Solar Urticaria

A Report of 25 Cases and Difficulties in Phototesting

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Background: Solar urticaria is a rare photosensitive disease, and its differential diagnosis with respect to polymorphous light eruption is sometimes difficult. We report our experience with 25 cases of solar urticaria and discuss the pitfalls in phototesting such patients.

Observation: The most important locations in this patient series are the V of the neck and the arms, which are similar to those of polymorphous light eruption. In all of the patients, however, the lesions appeared within 30 minutes of sun exposure or phototesting and disappeared within 24 hours. Notably, 12 (48%) of the patients had a history of atopy. Phototesting helps confirm the diagnosis, but, in some patients, this was difficult.

Conclusions: A negative phototest result from a single light source does not necessarily exclude a diagnosis of solar urticaria. In patients in whom phototesting elicits negative reactions, other light sources should be used, and, if the phototest result is still negative, a provocative test with natural sunlight should be done. Histamine-1-receptor antihistamines are a useful first-line therapy, although more severely affected persons may require prophylactic courses of phototherapy or photochemotherapy. The main problem is maintenance treatment.

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Solar Urticaria

Defined as an idiopathic photodermatosis characterized by the occurrence of itchy erythematous or urticarial lesions that are provoked only by sunlight and disappear rapidly, nearly always within 24 hours after the avoidance of sun exposure. The severity of the reaction depends on the length of time of the exposure and the intensity of the solar irradiation.

We describe the difficulties in diagnosing solar urticaria in a series of 25 patients. The clinical features needed for the diagnosis of solar urticaria are evaluated, and we discuss the necessary confirmation by phototesting. Indeed, the reproducibility of the lesions as a diagnostic element is not always easy. Finally, we analyze the different treatment modalities.

RESULTS

Clinical Findings

In this series of 25 patients with solar urticaria, the age at first consultation varied between 17 and 71 years, with a mean age of 35 years. The solar urticaria had started from 4 to 11 years before consultation.

All the patients had itching; 9 of the 25 also had a burning sensation, and 1 even had pain. Five patients had only itchy erythematous lesions, whereas in 20 patients whealing developed with exposure to the sun, and 14 of them had both erythematous lesions and whealing. In all of the 25 patients, erythema or urticaria and itching appeared 30 minutes or less after sun exposure. The time the urticaria persisted varied from 30 minutes or less (n=7) to 1 to 2 hours (n=6) to several hours (n=12). When the sun exposure was avoided, the lesions always disappeared within 24 hours, which is the most important clinical diagnostic criterion.

The location of the lesions varies. In our series, 24 patients had lesions on the V area of the neck; 24 on the arms; 24 on the legs; 20 on the shoulders; 19 each on the trunk, the neck, the hands, and the feet; 18 on the ears; 14 on the cheeks; 9 on the forehead; and 5 on the eyelids. Thus, the most frequent locations are the V area of the neck and the arms, which is similar to those of polymorphous light eruption.

Therefore, the location of the lesions is less important in confirming the diagnosis than is their short persistence. Only in 7 (28%) of our patients was there spontaneous improvement in the summer, which means that most of them did not develop a natural tolerance during the summer. One reason for this could be that most of them did not expose their skin regularly enough to the sun out of fear of a relapse, but another reason could simply be the climate in Belgium. That most of the reported cases occur in northern countries could be due to a continuous desensitization in southern
PATIENTS AND METHODS

PATIENTS

The 25 patients with solar urticaria were diagnosed on the basis of an exhaustive list of questions about the aspect of the lesions, the location, the relationship with sun exposure and sun intensity, the duration of sun exposure needed, the time during which lesions persist after the avoidance of exposure, the evolution during the summer, the personal and familial medical history, and the use of sunscreen ointments or other preventive measures.

PHOTOTESTING

Several lamps can be used for phototesting. Fluorescent tubes are inexpensive and available with several emission spectra. A more expensive but also more physiologic technique is to use a solar simulator (a xenon lamp). The most detailed phototesting procedure is to use a xenon lamp in combination with a monochromator to determine the minimum urticarial dose (MUD) at different wavelengths. The 25 patients were tested in this way with a Clinical Phototest box with Philips Cleo lamps (Alfasun, BLS, Wolvertem, Belgium), which combines a 900-W xenon lamp with a monochromator. The following wavelengths were used: 300, 320, 350, 400, 500, and 600 nm. Those patients who reacted to 600 nm were also tested at 700 nm.

