Experimentally Confirmed Induction of Sweet's Syndrome by Phototesting

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Accepted March 2, 2011.

Sweet's syndrome or acute febrile neutrophilic dermatosis is characterized by acute onset of painful, erythematous plaques and histological findings of a dense neutrophilic infiltrate without evidence for primary vasculitis. In 30–50% of cases Sweet's syndrome is associated with an underlying disease. In addition, several drugs can induce Sweet's syndrome. Rarely, Sweet's syndrome can be trigged by environmental factors. We report here a case in which, during the course of the disease, ultraviolet (UV) exposure became the main trigger of Sweet's syndrome, which could be experimentally induced by UV phototesting.

CASE REPORT

A 78-year-old Caucasian man presented with a 10-month history of erythematous well-demarcated papules and plaques on the head, upper trunk, lower arms, and legs, consistent with Sweet's syndrome. During the course of the disease skin lesions developed in sun-exposed areas, such as the forehead (Fig. 1A). The lesions were described as itching and almost painful.

The patient's medical history revealed a coronary heart condition and hypertension. In addition, the patient had been diagnosed with Hashimoto's thyroiditis. With the assumption that the skin lesions were drug-induced, the primary care physician had already changed the patient's antihypertensive medication, but without success. Colchicine was subsequently initiated, in combination with prednisolone; however, lesions recurred when the prednisolone dose was reduced to less than 10 mg per day.

The patient's work-up upon admission revealed anaemia, leukocytopaenia, and later during the course of disease thrombocytaemia. A bone marrow biopsy was taken, which was consistent with myelodysplastic syndrome without overt leukaemia. Since, during follow-up the skin lesions were localized in sunlight-exposed areas, we assumed an influence of ultraviolet radiation. Therefore, phototesting was performed on the upper back on three consecutive days with UVA1 (80 J/cm^2; UVAsun, Sellmeier, Germany), UVB (120 mJ/cm^2 = 1.5 MED, MED = 80 mJ/cm^2; UV 801 BL, Waldmann, Germany), and a combination of both UVA and UVB as described (1). Four weeks after phototesting the patient presented with erythematous itchy plaques and papules, which had already started to develop 72 h after UV exposure in the phototesting area on the back (Fig. 1B), and which were histologically consistent with Sweet's syndrome. In addition to oral prednisolone (1 mg/kg body weight) and colchicine 50 mg three times a day topical therapy with a high-protection sunscreen was used. This treatment regimen led to complete healing of all skin lesions, and systemic medication was reduced stepwise.

