Polymorphic light eruption

BRUCE TATE PhD, FACD

A holiday in the sun is disrupted by an unusual itchy rash.

Case presentation

A 17-year-old girl presented after returning from a 10-day holiday in north Queensland. Unfortunately, the holiday had been disrupted when she developed a very itchy rash that she initially thought was from insect bites. It had appeared mild a few days after she arrived, but had then worsened substantially.

The patient described bite-sized itchy red papules and 'small lumps' that seemed to turn into hives on her face, lips, neck, upper trunk and arms. She had noticed a few mosquitoes and lots of sandflies in north Queensland. She had spent a lot of time in the sun during her holiday, although she did apply sunscreen regularly and wore a hat.

When the patient attended the clinic on her return to Melbourne she had vesicles on her lips, and red papules and confluent plaques on her face, neck, upper trunk and shoulders (Figure 1). It settled quickly with a short course of prednisolone 37.5 mg a day and mometasone furoate cream twice a day. She gave a history of a milder version of the same rash starting in December of the previous two summers in Melbourne – each time after she had spent a weekend in the garden. Those episodes settled with mometasone furoate cream, although the rash tended to recur occasionally over the summer months. The patient was otherwise well and taking no other medications.

Diagnosis

The diagnosis of polymorphic light eruption (PLE; also called polymorphous light eruption) was made.

Dr Tate is a Dermatologist in St Albans, Vic, and with the Skin and Cancer Foundation of Victoria.

Differential diagnoses

Insect bite reactions

There was a possibility that the rash was due to a reaction to an insect bite given the patient's exposure to insects, but the history of previous summertime episodes and the distribution of the rash points strongly to PLE. Sandflies (biting midges) are common in tropical and temperate parts of Australia and New Zealand and can be very persistent. Their bites are relatively resistant to treatment and more potent topical corticosteroids are often required.

Photosensitive drug eruptions

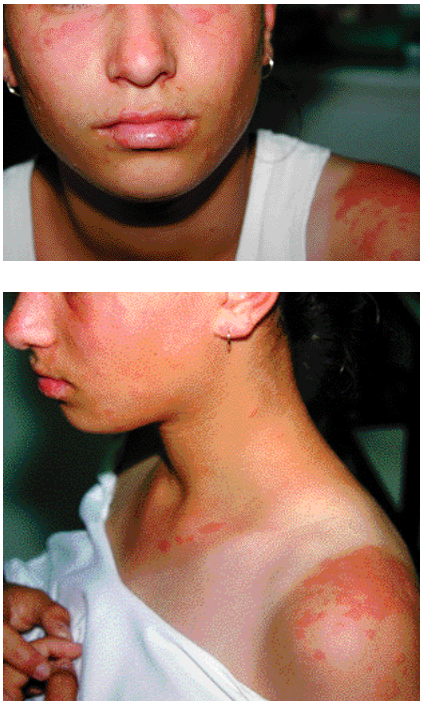
In the present case it was easy to rule out photosensitive drug eruptions as a cause of the rash because the patient was not taking any medications. These eruptions can be allergic or toxic in nature depending on the agent. They are usually not papular like PLE and usually involve all sun-exposed sites – typically face, neck, the V of the chest, hands and forearms. The most common cause of photoallergic drug eruptions is hydrochlorothiazide, often as part of a combination antihypertensive drug. The most common cause of phototoxic eruptions are tetracycline antibiotics, particularly doxycycline.

Solar urticaria

Solar urticaria is a rare form of physical urticaria, in which itchy hives appear minutes after sun exposure. It can be difficult to distinguish from PLE but delayed onset is not a feature of solar urticaria. It may be stimulated by ultraviolet (UV) A, UVB or visible light. Usually, it is not triggered by drugs or connective tissue diseases.

Lupus erythematosus

There are myriad presentations of the relatively uncommon disorder lupus erythematosus. The systemic form can resemble PLE although more typically presents with a butterfly distribution of facial erythema with photosensitivity. Usually other systemic features such as joint pains or organ involvement would be present to point to the diagnosis as well as positive lupus related blood tests. The discoid form of cutaneous lupus erythematosus is more common and only a small proportion of patients with this type have systemic lupus erythematosus. In discoid lupus erythematosus, small to large, slightly scaly red plaques form with a tendency to scar later. It is most often seen on the face.



Figures 1a (top) and b (bottom). Polymorphic light eruption on the face, neck and chest of a 17-year-old girl.

Photoaggravated dermatitis

Photoaggravated dermatitis is uncommon and can be seen with both atopic and seborrhoeic dermatitis. Photocontact dermatitis can be:

- phototoxic, with the culprit most often being psoralen chemicals found in various plants (certain weeds such as

wild parsley, angelica, fig and occasionally citrus)
- photoallergic where sunlight is required to alter the chemistry of the offending contact agent to make it allergenic. These are delayed reactions. The most common culprits are chemicals in sunscreens but occasionally some chemicals found in perfumes cause a photoallergic reaction. Photoallergic contact dermatitis is also an uncommon problem.