The immediate initial reading is the most important one for this condition. It is also advisable to read the tests after 6 hours because late reactions are possible, albeit rare. At a particular wavelength, the threshold dose (MUD) is the dose in millijoules or joules per square centimeter that induces a clearly visible itchy erythema or whealing during or shortly after irradiation. This reaction mostly disappears in a few minutes but may last for several hours. If the dose is below the MUD, there is no reaction or only slight immediate erythema without itching confined to the irradiated site. With doses at or slightly higher than the MUD, erythema precedes whealing. When the threshold dose is exceeded a great deal, whealing develops without the preceding erythema. In 2 of the 3 patients in whom monochromatic test results were negative, a positive reaction with itchy erythema and whealing was provoked with the use of the slide projector. Although the phototest results with a slide projector, a high-intensity UV-A source, a PUVA irradiation box, and total-spectrum xenon light remained negative in the third patient, a provocative test with sun exposure elicited a positive reaction (Table 2).

The most difficult differential diagnosis is polymorphous light eruption. The most frequent locations of solar urticaria are the V area of the neck and the arms, which is similar to those of polymorphous light eruptions, although more patients with solar urticaria also have facial lesions than do patients with polymorphous light eruptions. Unlike solar urticaria, the lag time for the appearance of polymorphous light eruption varies between 2 hours and 3 days after sun exposure, and, in the absence of further exposure, it generally takes 2 to 6 days or even longer before the lesions disappear. Solar urticaria also has to be differentiated from erythropoietic protoporphyria, in which sun-induced urticaria can be one of the symptoms. Erythropoietic protoporphyria usually starts in infancy, is often familial, and has skin pain as one of the symptoms.

The diagnosis of solar urticaria on clinical grounds is difficult because the lesions persist for such a short time, and most patients who consult a physician for this have no lesions at all. The diagnosis cannot be made with certainty merely on the basis of the history. Therefore, it has to be confirmed by phototesting, which is also necessary to determine the degree of photosensitivity and the action spectrum.

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From a practical point of view, when monochromatic phototest results are negative, it is important to continue provocative testing with other light sources. Only then may a diagnosis of solar urticaria be excluded. With the simpler tests, the precise action spectrum cannot be determined but only the diagnosis confirmed. Some patients require not only UV or visible irradiation or both to reproduce the urticarial wheals but also heat. Knowing the precise action spectrum can be important because it enables the physician to make a qualitative and quantitative evaluation of the photosensitivity, evaluate the effect of treatment, and observe the evolution of the condition over time. In addition, it can be useful in formulating guidelines for a patient.

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the predominant symptom. In addition, porphyrin dosages will show elevated protoporphyrin levels in the blood; in solar urticaria, these levels are normal by definition. Therefore, porphyrin determinations are only necessary when the solar urticaria starts in infancy, when skin pain is the predominant symptom, or when both are present.

The experimental induction of immediate whealing has also been reported in a case of porphyria cutanea tarda, but, again, the porphyrin dosages were abnormal. Cholinergic urticaria may occasionally mimic solar urticaria in that it can be precipitated by the heat occurring during sun exposure. Solar urticaria, however, will be more pronounced on the exposed skin areas, whereas cholinergic urticaria is usually more pronounced in the covered areas, where the temperature is higher. In patients with cholinergic urticaria, phototesting will elicit negative reactions.

The cutaneous lesions in lupus erythematosus may show a photodistribution, but they will not be urticarial. Moreover, lupus lesions will persist for at least several weeks, whereas solar urticaria will nearly always disappear within 24 hours after sun avoidance. Phototesting will induce immediate reactions in patients with solar urticaria and may induce delayed reactions in patients with lupus erythematosus. Finally, test results for antinuclear and Ro skin-sensitizing antibodies (Ro SSA) will be negative in patients with solar urticaria.

Once urticaria has developed, the treatment is only symptomatic. Topical or oral antihistamines can be used, as can topical steroids. The urticarial response can also be decreased by cooling the corresponding areas of the skin immediately after irradiation. It is more important to try to prevent whealing. The choice of the treatment and the response to the treatment will then depend mainly on 2 factors: the action spectrum to which the patient reacts and the severity of the lesions as expressed by the MUD.

