Case Report

Recalcitrant Solar Urticaria Induced by UVA and Visible Light: A Case Report

Metavee Insawang MD*,
Chanisada Wongpraparut MD*

** Department of Dermatology, Faculty of Medicine, Siriraj Hospital, Mahidol University, Bangkok, Thailand*

A 41 year-old man presented with a ten-year history of recurrent erythema and swelling of skin that occurred following sun exposure even as little as ten minutes. The lesion affected only on the sun exposed area. A phototesting was carried out and revealed that urticaria was induced following ultraviolet A (UVA) and visible light exposure. Solar urticaria (SU) from UVA and visible light was diagnosed. Many treatment options including combination of oral antihistamines, psoralen plus UVA (PUVA) photochemotherapy, narrowband UVB (NBUVB, 311 nm) phototherapy, and plasmapheresis were tried in the presented patient without significant response. Although various treatments are available, managing SU remains a challenging problem in many patients.

Keywords: *Solar urticaria, Ultraviolet, UVA, Visible light, Recalcitrant*

J Med Assoc Thai 2010; 93 (10): 1238-41
Full text. e-Journal: *http://www.mat.or.th/journal*

Solar urticaria (SU) is an uncommon form of physical urticaria that was first reported by Duke et al in 1923[1]. It is characterized by pruritic erythematous wheals that occur within minutes to a few hours after sunlight or artificial light source exposure. Occasionally, SU may present along with periorbital swelling, mucosal swelling, burning sensation, and angioedema. Severity and duration of the disease are also influenced by many factors such as the length of light exposure and light intensity[2]. The action spectrum of SU varies from ultraviolet (290-400 nm) to visible light (400-700 nm)[3,4]. Although there are many treatment modalities available, management of solar urticaria is still a major problem[5]. Here, the authors report a recalcitrant case of SU that was induced by visible light and ultraviolet A (UVA; 320-400 nm).

Case Report

A healthy 41-year-old male indoor worker presented at the Department of Dermatology, Faculty of Medicine Siriraj Hospital with a ten-year history of recurrent erythema and swelling of the skin that occurred following sunlight exposure for as little as ten minutes. The lesions were intensely erythema with mild pruritus. It affected only the sun-exposed areas mostly on the face, extensor surface of both forearms and dorsum of hands. The lesions resolved within an hour after he went indoors. He had no underlying diseases including photosensitivity, nor taking any medications before the onset of his symptoms. He did not have a family history of allergy. Physical examination was otherwise unremarkable. When he presented at our clinic, SU was suspected.

Phototesting was carried out on his back to determine the sensitivity to ultraviolet and visible light by using the following light sources; polychromatic UVA (UVASUN 3000, Mutzhas, Munich, Germany), 5-45 J/cm^2; polychromatic UVB (UV800, Walmann, Villiger-Schwenningen, Germany), 50-280 mJ/cm^2 and visible light (Kodak Carousel S-AV 2020 projector, Kodak AG, Germany) for 20 minutes. IL-1700 radiometer (International light Inc. Newburyport, MA, USA) was used for measure light intensity. Results were observed during irradiation, immediately after, 10, 30 minutes, and 1 hour after testing. In the presented patient, erythema and whealing were demonstrated immediately after phototesting by visible light test (Fig. 1) and UVA irradiation with the minimal whealing dose (MWD) for UVA at 15 J/cm^2 (Fig. 2). Solar urticaria from visible light and UVA was diagnosed in the presented patient

Correspondence to:
Wongpraparut C, Department of Dermatology, Faculty of Medicine, Siriraj Hospital, Mahidol University, 2 Prannok Rd, Bangkoknoi, Bangkok 10700, Thailand.
Phone: 0-2419-4318, Fax: 0-2411-5031
E-mail: chanisada@hotmail.com