Other idiopathic photodermatoses

There are many types of idiopathic photodermatoses but they are beyond the scope of this article. They include actinic prurigo,[1] chronic actinic dermatosis[2] and hydroa vacciniforme.[3]

Erythropoietic protoporphyria

Erythropoietic protoporphyria is a rare genetic porphyria. Sun exposure causes rapid onset of pain to the sun-exposed skin, but usually no rash. Specific blood tests can detect elevated red cell protoporphyrin levels.

Comment

PLE is common but its estimated prevalence varies according to latitude: up to 22% in northern Europe, 5% in Australia and 1% in Singapore. Most often it is seen in young adults and more often in women. All ages are susceptible and dark skin offers no protection.

The rash typically takes days to appear if it occurs during a sunny holiday in the winter months. Subsequently, after the first episode or during the spring or summer months the rash most often starts six to 12 hours, but sometimes only minutes, after sun exposure. The degree of sun exposure needed to trigger the rash is quite variable. As the summer (or less often the holiday) progresses, PLE becomes less active. It may also occur while skiing or when using a solarium.

As its name suggests, the clinical picture is variable in presentation, although for any given person the episodes tend to be similar. Most often itchy small red or skin-coloured papules or vesicles, small red nodules about the size of an insect bites or larger smooth to rough papular plaques appear. The papules may be almost confluent. Occasionally small blisters form or the skin can be itchy with minimal rash.

PLE mainly affects sun-exposed sites on the face, ears, neck (sparing under the chin), V of the chest, arms and hands or other areas depending on how much clothing is worn. 'Sun-hardened' sites, such as those that receive a lot of sun like the face and back of the hands, may be spared and it may occur under clothing if these offer little sun protection. A variant of PLE is juvenile spring eruption seen mainly in boys and usually affecting their ears with a papular/vesicular red rash.

Systemic symptoms such as malaise, headache, fever and nausea rarely occur in PLE. Symptoms usually improve over the years or entirely cease but may be life-long. There is an association with PLE and various forms of lupus erythematosus. PLE is more common in patients with lupus erythematosus and in relatives of patients with the disorder,[4] although this disorder is uncommon in people with PLE. A study from a large photodermatology clinic in Germany found that 12% of patients with PLE at some time had an elevated level of antinuclear antibodies and 0.6% were also Ro-antibody positive. Of the 70% of patients followed up, no one later developed lupus erythematosus.[5]

Histopathology is not diagnostic but can be helpful to rule out other conditions such as lupus erythematosus. In patients with PLE, histopathology shows a deep superficial perivascular infiltrate of lymphocytes and sometimes neutrophils and eosinophils. Dermal oedema can be marked, sometimes with vesicle or bulla formation. Interface change as seen in connective tissue diseases is not a feature.

Research into PLE is relatively limited. Based on immunohistochemistry and other work, it seems likely that it is a delayed hypersensitivity reaction. In 1994, heat shock proteins were suggested as a possible antigen[6] but the antigen(s) is still not known, although it is most likely to be an endogenous molecule altered by the sun to become immunogenic. Also, the sun has an immune suppressing effect on the skin, so it is possible that PLE is caused by a partial failure of this immunosuppression allowing a reaction to an antigen. It is also possible that sunlight alters the immune regulatory environment in the skin, allowing a suppressed allergy to manifest. Research supports a role for reactive oxygen species in the genesis of PLE.

Patients with PLE may be more likely than controls to have functional mutations in glutathione S-methyltransferase enzymes, which are important in protecting the skin from free radical damage.[7] Laboratory phototesting shows most patients are reactive to ultraviolet light, either long wavelength (UVA) or short wavelength (UVB) or a combination of the two but are rarely reactive to visible light. Most do not react much to monochromatic light, because they require a broader range of wavelengths to provoke PLE. Sunscreens block UVB much more efficiently than UVA and minimally block visible light. The degree of photoprotection offered by clothing is also variable. This explains why sun protection may be insufficient in protecting people with PLE.

Diagnosis is usually clinical, sometimes with the help of skin biopsy and, if indicated, blood tests to rule out connective tissue diseases or porphyrias. There are some specialised dermatology clinics where phototesting with broad-spectrum or monochromatic light sources can be used to try and characterise the wavelengths of light that trigger PLE. This testing can also be helpful to look into other photodermatoses. Photo-patch testing to rule out photoallergic contact dermatitis is also available in a few facilities.

Treatment

Sun protection, including the use of broad-spectrum sunscreen with a high sun-protection factor (30 or higher), is important but may not be sufficient on its own to prevent PLE. If the stimulating wavelength is known, this information can be used to tailor sun protection. For example, if the PLE is triggered by light through glass, it is likely to be due to UVA, which is less well filtered by most sunscreens. Reflective sunscreens containing titanium dioxide or iron oxide are most efficient in such cases but still may not be sufficient.

