

cysts filled with keratin material (Fig. 1b). The lobules were embedded in an abundant fibrous stroma. Mitotic activity was not high, but a few normal mitoses were observed in the tumour. These findings led to a diagnosis of trichoepithelioma.

Trichoepithelioma is a hamartoma of the pilosebaceous apparatus.¹ It occurs either as multiple lesions or as a solitary lesion.⁸ There is controversy regarding trichoepithelioma and trichoblastoma (also known as trichoblastic fibroma) and whether they are overlapping or distinct entities.^{9,10} Our patient had no family history of trichoepitheliomata such as common multiple trichoepithelioma, which is inherited as an autosomal dominant trait. She had multiple nodules on her face where common multiple trichoepithelioma usually appears, but these nodules were much larger than those of common trichoepithelioma.

After Czernobilsky reported a case of GST² in 1972 as a rare variant of trichoepithelioma, several cases of GST have been reported.³⁻⁷ GST arises as a solitary lesion mainly on the lumbosacral, thigh or perianal region, and not on the face. Taking these facts into consideration, a new term such as 'multiple symmetrical giant trichoepithelioma' might be suitable for describing the condition seen in our patient.

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Photosensitivity in Sweet's syndrome: two cases that were photoinduced and photoaggravated

SIR, Sweet's syndrome is a neutrophilic dermatosis which frequently affects photoexposed areas. However, the triggering or aggravating role of solar irradiation has not been mentioned in the large series reported.^{1,2}

Phototesting data in Sweet's syndrome with an action spectrum in the UVB range is not very extensive in the previous literature.^{3,4} We report two cases of photosensitive Sweet's syndrome with, for the first time, a UVA1 (340-400 nm) phototriggering obtained in one case.

Patient 1

A 54-year-old woman, skin type II, presented a cutaneous eruption that recurred three times, 3-4 days after intense exposure to the sun in June 1999 and July 2000 at a French coastal resort and in February 2000 in Guadeloupe. The eruption started on the anterior part of thighs with secondary diffuse extension (posterior part of the thighs, legs, abdominal area, back and arms which were sun-exposed), sparing the face and consisted of dull-red plaques or nodules. The eruption was associated with fever (38.5 °C), joint pains and elevated neutrophils (10368/mm³). The lesions resolved over 2 weeks with systemic corticosteroids or, for the last flare, spontaneously. The histological study showed the appearance of Sweet's syndrome with a subepidermal oedema and a dense neutrophilic infiltration in the upper and deep dermis with nuclear dust. A mild perivascular lymphohistiocytic infiltrate was also present in the upper and deep dermis. She had also a history of benign summer light eruption (BSLE)⁵ since the age of 25 years, less active at present, which started one day after strong sun exposure and affected the V area of the chest most. Photobiological testing was performed with two types of equipment: (i) a solar simulator (Dermolum UM-W[®], Müller Elektronik-Optik, Moosinning, Germany) equipped with two lamps (a 1000-W Xenon lamp and a 1000-W metal halide lamp), a water filter (absorbing the infrared beam), a dichroic mirror (stopping 95% of the wavelengths up to 600 nm), a Schott WG 305-1 mm filter (cutting the wavelengths shorter than 280 nm) and a thermopile giving the total irradiance of this polychromatic spectrum including the UVB range, UVA range, and almost all the visible range (polychromatic radiant energy: 55 mW cm⁻²); (ii) a high-pressure UVA lamp (UVA-700[®] Waldmann, Reichstett, France), emitting in the UVA1 range (340-400 nm) with an incorporated dosimeter (UVA1 radiant energy: 60 mW cm⁻²). All photobiological tests were performed on the back of the patient as usual, except the repeated UVA1 phototests which were performed both on the back and on the anterior part of the thigh (the site previously affected by Sweet's syndrome). For the phototests, polychromatic irradiation was performed on a 49 cm² area and UVA1 irradiation on a 23 cm² area. The polychromatic minimal erythema dose (MED) was normal at 720 mJ cm⁻² (normal value \geq 400 mJ cm⁻²). The simple UVA1 phototest (13 J cm⁻²)

