

Solar Urticaria Successfully Treated with Intravenous Immunoglobulins

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Key Words

Solar urticaria • Intravenous immunoglobulins • Phototest • Photobiological exploration

Abstract

Background: Idiopathic solar urticaria (SU) is a rare type of physical urticaria, occurring immediately after exposure to visible or ultraviolet (UV) light. Treatment is based on sun avoidance and on high doses of antihistamines, but is sometimes inefficient. **Methods:** We report on a 41-year-old patient with severe SU who was successfully treated with a single course of 2 g/kg of intravenous immunoglobulins (IVIG). **Results:** A dramatic improvement in UVA and UVB tolerance was rapidly observed, with an increase of up to 10 times the UVA minimal urticarial dose on day 3. The treatment with terfenadine was continued. Healing of photosensitivity was persistent since 100 days after the single course of IVIG, no urticarian reaction was provoked with polychromatic irradiation rising above 8.3 J/cm² or after UVA doses rising above 15 J/cm². **Conclusion:** Use of IVIG in severe SU can be discussed when high-dose antihistamines are inefficient and quality of life is affected.

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Introduction

Solar urticaria (SU) is a rare type of physical urticaria characterized by itching, erythema and whealing occurring immediately after exposure to visible or ultraviolet (UV) light. It may considerably restrict normal daily life because of the limitations upon outdoor activities [1]. Treatment is based on photoprotection and high doses of antihistamines, but may sometimes be inefficient.

We describe a patient with refractory severe SU who was successfully treated with a single course of intravenous immunoglobulins (IVIG) and who showed a fast dramatic normalization of provocative phototests.

Case Report

A 41-year-old man was referred to our unit for severe idiopathic SU evolving over the previous 5 years. His medical history was pertinent for asthma in infancy, obesity, hypertriglyceridemia and type 2 diabetes treated with metformin for 10 years. He had no personal or family history of photosensitivity or urticaria. Despite avoidance of the sun, use of sunscreens and high doses of antihistamines alone (terfenadine 540 mg/day) or in combination (levocetirizine 5 mg/day, fexofenadine 180 mg/day, loratadine 10 mg/day plus desloratadine 5 mg/day), the skin disease was not controlled and the patient, who was a travelling salesman, was declared unable to work.

Physical examination was normal; the patient had no clinical signs of lupus erythematosus or cutaneous porphyria. No exogenous or endogenous photosensitizer was found. Hemogram and serum protein electrophoresis were normal, and the search for antinuclear antibodies was negative. Photobiological explorations were performed with the solar simulator Dermolum® III (Müller Electronic, Moosinning, Germany). Action spectra included UVA and possibly UVB. The polychromatic minimal urticaria dose (MUD) was 2.13 J/cm² and the UVA MUD fell to 0.9 J/cm², compatible with the diagnosis of SU.

Because of the very low polychromatic and UVA MUD levels, we did not consider PUVA therapy as a therapeutic option. Considering the major effect on his daily life, we decided to start with IVIG. As the patient was obese (105 kg), we used the dose calculation for his ideal body weight, determined with Lorentz's formula (80 kg). The patient received a single course of 2 g/kg of IVIG, infused over 4 days (40 g/day; Tegeline®).

Photobiological explorations followed 7, 21 and 100 days after the last IVIG perfusion using the polychromatic phototest and the UVA phototest. The results are shown in table 1. A dramatic improvement in UVA and UVB tolerance was rapidly observed, with

an increase of up to 2.8 times the polychromatic minimal erythema dose and an increase of up to 10 times the UVA MUD on day 7. Healing of photosensitivity was persistent since 100 days after the single course of IVIG, no urticarian reaction was provoked with polychromatic irradiation doses rising above 10 J/cm² or after UVA doses rising above 15 J/cm².

Twelve months after, the patient is still receiving terfenadine, but the eruption has not relapsed, even after a long period of exposure to the sun. Each time the patient forgot terfenadine, he felt pruritus after sun exposure. However, he has regained a very good quality of life and has returned to his job.

