CLINICAL VIGNETTE

Polymorphous Light Eruption

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A 45-year-old woman presented with an erythematous petechial rash on her left lateral forearm that had started the preceding day, accompanied by burning, itching, and mild swelling. She noted some chills but no fever or rash elsewhere. She had applied topical hydrocortisone without improvement. Pertinent history included surgery 14 days prior to remove fragments of ceramic from her left palm. Post-operatively she had a rash in the area of providone-iodine prep, but it resolved after 2-3 days. Her hand wound had healed well. Otherwise she reported no new exposures other than a sunny drive to San Diego over the weekend. Examination and symptoms were concerning for herpes zoster infection in the setting of recent surgical stress. However, distribution and appearance were not classic.

Discussion

Polymorphous light eruption (PMLE) is an idiopathic primary photosensitivity that is the most common light-induced skin disease. It presents as a rash on sun-exposed skin that occurs within hours to days after sun exposure and lasts for several days before dissipating.[1] The rash is termed polymorphous because it can present in various forms although individuals tend to develop the same pattern of outbreak with each occurrence. The rash typically occurs and is most severe in the spring and early summer, after a winter period of minimal sun exposure, and lessens with progressive exposure during the summer, resolving in autumn or winter, and recurring the next spring. Lesions usually heal without scarring.

Epidemiology

PMLE occurs in all age groups and skin types although it is more common in younger adults, typically occurring in the second and third decades of life, and more common in fair skin than in in dark skinned individuals. The prevalence in the general population is 10-15% and may be higher than 20% in areas of central Europe and Scandinavia.[2] There is a female preponderance versus male (2:1). Although the actual cause of PMLE is unknown, genetic factors may play a role, with an increased concordance in monozygotic twins and a positive family history in up to 40% of cases.[2-4] A higher prevalence of PMLE among younger women suggests hormonal factors may be involved.[5] Individuals exposed to sunlight throughout the year rarely acquire PMLE and continued sun exposure leads to suppression of the condition over time.

Pathogenesis

PMLE is a type IV delayed hypersensitivity reaction to a compound in skin altered by exposure to UV radiation. This leads to impaired T-cell function and a reduction in the normal UV-induced immune suppression in the skin.[6] This abnormal response to UV exposure produces an inflammatory response to an endogenous photo-induced antigen which does not occur in non-affected individuals. The photoantigen causing PMLE has not been identified.[6]

Although PMLE is provoked by UVA radiation in 75-90% of cases, the rash can occur with exposure to sunlight through window glass. Standard sunscreens also may not be preventative. UVB, the chief cause of sunburn, and visible light also can provoke PMLE.[7] Sunburn reaction in individuals with PMLE is normal.

Clinical Features

Skin lesions typically appear within hours after sun exposure, although they may not occur for several days. The eruption appears on sun-exposed areas of the body, most commonly the V of the chest, the outer forearms, backs of hands, and lower legs. The face is usually spared. Lesions are accompanied by intense pruritus and can last for several days. They may not clear completely until the end of summer. Photosensitivity decreases with increased sun exposure, a phenomenon known as “hardening”, which involves increased melanin production, thickening of the stratum corneum, and possibly a normalization of the cutaneous cell-mediated immune response.[8]

PMLE presents as pruritic papules, papulovesicles, or plaques. The papular type is most common, appearing as dense clusters of 2 – 5 mm red bumps. Papules can coalesce into erythematous, raised, rough patches (plaques) from which groups of fluid-filled vesicles can arise. Less commonly lesions may be target-shaped and resemble erythema multiforme.[9] The rash is usually characteristic for each individual, appearing similar with each recurrence. The most common initial symptoms are itching, burning, and redness. Some individuals also experience headache, chills, lethargy, and nausea several hours after exposure, although symptoms typically last only 1 – 2 hours.

Diagnosis

PMLE is usually a clinical diagnosis based on the appearance of a rash, typically erythematous papules and plaques, with onset within hours after exposure to sunlight and clearance after a few days, generally occurring after an extended period of no sun exposure (e.g., seasonal). The differential may include: 1) herpes simplex varicella (HSV) with vesicular lesions; 2) solar urticaria, a hives-type rash that appears after brief sun exposure and disappears quickly when out of sun exposure; 3) photo-exacerbated dermatoses, which are not caused by the sun but which can flare with sun exposure (e.g., lupus erythematosus, rosacea); 4) drug-induced photosensitivity, which resembles sunburn and can occur on any skin, including the face; and 5) metabolic photodermatoses, such as porphyrias, which are caused by enzymatic defects that can lead to blisters and scarring on skin. Biopsy findings in PMLE show upper dermal edema and dense perivascular lymphocytic infiltrate. Direct immune fluorescence is negative.[7]

Treatment

Although PMLE resolves without intervention, symptomatic treatment includes oral antihistamines to control pruritus and high potency topical steroid creams. Oral corticosteroids may be considered for individuals with severe eruptions.

Prevention and Prognosis

Sun protection, including clothing and high SPF (50+) sun-screens, are primary prevention strategies. Desensitization phototherapy may be helpful, with repeated, controlled pre-exposure UV light treatments to promote skin hardening. Anti-malarial drugs (hydroxychloroquine), immune suppressing drugs (azathioprine), and pre-exposure systemic steroids have been tried for prevention, but have not become standard.[6] While PMLE may be associated with emotional distress, it resolves without treatment, leaves no skin scarring, and tends to improve with aging.

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