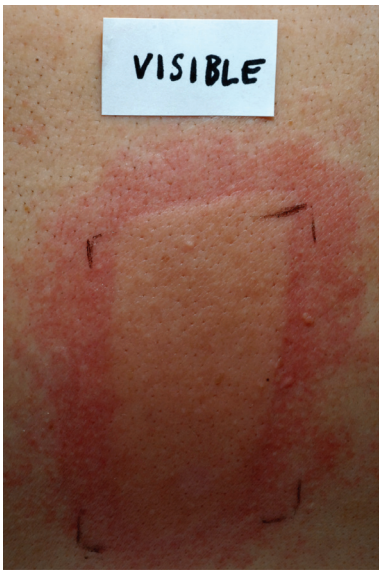

Fig. 1 Solar urticaria induced by visible light

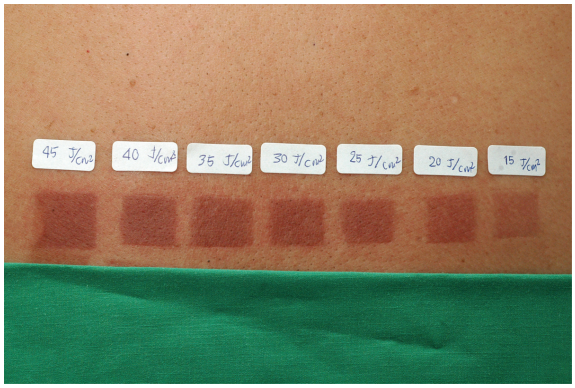

Fig. 2 Solar urticaria induced by UVA with a minimal whealing dose at 15 J/cm^2

according to the phototesting result. Antinuclear antibody was also done with negative result. Initially he was treated with H1 oral antihistamines; ceterizine 10 mg/day without any improvement. Consequently, the dose of ceterizine was increased to 20 mg/day and 180 mg/day of fexofenadine along with 10 mg/day of desloratadine were added without significant clinical improvement. Systemic PUVA photochemotherapy was then tried. However, after increasing the dose of UVA to 2 J/cm^2, his lesions were exacerbated. He was afterward received narrowband UVB phototherapy for 31 sessions without any benefit. Because of the poor improvement in prior treatments and he still felt that his lifestyle was restricted, plasmapheresis was suggested to the patient.

Intradermal serum factor testing was conducted to confirm the role plasmapheresis in the presented patient. Patient's serum was obtained and divided into two tubes. One tube was irradiated with UVA 50 J/cm^2, the other one was unirradiated. Serums from both tubes were then injected to two positions along the flexor aspect of the patient's forearm. Positive result was defined as wheal and flare response. Because of the positive result of intradermal serum factor testing in the presented patient suggested that he might have a circulating photoallergen in his serum or plasma. The authors discussed with the presented patient about the role of plasmapheresis and the patient decided to do this procedure. Before starting plasmapheresis, inform consent was obtained from the patient. Plasmapheresis was performed by conventional plasma separation with a hollow fiber membrane. Plasma was replaced by 20% solution of human albumin with the total volume of two liters each time. The schedule was given three times a week for two weeks as has been reported[6]. There was no complication such as anaphylactoid reaction observed in the presented patient.

After six sessions of plasmapheresis, photo-testing was performed to determine its effectiveness. Phototesting for MWD of UVA was repeated but the MWD did not change from the baseline (MWD of UVA 15 J/cm^2 as before plasmapheresis), however the degree of wheal was less than the baseline. Nevertheless, the presented patient still complained that his lifestyle was limited by sunlight exposure. The authors informed the presented patient to take a combination of oral antihistamines as well as wearing a long-sleeved shirt and hat when going out into the sun.

Discussion

Solar urticaria is a rare photodermatoses. It affects all races with a slight female predominance[2]. Most of the cases occur in the second to fourth decade. Atopic history was presented in a patient with SU ranged from 0-48%[2].

To date, the pathogenesis of SU is still unclear. Most of the papers were discussed mainly on action spectra, inhibition spectra and augmentation spectra of light[2,7,8]. Usually, each case has his or her own unique action spectra[9,10]. It was explained through the difference of patient's chromophore, geography and ethnicity[11]. According to Leenutaphong's

hypothesis, which supports immunologic nature of this disease, there are two types of SU. Solar urticaria type I is caused by IgE-mediated hypersensitivity to specific photoallergens and SU type II is caused by IgE-mediated hypersensitivity to a nonspecific photoallergen[7].

Management of SU is challenging[12]. Some authors proposed that SU is one of the most difficult photodermatoses to manage[3]. This may because the patients had their own action spectrum and small doses of exposure to UV and visible light were enough to provoke urticaria[5]. Various therapies of SU were described. The first line of treatment is oral H1 antihistamine[5]. For SU, the new generation of H1-receptor antihistamine is usually superior to the old generation antihistamine[13,14]. The effective dose needed in treating of SU is generally higher than conventional dosage for treating respiratory allergies[2,3]. To date, there are many reports of SU, however most of them were controllable by oral antihistamines. Recalcitrant cases of SU as in the presented case have rarely been reported.

