

Solar urticaria unresponsive to intravenous immunoglobulins

Mar Llamas-Velasco, Diego De Argila, Cristina Eguren, Patricia García-Martin, Sara Ibañes & Amaro García-Diez

Department of Dermatology, Hospital Universitario de La Princesa, Fundación de Investigación Biomédica, Madrid, Spain

Summary

Key words:

intravenous immunoglobulins;
photodermatoses; phototest; solar urticaria

Correspondence:

Mar Llamas-Velasco, Department of Dermatology,
Hospital Universitario de La Princesa, C/Diego de
León 62, CP 28006, Madrid, Spain.
Tel: +349 15 202 433
Fax: +349 15 202 435
e-mail: mar.llamasvelasco@gmail.com

Accepted for publication:

18 August 2010

Conflicts of interest:

No funding sources.

The treatment of solar urticaria (SU) can be difficult. Only a few cases of SU have been treated with intravenous immunoglobulins (IVIg) (as monotherapy or combined with phototherapy), with reported fast and durable increase of solar exposure tolerance. A 61-year-old female with severe UVB- and UVA-induced SU and a 62-year-old female with severe UVA and visible light-induced SU were both treated with a single course of IVIg (total dose of 2 g/kg), infused over 3 days. Phototest, performed 3 months after the treatment, showed only a slight minimal urticating dose improvement, and both patients reported just a moderate and 'transient' subjective improvement. Our patient's poorer response, compared with previous reports, may be due to differences in IVIg's treatment schedules, which are reviewed.

Idiopathic solar urticaria (SU) is an uncommon type of physical urticaria, probably due to sensitization to an endogen photoallergen. The eliciting factor can be UVA, UVB, visible light or, rarely, infrared radiation.

We have used intravenous immunoglobulins (IVIg) as second-line treatment for two patients with refractory and disabling SU.

Case reports

Case 1

A 61-year-old woman with neither a contributory medical nor drug intake history was referred to our unit because of severe idiopathic SU, evolving over the previous 6 years. Over photoexposed areas, she developed itchy wheals, minutes after exposure, which faded away in the next few hours. Patient's full blood count, urea, electrolytes, liver function tests and serum protein electrophoresis were all within normal limits. Autoantibody screening was negative (including antinuclear antibodies, extractable nuclear antigens, anticardiolipin and anti-double-stranded DNA antibodies and porphyrin screening test). Photobiological examinations were performed using a UV light handle device with fluorescent lamps (Gigatest UVB and UVA, Fritsh, Gottesheim, Alsacia, France), discharge lamps from a UVA cabinet (Dr Höenle, Gräfelting, Germany) and a slide projector (Kodak Carousel S-AV 2050 slide projector,

Stuttgart, Germany). The first patient had a UVB- and UVA-induced SU. Intradermal injection (0.1 ml) of her autologous serum irradiated with action spectra was followed by a whealing response, and the control injection of non-irradiated serum was not, which suggested that there was a circulating photoallergen. Despite strict sun avoidance, the use of sunscreens and high doses of antihistamines, her SU was poorly controlled. She was then treated with a single course of IVIg (total dose of 2 g/kg), infused over 3 days, at a dose of 1 g/kg/day on the first day and at 0.5 g/kg/day on days 2 and 3. No adverse events were observed. Outcomes 5 days before and 3 months after the treatment are shown in Table 1.

Case 2

A 62-year-old woman with no contributory medical history, nor drug intake history, presented with severe idiopathic SU evolving over the previous 5 years. Complementary tests were all within normal limits, ruling out associated diseases. She received the same treatment as the other patient, due to her UVA- and visible light-induced SU. Her photobiological tests results, before and after the treatment, are also shown in Table 1.

Both patients reported a moderate and transient subjective improvement of sunlight tolerance. Later on, UVA phototherapy, to induce hardening, and narrow-band UVB phototherapy, were prescribed, achieving temporary phototolerance.

