Indian skin are macule, papule, plaque, vesicular, and plaques with lichenification; as isolated lesions or in combinations. In India, the sites of lesions are influenced by the clothing. In women, the forearm and back are exposed and they are commonly involved.[Figure 2] The external aspect of the arms and forearms were involved in most of the cases possibly because these parts are placed horizontally while sitting or traveling and receive the maximum exposure.^[18] [Figure 3] On the other hand, the position of the face is vertical while walking or working and only rays fall on the cheek and nose which are affected. [Figures 4-7]. Neck may be exposed to the sun if the person is bending



Figure 2: Well-defined plaque on the back

Table 1: Morphological types of PMLE^[1,5,38-45]

Papular Papulovesicular Plaque type Vesiculobullous Urticarial Hemorrhagic or purpuric Eczematous Confluent edematous swelling of face Insect bite like (strophula) Prurigo like Erythema multiforme like Pinpoint papular variant Lichen nitidus like Lichen planus like Micropapular variant PLE sine eruption (erythema or pruritus alone) Erythema solareperstans Hydroavacciniforme like Solar urticarial like PMLE occurring over hypopigmented scars

forward. [Figure 8]. Covered areas were not affected irrespective of the type of clothing which suggests that it is probably preventable by all types of clothing.



Figure 3: Well-defined plaque on the face

Various morphological types of PMLE have been reported. They are summarized in Table 1.

The unique variants described include the pinpoint papular variant or micropapular variant of PMLE, which presents as multiple, monomorphic pinpoint, closely aggregated skin-colored to hypopigmented itchy lichenoid papules (1–2 mm) on sun-exposed areas[Figure 6].^[39] Erythema multiforme like PMLE is a less common type of PMLE. It typically presents as target-like lesions in photo distributed areas. This type has to be differentiated from photosensitive erythema multiforme based on histopathology and phototesting results.^[40,41]

Singh *et al.* described a distinctive pseudovesicular, monomorphic micropapular eruption predominantly involving the nose and adjoining cheeks that affects young to middle-aged people with no gender predilection. It may be photoaggravated in some cases and runs a chronic course. They proposed the term lichenoid pseudovesicular papular eruption of the nose for this condition. This may be variant of PMLE seen in India.^[46]

Pigmentary Changes and PMLE

In fairer skin types, the lesions are mostly erythematous papules or plaques and pigmentary changes are unknown.^[38]

Table 2: Difference between LE and PMLE ^[8,47]				
	LE	PMLE		
Typeoflesion	Polymorphic	Monomorphic		
Morphology	Malarrash, vasculiticlesion, discoidlesion,Itchy, groupedsmall/largeerytlperiunguallesionskin-coloredpapules, plaques/			
Sunlightexposure	Aggravates; canoccurinnonexposedareasalso	Induces		
Photosensitivity	Burning, erythema, edema	Itching, precipitationoflesion		
Orallesion	Erosion/erythematousplaquein >25%	Not seen		
Constitutionalsymptoms	Fever, arthralgia-common	Infrequent		
Systemicinvolvement	Frequent	Notseen		
Investigation	ANApositivity	Phototesting		



Figure 4: Facial plaque with a peripheral rim of hypopigmentation

Most of the Indians belong to type IV to type VI skin and pigmentary changes are commonly seen which varies from hypopigmented to hyperpigmented lesions. [Figures 7 and 8] These pigmentary changes may occur alone or in combination with erythematous or skin-coloured lesions. The pigmentary lesions are seen in more than 50% of lesions.^[18]



Figure 5: Multiple plaques on the face with a peripheral rim of hypopigmentation

The pigmentary chnges may persist after the lesions subside and have to be differentiated from other causes of hypopigmentation.

Differential Diagnosis

The important differential diagnosis is Lupus erythematosus. There are also a few reports that show that PMLE patients are associated with high titres of ANA and severe photosensitivity progressing into lupus erythematosus.^[47] The difference between PMLE and LE are summarized in Table 2.

