

Athena speciality certificate examination case for photodermatology

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A 7-year-old girl presented with a 2-year history of episodic pruritic erythematous papules on her right cheek, which coalesced to form a plaque (Fig. 1). This gradually resolved over a few weeks. The eruption subsequently recurred, also involving the left cheek, the end of the nose and upper arms. No other sites were affected. The rash persisted and worsened throughout the summer despite sunscreen with sun-protection factor 50, but she also had less severe episodes in winter and a similar rash started to develop on her buttocks. She was not on any medications and had no other medical history. Investigations included monochromator phototesting, which showed abnormal ultraviolet (UV) A sensitivity at 24 h after irradiation.

Question

Which of the following is the most likely diagnosis?

- (a) Actinic prurigo.
- (b) Erythropoietic protoporphyria.
- (c) Hydroa vacciniforme.
- (d) Polymorphic light eruption.
- (e) Solar urticaria.

Answer

- (a) Actinic prurigo.

Discussion

The distribution of the rash and the worsening in summer hints that light/photosensitivity plays a role.

Actinic prurigo (AP) is a rare immunological photodermatosis, which typically presents with pruritic papules, sometimes vesicles, and plaques with excoriations and scars over photodistributed sites. Infrequently, it also affects unexposed sites such as

the buttocks. Ocular involvement and cheilitis may occur but is less frequently reported in UK cases than in those from South America. AP is more common in females and is much more frequently reported in people of Latin American or Amerindian descent with the human leucocyte antigen (HLA) DR4 subtype (HLA DRB1*0407).¹ A positive family history may be present in 75% of this patient cohort, whereas it typically occurs in approximately 50% of cases in the UK. Our patient was positive for HLA DRB1*0407 but had no known family history or Latin American/Amerindian ancestry. AP may spontaneously resolve by the late teens but can also persist into adulthood.²

Erythropoietic protoporphyria (EPP) typically presents with immediate pain and oedema on light-exposed sites and will usually present in early childhood. The genetic background of EPP is typically that of inheritance of a low-expression allele polymorphism in addition to a recessive mutation in the gene encoding the enzyme ferrochelatase. This can cause accumulation of a haemoglobin precursor, protoporphyrin IX, which is a photosensitizer to visible light. This significantly accumulates in red blood cells, plasma, liver and skin. Thus, photoactivation of protoporphyrin IX causes phototoxic insult, resulting in tissue damage, which manifests as skin pain and sometimes oedema, erythema and waxy scarring on photoexposed sites caused by photosensitivity to visible light. If visible light exposure is not prolonged, the patient will usually have symptoms of pain only on exposed sites, with visible skin changes evident only with more prolonged or repeated exposures. Protoporphyrin IX localizes within the erythrocytes as opposed to the basement membrane localization of uroporphyrinogen III in porphyria cutanea tarda.^{3,4}

Hydroa vacciniforme is a very rare chronic, scarring photodermatosis. It is associated with Epstein–Barr virus infection⁵ in some cases, although not all and its aetiology is poorly understood. It affects children aged 3–15 years; it can affect boys and girls whereas it may develop later in boys and continue for longer. Affected children present with symmetrical, pruritic, often haemorrhagic crusted, oedematous papules, vesicles/bullae

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Figure 1 (a) A 7-year-old girl with pruritic erythematous papules on the right cheek, which (b) coalesced to form a plaque; (c) on subsequent recurrence, the eruption spread to the left cheek and later involved the end of the nose, upper arms and buttocks.

on photodistributed sites, presenting 15 min to a few hours after UV light exposure. The lesions heal with varioliform scarring, usually improving by late teens.

Polymorphic light eruption (PLE) is the most common immunological photodermatosis. PLE occurs in 18% of the population in northern Europe, with 20% of cases presenting in childhood, and usually occurs only in spring and summer. Typically, several hours of sun exposure may be required to trigger an eruption later in the day or on the following day. Generally, the rash lasts from a few days to up to 2 weeks, often resolving without scarring. Papules and vesicles are the typical morphology of PLE (it sometimes can have a varied appearance) and commonly, it is limited to photoexposed sites. In children, the face is usually involved. The phenomenon of 'hardening' is often seen in PLE, with habitually exposed sites, such as the face and dorsum of the hand, becoming less affected. Patients are able to 'harden' themselves over the summer by low-level repeated exposure, which can improve tolerance, and sometimes hospital-based phototherapy can be used to induce hardening.⁶

Solar urticaria is a rare immunological photodermatosis, which can be most disabling as it typically presents within minutes of light exposure. The urticarial eruption will occur on light-exposed sites and is typically triggered by both UVA and visible light, so can occur throughout the year and can be triggered with light through clothing or windows and sometimes by artificial light. Lesions usually resolve within an hour and do not result in scarring. The underlying cause is usually idiopathic but photoactive drugs, lupus and cutaneous porphyrias must be excluded.

Conflict of interest

The authors declare that they have no conflict of interest.

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None.

Ethics statement

Ethics approval was not applicable. The patient provided informed consent for publication of their case details and images.

Data availability

Data are available under Green Open Access.

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