Phototherapy by using ultraviolet exposure with an artificial light source to induce tolerance is another way to treat SU patients[15,16]. At least 22 patients had been reported successfully treated with phototherapy or photochemotherapy. Different types of phototherapy and photochemotherapy had been used for SU including PUVA, UVA alone, combination of UVA and UVB, broadband UVB and narrowband UVB[16-24]. The presented patient was tried for PUVA photochemotherapy and narrowband UVB phototherapy, unfortunately, he failed to respond to these treatments.

Plasmapheresis is one of the treatment modalities for recalcitrant SU patients[25,26]. The mechanism of this treatment was explained through removal of the patient's circulating serum factors or photoallergens[6]. To the authors' knowledge, eight patients have been treated with plasmapheresis. Six of them were reported successfully treated[2,6,25-27]. Collins et al suggested that plasmapheresis should only be used for patients who have a demonstrable circulating serum factor[27]. Because of its expensive cost with variable results and possible side effects, this type of treatment for SU is still in limited use. As far as the authors know, there is no previous report of successful treatment by using plasmapheresis in patients with SU in Thailand. Unfortunately, plasmapheresis did not make benefits to the presented patient. Other treatments that have been recently reported for a recalcitrant case of SU included intravenous immunoglobulin and anti-IgE therapy[28,29]. However, because of the financial problem, the presented patient could not afford these expensive new therapies.

In conclusion, the authors report a refractory case of SU induced by UVA and visible light who did not respond to medications, phototherapy, photochemotherapy, and plasmapheresis. Although various treatments are available, managing SU remains a major problem in many patients.

References

1. Duke WW. Urticaria caused by light. JAMA 1923; 80: 1835-8.
2. Botto NC, Warshaw EM. Solar urticaria. J Am Acad Dermatol 2008; 59: 909-20.
3. Roelandts R, Ryckaert S. Solar urticaria: the annoying photodermatosis. Int J Dermatol 1999; 38: 411-8.
4. Webb LM, Mikita CP. Solar urticaria. Allergy Asthma Proc 2009; 30: 563-5.
5. Roelandts R. Diagnosis and treatment of solar urticaria. Dermatol Ther 2003; 16: 52-6.
6. Leenutaphong V, Holzle E, Plewig G, Kutkuhn B, Grabensee B. Plasmapheresis in solar urticaria. Dermatologica 1991; 182: 35-8.
7. Leenutaphong V, Holzle E, Plewig G. Pathogenesis and classification of solar urticaria: a new concept. J Am Acad Dermatol 1989; 21: 237-40.
8. Leenutaphong V, Holzle E, Plewig G. Solar urticaria: studies on mechanisms of tolerance. Br J Dermatol 1990; 122: 601-6.
9. Leenutaphong V, von Kries R, Holzle E, Plewig G. Solar urticaria induced by visible light and inhibited by UVA. Photodermatol 1988; 5: 170-4.
10. Tuchinda C, Leenutaphong V, Sudtim S, Lim HW. Fixed solar urticaria induced by UVA and visible light: a report of a case. Photodermatol Photoimmunol Photomed 2005; 21: 97-9.
11. Horio T, Yoshioka A, Okamoto H. Production and inhibition of solar urticaria by visible light exposure. J Am Acad Dermatol 1984; 11: 1094-9.
12. Uetsu N, Miyauchi-Hashimoto H, Okamoto H, Horio T. The clinical and photobiological characteristics of solar urticaria in 40 patients. Br J Dermatol 2000; 142: 32-8.
13. Diffey BL, Farr PM. Treatment of solar urticaria with terfenadine. Photodermatol 1988; 5: 25-9.
14. Rajatanavin N, Bernhard JD. Solar urticaria: treatment with terfenadine. J Am Acad Dermatol