Discussion

SU is a rare photodermatosis and must be differentiated from heat-induced urticaria, which is triggered by infrared but not UV or visible light [1]. The provocative allergen for SU remains unknown and may be different from one patient to another. It might be a chromophore localized in the epidermis, which could become antigenic after UV or visible light exposure, allowing a link with IgE. The histamine-releasing activity could be secondary to the cross-linking of mast cells and basophil leucocytes IgE receptors. The phenomenon of tolerance could be explained by the saturation of the IgE-binding sites by the allergen. First-intention treatments of SU include photoprotection and antihistamines. When high doses of antihistamines are ineffective, PUVA therapy is indicated, often after UVA desensitization. In very debilitating SU, different systemic treatments have been tested, based on the suspected existence of circulating antigen: plasma-

Table 1. Evolution of photobiological explorations before and after the single course of IVIG in our patient

Phototests	Before IVIG	Day 7	Day 21	Day 100
Polychromatic phototest				
MUD, J/cm ²	2.1	>6	10	>10
UVA phototest				
MUD, J/cm ²	0.9	>9	13	>15

IVIG = Intravenous immunoglobulins; MUD = minimal urticarial dose.

Table 2. Characteristics of patients who received IVIG for SU

Patient	Sex	Age years	Treatment received before IVIG	IVIG administration modalities	MUD before IVIG, J/cm ²	MUD after IVIG, J/cm ²	Clinical results
Darras et al. [5]	F	32	antihistamines UVA desensitization PUVA therapy	Tegeline® 0.5 g/kg (day 1) 1 g/kg (days 2, 3) 3 courses: months 1, 3, 12	1 (UVA) 0.1 (UVB)	15.6 (UVA) 1.6 (UVB)	no relapse 1 year after the 3rd course; antihistamines still required
Puech-Plottova et al. [4]	F	55	antihistamines UVB therapy PUVA therapy hydroxychloroquine corticosteroids	Sandoglobulin® 0.4 g/kg/day (5 days) 3 courses: months 1, 3, 5	0.025 (UVA)	27 (UVA)	no relapse 1 year after the 3rd course; antihistamines still required
Our patient	M	41	antihistamines	Tegeline® 0.5 g/kg/day (4 days) 1 course	0.9 (UVA) 2.13 (polychromatic)	13 (UVA) 10 (polychromatic)	no relapse 1 year after the course; antihistamines still required

pheresis, extracorporeal photochemotherapy [1] and cyclosporine [2].

Immunoglobulins, a polyvalent antibody product derived from the pooled plasma of healthy donors, are increasingly used to treat dermatological diseases, including urticaria [3]. Open studies performed on several patients affected with idiopathic chronic urticaria [3] or with delayed pressure urticaria showed 20–50% with complete response and 30% with partial remission. Two cases of refractory idiopathic SU successfully treated with IVIG have been previously reported (table 2) [4, 5]. In one case, IVIG were given alongside PUVA therapy. The administration schedule differed from one patient to another: 2–2.5 g/kg infused over 3–5 days. The number of courses necessary to obtain clinical remission varied from 1 course (our patient) to 3 courses, with variable time lags between the courses. In all cases, a strong and persistent improvement of MUD was obtained. Nevertheless, all patients still require antihistamines.

IVIG are usually well tolerated. Minor side effects, such as headache, fever and myalgia, are frequent and occur during infusion or 1–3 days after administration. Severe adverse events, such as acute renal failure, aseptic meningitis, stroke and cardiac infarction, are rare and develop mainly in patients with additional risk factors. The mode of action of IVIG in SU is unknown, but it might consist of a blockade of receptors by the fragment crystallizable region [3].

Despite their expensive cost, use of IVIG in SU must be discussed when quality of life is affected and when antihistamines are ineffective. Additional prospective studies in a larger number of patients are required to demonstrate their effectiveness in SU and to determine the optimal schedule of administration.

References

- 1 Horio T: Solar urticaria – idiopathic? *Photodermatol Photoimmunol Photomed* 2003; 19:147–154.
- 2 Edström DW, Ros AM: Cyclosporin A therapy for severe solar urticaria. *Photodermatol Photoimmunol Photomed* 1997;13:61–63.
- 3 Prins C, Gelfand EW, French LE: Intravenous immunoglobulin: properties, mode of action and practical use in dermatology. *Acta Derm Venereol* 2007;87:206–218.
- 4 Puech-Plottova I, Michel JL, Rouchouse B, Perrot JL, Dzvinga C, Cambazard F: Solar urticaria: one case treated by intravenous immunoglobulin. *Ann Dermatol Venereol* 2000;127:831–835.
- 5 Darras S, Ségard M, Mortier L, Bonnevalle A, Thomas P: Treatment of solar urticaria by intravenous immunoglobulins and PUVA therapy. *Ann Dermatol Venereol* 2004;131: 65–69.