Photosensitive Sweet syndrome: An uncommon entity

Sir,

Sweet syndrome, also called acute febrile neutrophilic dermatosis is an inflammatory disorder characterised by tender, erythematous plaques and nodules, associated with fever and leucocytosis.[1] Although most cases are idiopathic, the disease can also be associated with drugs and malignancies. We describe a relatively rare presentation of Sweet syndrome, limited to photo-exposed areas.

A 39-year-old woman presented with recurrent, erythematous, raised, painful lesions on the face and hands for four years, associated with high-grade fever (>38°C). There was a history of exacerbation of lesions on exposure to sunlight. There were no associated joint pains, Raynaud phenomenon, muscle weakness, fatigue, recurrent infections, gum bleeding, menstrual irregularities or history of drug intake preceding the onset of lesions. The lesions would show remission with systemic corticosteroids but recur as soon as the steroids were tapered or stopped. Cutaneous examination revealed multiple well-defined, erythematous, tender, succulent papules and plaques with pseudo-vesiculation, strictly conforming to photo-exposed sites including face (bridge of nose), helix of ear, V-area of the neck and dorsum of hands [Figure 1] with no lesions on non-photo-exposed sites. Systemic examination was unremarkable. There was no lymphadenopathy.

On investigations, there was elevated erythrocyte sedimentation rate (52 mm/h; normal: 0–20 mm/h). Peripheral neutrophils were marginally raised (7100/mm^3). Urine microscopy and serum complement levels were normal. Antinuclear antibody using indirect immunofluorescence and antistreptolysin-O testing was negative. No atypical cells were seen in blood. Skin biopsy revealed diffuse dense neutrophilic infiltrate in dermis with some oedema in the papillary dermis. The neutrophilic infiltrate extended up to the subcutaneous tissue without any evidence of leukocytoclastic vasculitis [Figure 2]. Direct immunofluorescence study was negative. In view of the clinical, laboratory and histopathological findings, along with exclusive distribution of lesions on photo-exposed areas, a diagnosis of photosensitive Sweet syndrome was made. She was started on oral prednisolone (40 mg/day) which showed significant improvement within two weeks of therapy. Photoprotection using sunscreens and protective clothing was advised. Steroids were tapered, and the patient was maintained on dapsone (100 mg/day) and continues to be in remission. Phototesting was precluded because of the patient's refusal to undergo the same.

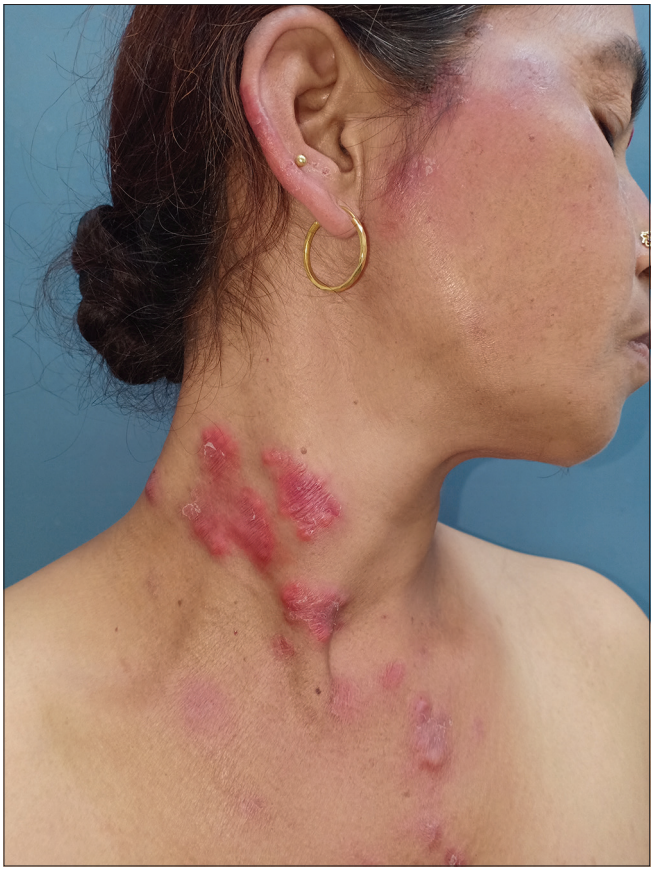

Figure 1a: Multiple well-defined, erythematous, succulent papules and plaques with pseudo-vesiculation, on photo-exposed sites including helix of ear and V-area of the neck

Classical Sweet syndrome predominantly affects women in the age group of 30–60 years, although any age or gender can be affected.[2] Raised acute-phase reactants such as leucocytosis with neutrophilia, as well as elevated erythrocyte sedimentation rate, are often reported. Histopathological