Table 1. Values of the Minimal Urticarial Dose in Joules per Square Centimeter at Different Wavelengths in 25 Patients With Solar Urticaria as Determined With a Xenon Lamp and a Monochromator After Immediate Reading*

<table>
<thead>
<tr>
<th>Patient</th>
<th>Wavelengths, nm†</th>
<th>300±5</th>
<th>320±10</th>
<th>350±32</th>
<th>400±32</th>
<th>500±32</th>
<th>600±32</th>
<th>700±32</th>
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<td>−</td>
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<td>≤35</td>
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</tbody>
</table>

* The minus sign means that the phototest results at this wavelength are negative; ND, not done.
† The plus or minus sign signifies the band width in nanometers of the monochromator at this wavelength.

Table 2. Induction of Whealing in Patients With Solar Urticaria Using Different Light Sources*

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Monochromatic Phototesting, 300 to 600 nm†</th>
<th>Slide Projector</th>
<th>UVASUN 3000</th>
<th>PUVA Irradiation Box</th>
<th>Total-Spectrum Xenon Light</th>
<th>Sunlight</th>
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<td>−</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>+</td>
</tr>
</tbody>
</table>

* See the text for the descriptions and manufacturers of the different light sources. A minus sign indicates no wheal; a plus sign, whealing.
† See Table 1 (details are given for 300, 320, 350, 400, 500, and 600 nm for these 3 patients).
Antimalarial agents, oral β-carotene, and oral steroids are of little value in the treatment of solar urticaria. Antihistamines are still the therapy of first choice because histamine may be an important mediator in solar urticaria. Antihistamines can reduce the whealing and itching in many patients. Especially interesting are the antihistamines of the new generation, among which the H1-receptor antihistamines are generally superior. Many studies conducted to date have been done with terfenadine. To obtain good therapeutic results, much higher doses than the conventional dosage are needed (180-360 mg/d), with which success rates as high as 77% have been reported. Edema and itching can thus be prevented, although macular erythema can still be present. After a while, the dose can be decreased. In many cases, 120 mg/d is sufficient as maintenance treatment. A higher dosage of terfenadine than the conventional dosage can cause adverse effects and lead to rare but life-threatening cardiac dysrhythmias. The question is how this medication achieves its effect because effective doses are much higher than would be needed to only block the histamine liberator. Some patients have clinical improvement with terfenadine therapy but with little modification of the phototest results.

In patients in whom antihistamines do not give a satisfactory result, phototherapy and photochemotherapy are worth trying. Even if detailed phototesting was done before, it is advisable to determine the MED again with the lamps and, if possible, with the light box used for the therapeutic irradiations. The main reason for this is to avoid a generalized flare-up. Phototherapy has the advantage over photochemotherapy of not provoking phototoxic adverse effects and probably of not having the same adverse effects over the long term. It can be done with repeated exposures to narrowband UV-B, broadband UV-B, and a combination of UV-B and UV-A. Phototherapy can also be done with visible light. It has been shown that daily irradiations with fluorescent lamps emitting mainly blue light can provide good protection. The tolerance obtained with phototherapy decreases quickly and is mostly gone in a few days. Better results are obtained with PUVA therapy, especially in those patients for whom all other therapy was unsuccessful. In contrast with the tolerance induced by sun exposure or by UV phototherapy, the protection obtained with PUVA lasts many weeks. Usually the UV-A irradiations are given 2 hours after oral intake of 0.6 mg/kg of 8-methoxypsoralen 3 times a week. We start the regimen at a dose under the MUD and increase it by 20% twice a week. In those patients in whom the MUD is so low that PUVA treatment would be nearly impossible, pre-PUVA UV-A desensitization can be done. Before starting PUVA treatment, the patient is irradiated with UV-A without psoralens for 3 days with 6 irradiations, increasing the dosages every day hourly. In this way, the MUD will gradually increase. As soon as the MUD is at least 1.5 J/cm2 of UV-A, the classic PUVA treatment with psoralens can be started. Repeating the phototesting after 10 sessions, we have obtained spectacular increases in the MUD by a factor of as high as 200. Nevertheless, the MUD value returns to the initial level within 3 months. With this in mind, we are considering maintenance therapy with UV-A only, for example, once or twice a week. The literature is silent on this subject.

Another successful but more invasive and, therefore, less practical therapy is plasmapheresis. It can be used in those patients in whom a photoallergen can be found in plasma or serum. This treatment can also be combined with PUVA.

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REFERENCES