Table 1. Photobiological test results of our two cases

	Basal phototest	Phototest 90 days after treatment
Patient 1		
UVB MUD	61 mJ/cm ²	130 mJ/cm ²
UVA MUD	15 J/cm ²	15 J/cm ²
Visible light	Negative	Negative
Patient 2		
UVB MUD	Negative	Negative
UVA MUD	1.46 J/cm ²	5.2 J/cm ²
Visible light	Positive (10 min)	Positive (10 min)

MUD, minimal urticating dose.

Discussion

SU is a difficult to treat and disabling disease that considerably limits outdoor activities. First-line treatment with H1-blockade and sun blockers may be enough for some patients, but induction of tolerance with an artificial UV source is sometimes recommended, in more severe cases (1). Other treatment options such as plasma exchange or photopheresis are based on the suspected existence of a circulating factor (2).

IVIg is a polyvalent antibody product, derived from the pooled plasma of healthy donors, which is being increasingly used to treat dermatological diseases, including idiopathic chronic urticaria or delayed pressure urticaria (3). It is a well-tolerated drug with minor side effects like infusion reactions. The IVIg postulated action mechanism in autoimmune diseases is thought to be due to: (1) the functional blockade of the Fc portion of IgG molecules, producing an inhibition of B- and T-cell function of some receptors, (2) cytokines' modulation or (3) complement-mediated damage inhibition (2). In SU, it could also provide antiidiotypic antibodies, capable of suppressing histamine releasing autoantibodies (1). Circulating photoallergen binding to antibodies on mast cells could be blocked by IVIg; hence, IVIg could be especially useful for patients with demonstrated photosensitizers in their serum.

There have only been a few reports in the literature of SU successfully treated with IVIg (1–5). Patients were aged from 27

to 55 years and their SU disease duration varied from 1 month to 5 years (1–5). The administration schedule differed from one patient to another: 2–2.5 g/kg, infused over 3–5 days and a wide range number of necessary courses (from 1 to 6), with variable time lags (from 1 to 3 months between the courses). They were previously treated with antihistamines and photoprotection (all cases), PUVA desensitization (all except one case), oral corticosteroids (two cases) and ciclosporine (one case). All cases except for one achieved complete remission (3). Some of them maintained antihistamines along with IVIg treatment (5). In one patient, IVIg was administered in combination with PUVA (3).

We have reported on two patients with treatment-refractory and severe SU, who were treated with a single course of IVIg, with a slight and temporary subjective improvement, but with no clinically relevant increase in the minimal urticating dose. In our experience, IVIg given at a total dose of 2.5 g/kg over 3-day courses is not as effective as the literature has reported previously. The poorer response of our patients could be due to different IVIg commercial trademarks, variable dosage schedules or individual patient factors, as they have a wide action spectrum heterogeneity and we cannot rule out a positive results' publication bias. Finally, the presence of a SU circulating factor does not seem to predict a good response of SU to IVIg.

References

1. Hughes R, Cusack C, Murphy GM, Kirby B. Solar urticaria successfully treated with intravenous immunoglobulin. *Clin Exp Dermatol* 2009; **34**: 660–662.
2. Correia I, Silva J, Filipe P, Gomes M. Solar urticaria treated successfully with intravenous high-dose immunoglobulin: a case report. *Photodermatol Photoimmunol Photomed* 2008; **24**: 330–331.
3. Darras S, Segard M, Mortier L, Bonneville A, Thomas P. Urticaire solaire traitée par l'association immunoglobulines polyvalentes et PUVAthérapie. *Ann Dermatol Venerol* 2004; **131**: 65–69.
4. Puech-Plotova I, Michel JL, Rouchouse B, Perrot JL, Dzviga C, Cambazard F. Urticaire solaire: un cas traité par immunoglobulines polyvalentes. *Ann Dermatol Venerol* 2000; **127**: 831–835.
5. Maksimovic L, Fremont G, Jeanmougin M, Dubertret L, Viguier M. Solar urticaria successfully treated with intravenous immunoglobulins. *Dermatology* 2009; **218**: 252–254.