Clothing is variable in its ability to protect the skin from UVA or UVB. Broad-brimmed hats alone are estimated to reduce light exposure on the face by about 40%. Addition of the antioxidants α-glucosylrutin plus vitamin E to a broad-spectrum sunscreen was substantially better than the sunscreen alone in preventing onset of PLE.[8,9] This product is available overseas but is not sold in Australia. Depending on the situation, it is prudent to check baseline and later vitamin D_3 levels if the patient is particularly diligent in sun protection.

Potent topical corticosteroids will often settle an attack within a few days to a week but will usually be needed repeatedly for those with more active disease. Patients should use mometasone furoate (Elocon, Novasone) or betamethasone dipropionate (Diprosone Dermatologicals, Diprosone OV, Eleuphrat) cream twice a day on non-facial sites and methylprednisolone aceponate (Advantan) on the face until it settles and as needed for recurrences. For a more substantial bout of PLE, patients should use a short course (three to 10 days depending on severity) of prednisolone (Panafcortelone Tablets, Predsolone Tablets, Solone) 37.5 mg a day. Dose weaning is not required. Topical corticosteroids are then used to treat milder recurrences. Occasionally patients need other treatments to deal with more severe or oral corticosteroid-dependent PLE. Options in such cases are listed below:

- Hydroxychloroquine (Plaquenil) 200 to 400 mg a day is a slow-acting agent whose efficacy is good to excellent in about 70% of those treated[10]
- Paradoxically, narrow band UVB phototherapy in a gradually escalating dose protocol over four to eight weeks can be very useful in reducing the severity of attacks. Treatment is started in the early spring to try and prevent the onset of significant PLE. If the rash does not normally develop on the face, the face is shielded during treatment
- In those with debilitating attacks and who have no contraindications to therapy, azathioprine (Azahexal, Azamun, Azapin, Imuran, Thioprine) 100 to 150 mg a day or cyclosporin (Cicloral, Neoral Sandimmun) less than 5 mg/kg a day can be used.

PLE is not a PBS indication for cyclosporin, so it is an expensive treatment. The use of β-carotene as an antioxidant has been advocated for treating PLE but not shown to be effective. Also the herbal antioxidant product Heliocare containing extracts of *Polypodium leucotomos*, green tea and β-carotene has also been suggested to be useful but there has been no formal research of its role in PLE treatment.

Summary

PLE is a common photodermatosis that can usually be easily managed with sun protection and topical or oral corticosteroids. Other causes of photosensitivity should be considered in the differential diagnosis. Occasionally more debilitating cases occur requiring other treatments. MT

References

1. Castanedo-Cazares JP, Moncada B, Torres-Alvarez B, Lepe V. Actinic prurigo. Available online at: http://www. emedicine.com/DERM/topic927.htm (accessed November 2008).
2. Sebastian QL, Del Rosario R. Hydroa vacciniforme. Available online at: http://www. emedicine.com/ DERM/topic181.htm (accessed November 2008).
3. Chronic actinic dermatitis. Available online at: http://dermnetnz.org/ reactions/chronic-actinic-dermatitis.html (accessed November 2008).
4. Millard TP, Lewis CM, Khamashta MA, et al. Familial clustering of polymorphic light eruption in relatives of patients with lupus erythematosus: evidence of a shared pathogenesis. Br J Dermatol 2001; 144: 334-338.
5. Tzaneva S, Volc-Platzer B, Kittler H, et al. Antinuclear antibodies in patients with polymorphic light eruption: a long term follow up study. Br J Dermatol 2008; 158: 1050-1054.
6. McFadden JP, Norris PG, Cerio R, et al. Heat shock protein 65 immunoreactivity in experimentally induced polymorphic light eruption. Acta Derm Venereol 1994; 74: 283-285.
7. Millard TP, Fryer AA, McGregor JM. A protective effect of glutathione-S-transferase GSTP1*Val105 against polymorphic light eruption J Invest Dermatol 2008; 128: 1901-1905.
8. Hadshiew IM, Treder-Conrad C, Bulow Rv R, et al. Polymorphous light eruption (PLE) and a new potent antioxidant and UVA-protective formulation as prophylaxis. Photodermatol Photoimmunol Photomed 2004; 20: 200-204.
9. Jeanmougin M, Peyron JL, Thomas P, et al. Polymorphic light eruption: prophylaxis using a topical combination of antioxidants and UVA protection formulations. Ann Dermatol Venereol 2006; 133: 425-428.
10. Pareek A, Khopkar U, Sacchidanand S, et al Comparative study of efficacy and safety of hydroxychloroquine and chloroquine in polymorphic light eruption: a randomized, double-blind, multicentric study. Indian J Dermatol Venereol Leprol 2008: 74: 18-22.

Further reading

Polymorphic light eruption. Available online at: http://dermnetnz.org/reactions/pmle.html (accessed November 2008).
Scheinfeld NS, Shirin S, Del Rosario R. Polymorphic light eruption. Available online at: http://www.emedicine.com/ DERM/topic342.htm (accessed November 2008).

COMPETING INTERESTS: None.