

Solar urticaria

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ABSTRACT

A case of solar urticaria is presented, followed by a discussion of the clinical characteristics, pathophysiology, diagnosis, and management of this disease. Special emphasis is given to clinical pearls and pitfalls for the practicing allergist. Solar urticaria is a physical urticaria that can be difficult to diagnose and distinguish from other photodermatoses. There are some characteristic features that are important to remember when evaluating a patient with suspected solar urticaria. Testing can be difficult without the assistance of an experienced dermatologist because there are several different wavelengths of light that can lead to a patient's symptoms. Solar urticaria tends to be a chronic disease with a low 5-year resolution rate but can usually be effectively managed with multiple antihistamines.

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Key words: Chromophore, dermatographism, erythrocytic protoporphyria, minimal urticarial dose, photodermatoses, phototherapy, physical urticaria, polymorphic light eruption, solar urticaria

Question

A 28-year-old African American woman presents with a 6-year history of hives on sun-exposed areas of her body. The lesions erupt rapidly after 15–30 minutes of direct sunlight and resolve spontaneously over the course of several hours. The severity of symptoms is directly proportional to the intensity of the sunlight. She denies lesions on non-sun-exposed areas or episodes of urticaria not elicited by sun exposure. She denies associated angioedema or recurrence of symptoms with other physical stimuli such as pressure, cold, vibration, exercising, or hot showers. Her exam reveals dark skin (phototype 5) without evidence of dermatographism, rash, urticaria, angioedema, or chronic changes and no other significant physical findings. She has self-treated her condition with diphenhydramine, achieving partial response, but sunscreen does not seem to prevent or delay the onset of hives. Which of the following is true?

- A. This patient's diagnosis is consistent with polymorphic light eruption.
- B. Her action spectrum is most likely to be in the visible light range.
- C. This condition is inherited in an autosomal recessive pattern.
- D. Her condition will likely resolve over the next 5 years.
- E. This condition is more common in African American populations than in white populations.

INTRODUCTION

Solar urticaria is a rare physical urticaria that should be considered in patients with a history of recurrent urticaria after direct exposure to sunlight or other light sources.^{1,2} First described in 1904, solar urticaria comprises 0.08–0.5% of all cases of urticaria, based on retrospective analyses.^{3,4} The diagnosis of solar urticaria can be confirmed by reproducing symptoms with light exposure. Identifying the light spectrum and minimal urticarial dose (MUD) required to trigger symptoms will help guide management options.⁵ The average delay from symptom onset to diagnosis of solar urticaria ranges from 4 to 11 years in various studies.⁴

CLINICAL CHARACTERISTICS AND PATHOPHYSIOLOGY

Solar urticaria is characterized by the appearance of a wheal and flare reaction on sun-exposed skin within a relatively short period of time, usually <30 minutes.

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The lesions are evanescent over the next few hours, occasionally taking up to 24 hours to resolve. Erythema and urticaria are often preceded by itching or burning at the site of exposure. Exposure time to provoke symptoms and disease severity can be influenced by many factors such as season, temperature, time of day, and latitude.⁶ Skin more commonly exposed to light has been shown to be less sensitive than areas more commonly kept covered and can often become more resistant to the sun as the year progresses, a process known as "hardening."³ In these patients, longer exposures are required to elicit symptoms. This phenomenon is the basis that underlies phototherapy. Angioedema and systemic symptoms including anaphylactic shock have rarely been reported in some cases of solar urticaria.⁷ Dermatographism is common, found in 25% of patients in one series of 57 cases.⁸ Most studies show a female preponderance with an average age of onset in the third or fourth decades of life and no familial pattern of inheritance.³

Although the exact mechanisms underlying solar urticaria are unknown, the prevailing theory is an IgE-mediated disease in which mast cells are activated by the presence of a substance known as a chromophore. Chromophores become immunologically active after exposure to a particular part of the electromagnetic spectrum in each patient. This is known as the action spectrum. Variable action spectra among patients contribute to the idea that multiple chromophores exist. One study showed 60% of patients were sensitive only to visible light, 12.5% reacted to visible light plus UVA and UVB, 10% reacted only in the UVA range, and 10% reacted only in the UVB range. The MUD also varies widely in patients studied. Serum total IgE, however, is usually within the normal range. Solar urticaria can also be caused by exogenous substances, such as chlorpromazine, secondary to metabolic diseases, as in erythropoietic protoporphyria (EPP), and coexistent with other photodermatoses, such as with polymorphic light eruption (PMLE).⁹ This usually precedes solar urticaria, sometime by several years, occasionally delaying the diagnosis because patients may not recognize the development of a new disease process.⁶