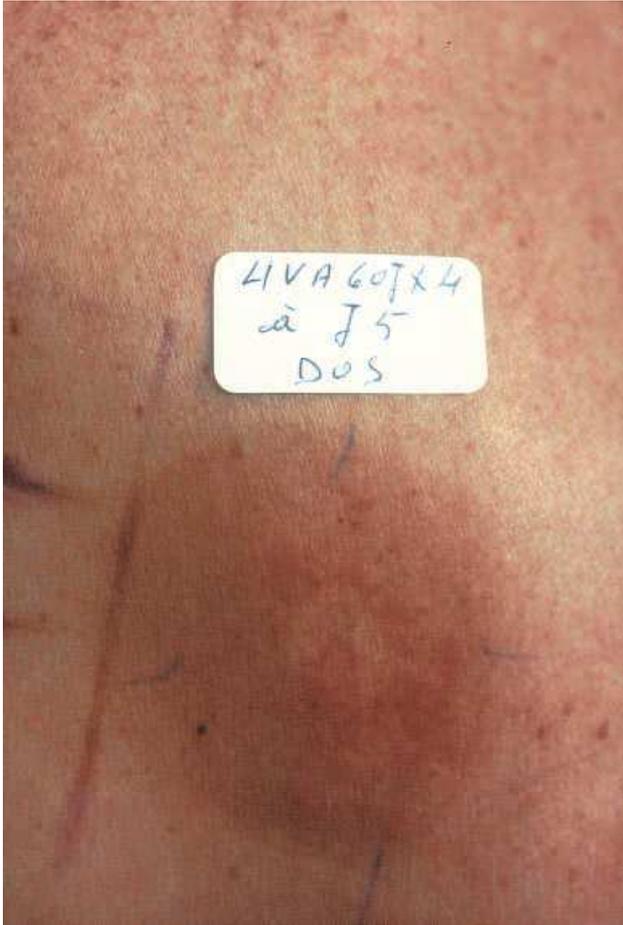


Figure 1. Repeated UVA1 phototest ($60 \text{ J cm}^{-2} \times 4$ days) at the 5th day (Patient 1).

was negative at 24 h. The repeated polychromatic phototest ($3 \text{ MED} \times 3$ days) was negative on the fourth day with a phototoxic erythema. The repeated UVA1 phototest ($60 \text{ J cm}^{-2} \times 4$ days) performed on the back and on the anterior part of the thigh was positive on the 4th day with an erythematous papular reaction on the pigmented area of the phototest. The reaction was stronger on the 5th day (Fig. 1). Histologically, the photoinduced lesions showed a neutrophilic infiltration with nuclear dust and a perivascular lymphohistiocytic infiltrate in the upper dermis (Fig. 2). There were no associated diseases. Lupus serology was negative.

Patient 2

A 61-year-old woman presented typical lesions of Sweet's syndrome with painful erythematous, purple, well demarcated plaques, strictly photodistributed, clearly limited by the clothes, 3 days after a mild sun exposure in May 1992, with fever ($38.5 \text{ }^\circ\text{C}$ to $40 \text{ }^\circ\text{C}$), abdominal pain and asthenia (Fig. 3, 4). Histological study showed typical lesions of Sweet's syndrome with a subepidermal oedema and a dense

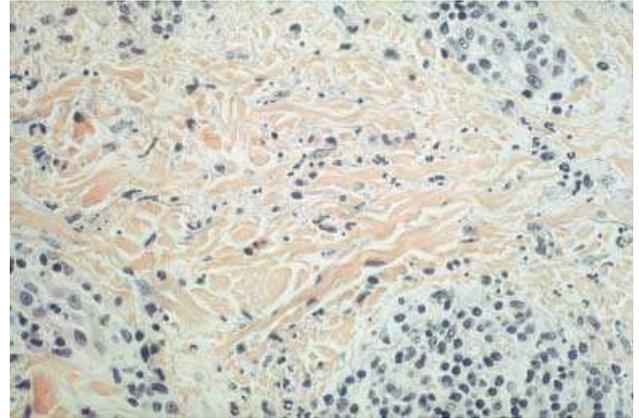


Figure 2. Histological aspect of the UVA1 photoinduced lesions (haematoxylin and eosin $\times 40$) (Patient 1).



Figure 3. Sun-exposed distribution of lesions of Sweet's syndrome (Patient 2).

neutrophilic infiltration in the dermis. The laboratory data showed an elevated erythrocyte sedimentation rate (100 mm in the first h), elevated neutrophils ($8.25 \times 10^9 \text{ L}^{-1}$) and an IgA κ monoclonal gammopathy secondary to an IgA myeloma. The polychromatic irradiation (UVB, UVA, visible ranges) was performed with a single 1000-W Xenon lamp (Dermolum III[®] Müller Elektronik-Optik) filtered with a Schott WG 295 filter. The UVA1 irradiation was performed with a high pressure UVA lamp (2000-W, Sunlab[®] 340–400 nm). The intensity, measured with the Centra UV[®] dosimeter (Osram[®], Munich, Germany) was 1.4 mW m^{-2} in the UVB range and 40 mW cm^{-2} in the UVA1 range. The UVB MED was normal at 56 mJ cm^{-2} (normal value $\geq 25 \text{ mJ cm}^{-2}$). The simple (13 J cm^{-2}) and repeated ($40 \text{ J cm}^{-2} \times 3$ days) UVA1 phototests and the repeated polychromatic phototest ($3 \text{ MED} \times 3$ days) performed on the back were negative. The polychromatic (5 MED) and UVA1 (13 J cm^{-2}) irradiations performed in previously affected areas were negative too. Thus, phototesting was negative in this case, whereas the



Figure 4. Lesions of Sweet's syndrome in sun-exposed distribution (Patient 2).

patient reported a photosensitivity for which she regularly used sunscreen on her face with good results (no lesions on the face), and explaining the photodistributed lesions.