The differential diagnosess for hypopigmented macular PMLE lesions are pityriasis alba, pityriasis versicolor, indeterminate leprosy, previtiligo, post-inflammatory

Hypopigmentation, and nevus anemicus.^[48,49] In India, dermatophytosis is also an important mimic of PMLE.^[50]

The infiltrated plaques have to be differentiated from sarcoidosis, Borderline leprosy, Jesse's lymphocytic infiltrate, lupus vulgaris, granuloma annulare, and granuloma multiforme. Another important photodermatosis which has to be differentiated from PMLE is actinic prurigo.

PMLE in Children

PMLE is the most common photodermatosis in childhood.^[35] It occurs in children during the summer. They are commonly seen over the face, the "V" area of the chest, the back of the neck, and the dorsolateral aspects of the forearms. The face is the common site of occurrence in children. [Figures 6 and 7] The morphological patterns observed include grouped papules, plaques, vesicles, and eczematous plaques.

Histopathology of PMLE

The histopathology of PMLE is characterized by the presence of hyperkeratosis, spongiosis with or without the presence of liquefactive degeneration in the epidermis. Dermal changes in the upper and mid dermis include the presence of dense perivascular lymphocytic infiltrate. The dermal infiltrate is composed primarily of T lymphocytes. Sunburn cells are notably sparse or absent.^[25] The histopathology varies



Figure 6: Hypopigmented pinpoint papules on forearm

depending upon the morphological type. Histopathological differential diagnoses for polymorphous light eruption includes reticular erythematous mucinosis, Jessener's lymphocytic infiltrate, lupus erythematosus, and actinic reticuloid. The histopathology of PMLE can be graded as follows [Table 3].^[17]

Phototesting

In an Indian study, phototesting of patients with PMLE showed that UVB rays were the most relevant wavelength. This increased sensitivity could be due to the geographical conditions, heat, and humidity in the subtropical climate. Photo-patch testing is not helpful indiagnosing PMLE.^[51]

Management of PMLE

The management of PMLE includes both preventive measures and medical management.

Physical protection plays a very important role. The use of an umbrella can be very useful. The patients should wear a



Figure 7: Hypopigmented lesions on face

Table 3: Histopathology grading of PMLE ^[42]				
Grading	Epidermis	Basal cell degeneration	Dermis	
Diagnostic	Hyperkeratosis/ atrophy/spongiosis	Liquefactive degeneration present	Dense perivascular lymphocytic infiltrate in upper and mid dermis	
Possible	Atrophy/spongiosis	Not present	Sparse perivascular lymphocytic infiltrate	
Probable	No changes	Not present	Minimal perivascular lymphocytic infiltrate.	

Table 4: Instructions to patients

Sun protection should be advised a preventive measure during summer months by the following methods Avoid sun exposure between 11.00 am and 3.00 pm. Regular application of broad-spectrum sunscreens of new generation with high SPF Apply sunscreens half an hour before sun exposure. Reapply every 2 h Follow "Teaspoon rule": 3 ml each for face, arms and 6 ml each for trunk and legs Cover all sun-exposed sites including temples, ears, lateral, and posterior neck.

Avoid wearing tight and wet clothes

Table 5: Treatment ladder of PMLE^[6,8,24,45]

First-line

Sun avoidance

Broad-spectrum sunscreens with patient education on application technique

Topical corticosteroids + phototherapy or photochemotherapy Second line

Short course corticosteroids for 4-5 days

Mild cases: self-conditioning by graduated exposure to sunlight in springtime; severe cases: medically supervised conditioning/ desensitization

Third line

Hydroxychloroquine Systemic immunosuppressants: Azathioprine Omega-3 fatty acids Beta-carotene: 25mg TDS Antioxidants Nicotinamide: 1g TDS *E. coli* filtrate Thalidomide

dress covering the exposed areas to provide sun protection. Light-colored clothing is preferred over darker shades. Cotton fabric is recommended for people who work in sun. The patient advice pamphlet is given in Table 4. The various lines of management available are listed in Table 5.

Treatment should be based on the age, sex, occupation, and site of involvement. In Indian context, economic conditions also should be considered. The management designed individually to suit the patients can be successful in the treatment and prevention of the disease.