1988; 18: 574.
15. Plewig G, Holzle E, Lehmann P. Phototherapy for photodermatoses. Curr Probl Dermatol 1986; 15: 254-64.
16. Collins P, Ferguson J. Narrow-band UVB (TL-01) phototherapy: an effective preventative treatment for the photodermatoses. Br J Dermatol 1995; 132: 956-63.
17. Bernhard JD, Jaenicke K, Momtaz T, Parrish JA. Ultraviolet A phototherapy in the prophylaxis of solar urticaria. J Am Acad Dermatol 1984; 10: 29-33.
18. Wolska H, Kleniewska D, Kowalski J. Successful desensitization in case of solar urticaria with sensitivity to UVA and positive passive transfer test. Derm Beruf Umwelt 1982; 30: 84-6.
19. Parrish JA, Jaenicke KF, Morison WL, Momtaz K, Shea C. Solar urticaria: treatment with PUVA and mediator inhibitors. Br J Dermatol 1982; 106: 575-80.
20. Holzle E, Hofmann C, Plewig G. PUVA-treatment for solar urticaria and persistent light reaction. Arch Dermatol Res 1980; 269: 87-91.
21. Addo HA, Sharma SC. UVB phototherapy and photochemotherapy (PUVA) in the treatment of polymorphic light eruption and solar urticaria. Br J Dermatol 1987; 116: 539-47.
22. Dawe RS, Ferguson J. Prolonged benefit following ultraviolet A phototherapy for solar urticaria. Br J Dermatol 1997; 137: 144-8.
23. Darras S, Segard M, Mortier L, Bonnevalle A, Thomas P. Treatment of solar urticaria by intravenous immunoglobulins and PUVA therapy. Ann Dermatol Venereol 2004; 131: 65-9.
24. Beissert S, Stander H, Schwarz T. UVA rush hardening for the treatment of solar urticaria. J Am Acad Dermatol 2000; 42: 1030-2.
25. Bissonnette R, Buskard N, McLean DI, Lui H. Treatment of refractory solar urticaria with plasma exchange. J Cutan Med Surg 1999; 3: 236-8.
26. Hudson-Peacock MJ, Farr PM, Diffey BL, Goodship TH. Combined treatment of solar urticaria with plasmapheresis and PUVA. Br J Dermatol 1993; 128: 440-2.
27. Collins P, Ahamat R, Green C, Ferguson J. Plasma exchange therapy for solar urticaria. Br J Dermatol 1996; 134: 1093-7.
28. Puech-Plottova I, Michel JL, Rouchouse B, Perrot JL, Dzviga C, Cambazard F. [Solar urticaria: one case treated by intravenous immunoglobulin]. Ann Dermatol Venereol 2000; 127: 831-5.
29. Guzelbey O, Ardelean E, Magerl M, Zuberbier T, Maurer M, Metz M. Successful treatment of solar urticaria with anti-immunoglobulin E therapy. Allergy 2008; 63: 1563-5.

รายงานผู้ป่วย 1 ราย โรคลมพิษจากแสงแดดถูกกระตุ้นด้วยรังสีอัลตราไวโอเลต เอ และ แสงที่มองเห็นได้ที่ดื้อต่อการรักษา

เมธาวี อินสว่าง, ชนิษฎา วงษ์ประภารัตน์

ผู้ป่วยชายไทย อายุ 41 ปี มาโรงพยาบาลด้วยผื่นผิวหนังที่มีลักษณะบวมแดงยุบ เป็น ๆ หาย ๆ มาเป็นเวลา 10 ปี รอยโรคเกิดหลังถูกแสงแดดแม้เพียง 10 นาที และเกิดเฉพาะบริเวณที่ถูกแสงแดด ผู้ป่วยได้รับการทดสอบแสงและพบว่าผื่นลมพิษเกิดจากรังสีอัลตราไวโอเลตเอและแสงที่มองเห็นได้ ผู้ป่วยได้รับการวินิจฉัยเป็นโรคลมพิษจากแสงแดด คณะผู้รายงานได้ให้การรักษาผู้ป่วยหลายวิธี ได้แก่ การให้ยาต้านฮิสตามีนรับประทานหลายชนิดร่วมกัน การรักษาโดยการฉายรังสีอัลตราไวโอเลต เอ ร่วมกับรับประทานยา psoralen (PUVA) การฉายรังสีอัลตราไวโอเลต บี ชนิดคลื่นแคบ (311 นาโนเมตร) รวมถึงการรักษาโดยการเปลี่ยนถ่ายพลาสมา แต่ผลการตอบสนองต่อการรักษายังไม่เป็นที่น่าพอใจ แม้จะมีการรักษาหลายชนิดที่สามารถให้ในผู้ป่วยโรคลมพิษจากแสงแดด แต่การดูแลรักษาผู้ป่วยกลุ่มนี้ยังคงเป็นปัญหาอยู่ในปัจจุบัน