We report two cases of Sweet's syndrome with photosensitivity. The first case was photoinduced by a strong summer sun exposure and by UVA1 phototesting. The second case was not photoinduced or phototriggered by phototesting but only photoaggravated with presence of photosensitivity and photodistributed lesions. There are few data on the photosensitivity in Sweet's syndrome in the literature. Horio³ reported two cases with photoaggravation. One case was aggravated during the summer and the second case was photoinduced by strong sun exposure. In one case, the similar clinical lesions were triggered by supra UVB MED irradiation (5 MED) and by a chemical irritation test performed with 10% sodium lauryl sulphate solution at the site previously involved and by 5 UVB MED at the adjacent area of lesions. Furthermore, the same irradiation induced an abnormal papular reaction at uninvolved skin. The histological study of these lesions showed a perivascular lymphohistiocytic infiltrate with a smaller proportion of neutrophils and nuclear dust in the

dermis. These clinical and histological aspects of the photo-induced lesions were very similar to those that we obtained in our first case. More recently, Bessis et al⁴ reported one case photoinduced by strong summer sun exposure. The polychromatic and monochromatic UVB irradiation at 290 nm triggered typical clinical and histological lesions of Sweet's syndrome. The localized phototests performed with longer UVB and UVA irradiation were negative. The pathomechanism of photosensitivity in Sweet's syndrome could involve an isomorphic Koebner reaction classically described in this dermatosis with lesions localized on old vaccination areas or scars and/or recent injection or trauma sites. The direct action of UVB on neutrophil activation and recruitment in the skin through the production of cytokines such as IL-8 and TNF- α and through the upregulation of E-selectin⁶ has also been reported in normal human skin after UVB exposure.

The BSLE of French authors corresponds with a minor form of polymorphous light eruption.⁵ The action spectrum is in the UVA range and phototriggering can be obtained with high and repeated UVA irradiation performed in the area usually affected.^{5,7} Despite the presence of a dermal perivascular lymphohistiocytic infiltrate, we did not trigger BSLE lesions in our patient because the repeated UVA1 phototests were not performed in the areas usually involved by her BSLE and prior to the histological appearance of the phototriggered lesions with presence of a neutrophilic infiltrate in the dermis and nuclear dust. Furthermore, we observed the same perivascular lymphohistiocytic infiltrate in spontaneous lesions in our first case and such an infiltrate has already been reported in Sweet's syndrome lesions in association with⁸ or preceding⁹ the typical dermal neutrophilic infiltration.

We report for the first time an UVA1 phototriggering in Sweet's syndrome. Our results do not suggest a Koebner phenomenon previously evoked, as the lesions were obtained without the induction of an actinic erythema. We recommend phototesting in Sweet's syndrome. Such patients need to be aware of the harmful effects of the sun on their dermatosis. Strict external broad-spectrum (UVB-UVA) photoprotection must be used as well as avoidance of sun exposure during the sunny months.

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Panniculitis in a patient with chronic myelogenous leukaemia treated with imatinib

SIR, The tyrosine kinase inhibitor imatinib (Gleevec, formerly STI571; Novartis, Basel, Switzerland) selectively impairs the function of the platelet-derived growth factor receptor and c-kit (CD117). Moreover, imatinib inhibits the BCR-ABL fusion protein¹ resulting from the Philadelphia (Ph)

translocation t(9;22)(q34;q11) that plays a crucial role in the pathogenesis of chronic myelogenous leukaemia (CML). Imatinib has been shown in various clinical trials in CML and gastrointestinal stromal tumours to be a well-tolerated oral drug with frequent but mild toxicities.²

We report a 64-year-old woman diagnosed as having chronic phase Ph+ CML in October 1995. After haematological failure of interferon alfa-based therapy oral imatinib 400 mg daily was commenced in August 2001. Complete haematological response was achieved after 2 months of therapy. In January 2002, self-limiting erythematous skin lesions were first observed on the lower legs. During the following months, similar lesions recurred, some of which persisted, in different locations and with variable intensity on both legs and arms. In September 2002, painful erythematous skin lesions appeared on both arms, accompanied by fever. Under the suspicion of streptococcal cellulitis a variety of antibiotic therapies was administered. In November 2002 the clinical situation had deteriorated, with erythematous, swollen, deeply indurated and very painful skin lesions on the right forearm and the upper and lower left leg (Fig. 1A,B). Further symptoms were a fever of 38.5 °C, night sweats, malaise and fatigue. No associated lymphadenopathy or hepatomegaly were detected. The spleen was slightly enlarged (14 × 6 cm). Laboratory tests showed a C-reactive protein level of 141 mg L⁻¹ and a hypochromic, normocytic anaemia with a haemoglobin level of 7.6 g dL⁻¹ requiring transfusion. CML was in the accelerated phase (white blood cell count 9.3 × 10⁹ L⁻¹; 12% bands, 81% polymorphonuclear cells, 2% lymphocytes, 3% monocytes and 2% basophils; platelets 741 × 10⁹ L⁻¹). Bone marrow aspiration cytology revealed a hypercellularity of myelo- and megakaryopoiesis with 2% blasts; cytogenetic evaluation



Figure 1. At presentation, painful and indurated skin lesions were evident on (A) the left leg and (B) the right forearm.