DIAGNOSIS

The patient with solar urticaria most often presents to the physician without visible evidence of skin abnormality. Confirmatory testing with phototesting should be performed with visible light (slide projector or monochromator), UVA, UVB, and sunlight if necessary to establish an MUD. The wavelength of light that elicits symptoms is known as the action spectrum. Although a majority of patients will develop symptoms in <15 minutes of sun exposure, some patients required much higher doses of radiation, and some

patients even require heat in addition to light from a particular action spectrum. This sometimes explains why patients will have negative tests initially to phototesting, if not performed adequately. Occasionally, direct sunlight testing is necessary to achieve results.⁵ A few patients have been identified reacting only to the infrared spectrum of light, not found in UVA, UVB, or visible light testing.³ Phototesting is most efficiently performed by an experienced dermatologist with follow-up with the allergist 48–72 hours after testing. This will allow the patient's disease severity to be quantified, provide a baseline measurement for response to therapy, and direct disease management or identify other disease processes, such as PMLE. Unlike solar urticaria, PMLE is characteristically more delayed in onset with resolution of symptoms over several days or weeks and is more prevalent in African American populations than in white populations.¹⁰ The rash morphology is less urticarial and more papular, plaque-like, or papulovesicular. If there are elements of the history suggestive of systemic lupus erythematosus, autoantibody screening should be performed. Lesions in systemic lupus erythematosus, however, are not classically pruritic and will persist for weeks. Once described as type VI solar urticaria, EPP is a metabolic disorder due to ferrochelatase deficiency, evidenced by an elevated erythrocyte protoporphyrin level. EPP should be considered when the diagnosis is made in a child, when there is a familial pattern of inheritance, or when the lesions are described as painful.

MANAGEMENT

Second-generation antihistamines should be considered as first-line therapy for solar urticaria.⁵ In patients not responding initially to daily second-generation antihistamines, the addition of multiple antihistamines twice daily, plus daily montelukast, has been shown to be well tolerated and beneficial in patients with severe forms of solar urticaria. These medications can likely be tapered after 1–2 months as hardening is achieved.¹¹ It should be kept in mind that antihistamines may only block the wheal and flare but not the erythema associated with solar urticaria.³ The addition of broadband sunscreen can be beneficial for patients with UVB and some UVA action spectra but is largely ineffective in patients sensitive to visible light.⁵ For patients that do not respond to antihistamines plus sunscreen, phototherapy or photochemotherapy is an effective option, although there are concerns about high cumulative doses of ultraviolet radiation with these therapies.¹¹ Patients receiving phototherapy must maintain treatment every few days or therapeutic effects will wane. Intravenous immunoglobulin and plasmapheresis have also been used effectively in selected patients with severe and treatment-resistant solar urticaria.³

Little prognostic data for solar urticaria is available, although a few studies have reported on long-term follow-up of these patients. The disease course is most often chronic with slow resolution or improvement. In a case series including 87 patients, Beattie *et al.* showed a resolution of symptoms in 12% of patients at 5 years, 26% of patients at 10 years, and 36% at 15 years after diagnosis with another one-third of patients showing clinical improvement over the same period of time.⁶ Botto *et al.* showed resolution of disease in 46% of patients at 15 years.³

Answers to the Questions:

- A. False—This clinical picture is more consistent with solar urticaria than PMLE.
- B. True—Less than 20% of patients studied tested negative to the visible light spectrum.
- C. False—This disease is not known to have a familial pattern of inheritance.
- D. False—Five-year resolution has been estimated to range from only 12–15%.
- E. False—Solar urticaria is more common among white populations, although PMLE is more common among African Americans.

Pearls

- Solar urticaria should be suspected when urticarial lesions occur rapidly on sun-exposed skin.
- Combining multiple second-generation antihistamines is usually required for effective therapy.
- Solar urticaria is usually a chronic disorder but can be managed effectively to resolve or improve symptoms in the vast majority of cases.

Pitfalls

- Solar urticaria must be differentiated from other photodermatoses such as PMLE or porphyria.

- Patients with solar urticaria may sometimes have coexisting photodermatoses.
- Consider exogenous causes or metabolic disorders such as EPP.
- Various parts of the UV, visible, and infrared light spectrum can trigger symptoms in different patients, rendering some therapies useless in certain cases and leading to false negative results with some testing modalities. Phototesting must be performed to visible light, UVA, UVB, and sometimes direct sunlight.

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ERRATUM

In “Perception of acute airway function changes by patients with mild asthma” by Kestutis Malakauskas, Sandra Ragaisiene, and Rainundas Sakalauskas, in the May–June 2009 issue, Vol. 30, No. 3, page 277, the last author’s name was misspelled. It should be Raimundas Sakalauskas.

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