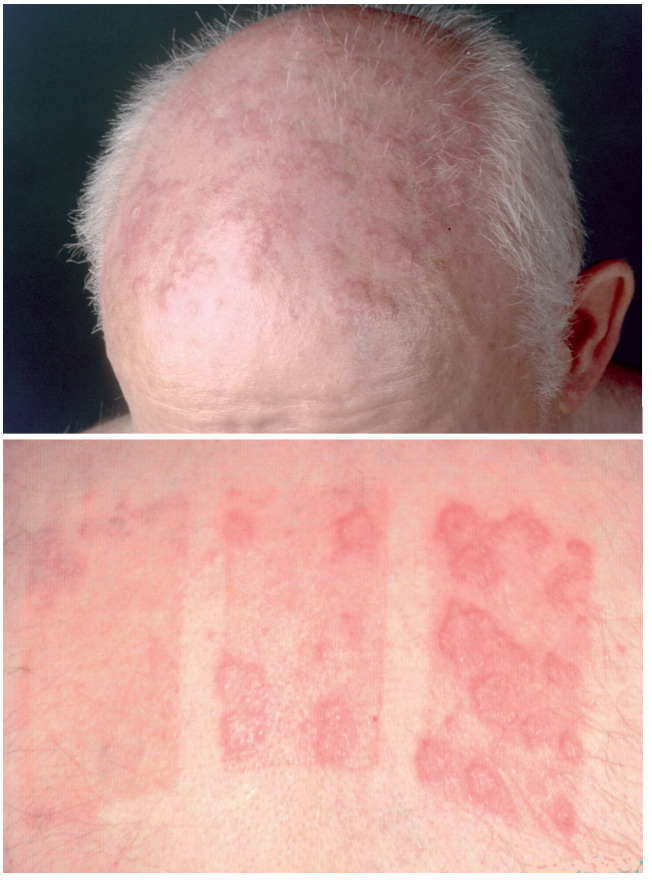

Fig. 1. (A) Erythematous confluent papules and plaques on the forehead after sun exposure. (B) Oedematous confluent papules and succulent plaques consistent with Sweet's syndrome in the phototesting area on the back. Experimental phototesting was performed from left to right with ultraviolet 1 (UVA1), a combination of UVA1 and UVB, and UVB on three consecutive days.

DISCUSSION

Classical Sweet's syndrome is an acute febrile neutrophilic dermatosis with an underlying disease in up to 50% of cases, which is in accordance with the myelodysplastic syndrome in our patient (2–4). In addition, an increasing number of drugs has been reported to induce Sweet's syndrome (5). However, drug-induced Sweet's syndrome appears to play a rather minor role in this case, since after onset of skin lesions several drugs prescribed for cardiac insufficiency or thyroid function

had been changed, but did not influence the lesion appearance. During the course of the disease lesion development correlated increasingly with sun exposure. Accordingly, lesions could even be experimentally induced by UVA1, UVB, and both UVA1 plus UVB phototesting. These findings therefore strongly indicate an important role of UV irradiation in the induction of Sweet's syndrome in our patient. UV-induced Sweet's syndrome is a rare condition, which has been reported to mimic photoallergic contact dermatitis or rosacea (6, 7). In one report Sweet's syndrome was inducible by UVA1, but not UVB phototesting, and the lesions developed after 4 days (8). In another report Sweet's syndrome lesions could be induced in the healed skin of previous lesions and the adjacent area of lesions by irradiation with 5 MED UVB (9). Interestingly, in one patient Sweet's syndrome lesions were inducible only by monochromatic UVB 290 nm irradiation (10). UV exposure can induce several inflammatory skin disorders, such as solar urticaria, polymorphous light eruption (PLE) or cutaneous lupus erythematosus (CLE) (1, 11). After phototesting lesion development normally occurs in solar urticaria minutes after UV exposure, hours to several days later in PLE, and 8–10 days in photosensitive CLE (1). In the present case skin lesions induced by phototesting and consistent with Sweet's syndrome had already started to develop after 72 h, and continued to develop and persisted over several weeks (Fig. 1B). UV irradiation is usually associated with immunosuppression, which is mediated by inhibitory cytokines and/or regulatory T-cell subsets (11). However, UVB irradiation in particular is also able to induce pro-inflammatory mediators in the skin, such as prostaglandin E_2, interleukin-1 alpha, tumour necrosis factor-alpha, and granulocyte colony-stimulating factor (G-CSF) (9, 12). Interestingly, drug-induced Sweet's syndrome most commonly occurs in patients receiving G-CSF therapy (5). Therefore, we speculate that UVB-induced cutaneous G-CSF expression might play a role in UV-induced Sweet's syndrome. Accordingly, skin lesions were primarily induced in experimentally UVB irradiated skin areas (Fig. 1B). Another pathomechanism in photosensitive Sweet's syndrome could include the isomorphic Koebner reaction also described in this dermatosis, with lesion development localized in scars, injection, inflammatory or trauma sites (8). In conclusion, our findings suggest that Sweet's syndrome should be considered in the differential diagnosis of lesions presenting in UV-exposed skin areas and persisting over several weeks.

ACKNOWLEDGEMENT

This publication is dedicated to Prof. Luger's 60th birthday.

The authors declare no conflicts of interests.

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