Sun avoidance and Sun Protection

Sun avoidance is the only definitive way of preventing PMLE. But this is not possible in a largely agricultural country like India. Sun protection may be a useful alternative. Sun protection should be advised as a preventive measure during summer months with sun avoidance between 11.00 am and 3.00 pm accompanied by regular application



Figure 8: Hyperpigmented plaque on the neck

of sunscreens. Patients should also be advised regarding the use of protective clothing and should be informed that a tight and wet fabric increases the transmission of UV light.^[33]

Sunscreens

Broad-spectrum sunscreens of the new generation have high SPF and they also provide protection against longer wavelength UVA. Hence, their regular use will provide partial or complete protection against PMLE in almost 90% of patients. In contrast, the old generation sunscreens protect primarily against UVB and does not protect against UVA can provoke PMLE. Patient education regarding proper application technique, quantity to be used, and the necessity to cover all sun-exposed sites including temples, ears, lateral and posterior neck.

In India, most of the sunscreens available are combination products which include physical and chemical sunscreens. They are available in a wide range of sun protection factors. Sunscreens are useful in the prevention of PMLE in patients. But it has been observed that most individuals are not aware of sunscreens and they do not apply sunscreens properly.^[52-54]

Further application is necessary after sweating. In the Indian subcontinent where humidity is high and most of the patients are laborers, the role of sunscreen is debatable.

Topical Corticosteroids

High potent topical steroids can be used in patients with mild episodes of PMLE to relieve itch. When used in combination with phototherapy or photo-chemotherapy, they help in reducing the severity and incidence of rash associated with treatment. Further in localized lesions occurring in specific areas, topical steroids are particularly useful.^[4]

Topical Antioxidants

A combination of topical antioxidants like alphaglycosylrutin, ferulic acid, and tocopherol acetate was found useful in PMLE. This protects against the inflammation reactions that are most likely to be mediated by the generation of free radicals in the skin.^[54]

Systemic Corticosteroids

Systemic steroids are indicated in acute and severe cases. Daily prednisolone in the dosage of 25 mg for 4–5 days at the onset may help to settle the attacks. Prednisolone 1 mg/kg for 1–2 weeks may be initiated during acute and severe exacerbations. Prolonged courses of prednisolone must be avoided due to its potential long term side effects. Therefore, it can be cautiously used in patients with occasional, symptomatic attacks of PMLE.^[55]

Photochemotherapy

The frequency and severity of PMLE decreases with summer progression as a result of desensitization phenomenon called "hardening." This phenomenon is used in the treatment of PMLE. For mild cases, a self-conditioning programme by graduated exposure to sunlight is recommended. For severe cases, medically supervised conditioning is preferential. In Indian context, with increasing summer, the hardening occurs as a natural phenomenon.^[55]

The mechanism of induction of photoprotection is probably due to the following reasons:

- a. induction of melanization
- b. induction of epidermal thickening
- c. UV induced immunomodulatory and anti-inflammatory effects

Antimalarials

Hydroxychloroquine 400 mg daily for the first month followed by 200 mg daily for 12 weeks has shown to have a mild benefit in PMLE with a reduction in the severity of the rash. Its membrane stabilizing properties cause proteolytic enzyme inhibition. Hydroxychloroquine 400 mg/day has to be started a few days prior to exposure and it should be reduced to 200 mg/day after optimal drug levels have been reached. In an Indian study, the efficacy of hydroxychloroquine in PMLE has been demonstrated in the trial showing reduction in eruption. Short-term treatment courses with hydroxychloroquine seem to be well-tolerated with a minimized risk of ocular lesions.^[56,57,58,59]

Systemic Immunosuppression

Systemic immunosuppressants like azathioprine may be recommended in PMLE in the following situations:

- a. extreme sun sensitivity
- b. intolerance to phototherapy
- c. patients in whom sunscreens are ineffective

The azathioprine was used in the dose of 50mg to 150mg and was effective in a small series of patients but it takes few weeks to act. Cyclosporine has also been found to act in severe cases of PMLE.^[60]

Thalidomide has been used with some success in certain cases, but the serious adverse side-effects associated with this medication have limited its use.^[61]

Conclusion

Polymorphous light eruption is a common disease with varied presentation in the Indian skin. Large scale studies are sparse in this largely neglected disease. It has to be differentiated from its close mimics and appropriately managed. A concord in management between the patient and treating physician is cornerstone for successfully overcoming this disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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