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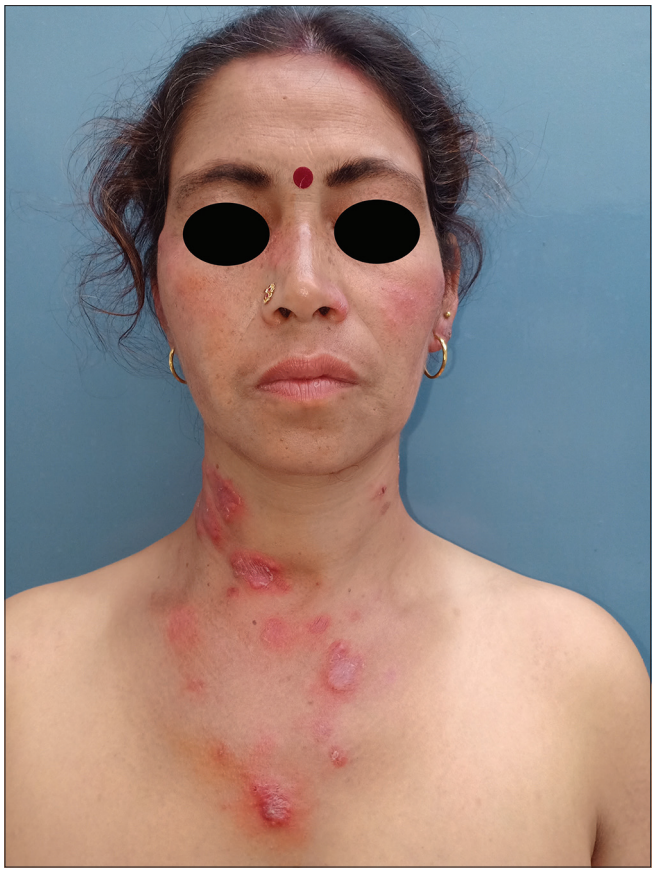

Figure 1b: Erythematous papules and plaques on the cheeks and bridge of nose

Figure 1c: Erythematous papules and plaques on dorsum of the hands

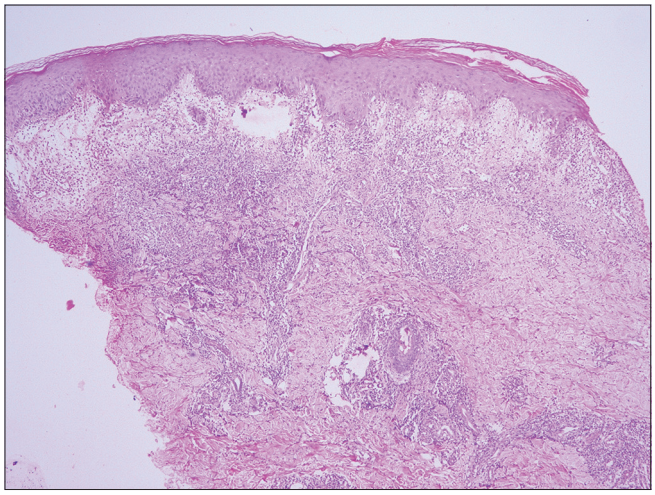

Figure 2a: Mild acanthosis and cellular infiltrate in the dermis (haematoxylin and eosin, ×40)

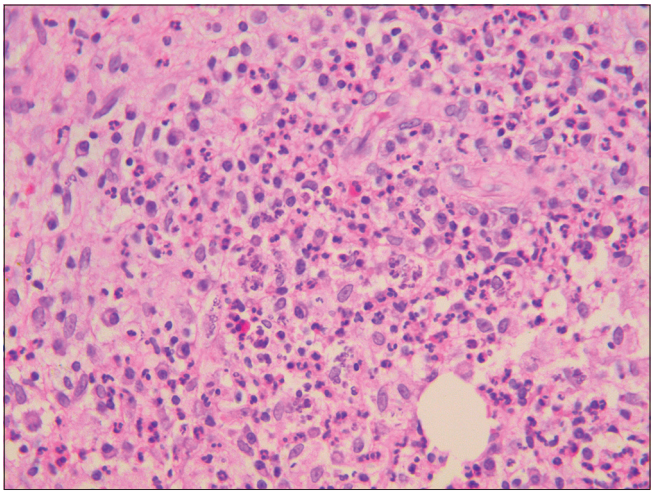

Figure 2b: Diffuse inflammatory infiltrate composed of polymorphonuclear leucocytes and nuclear debris. The neutrophilic infiltrate is present without any evidence of leukocytoclastic vasculitis (haematoxylin and eosin, ×400)

appearance is characteristic with a dense and diffuse neutrophilic infiltrate in the papillary and upper reticular dermis, in the absence of leukocytoclastic vasculitis.[3]

Photo-induction and photo-aggravation of Sweet syndrome have been described previously. Lesions consistent with Sweet syndrome can be induced by both ultraviolet-B (290 nm) and ultraviolet-A (340–400 nm) radiation on phototesting.[4,5] Putative mechanisms involved in photosensitivity include either Koebner phenomenon or ultraviolet B-induced neutrophil recruitment and activation, due to enhanced interleukin-8 and tumour necrosis factor-α production.[1,5] Systemic corticosteroids comprise first-line therapy which were effective in our patient. Other therapies

Table 1: Key distinguishing features between photosensitive Sweet syndrome and bullous lupus erythematous

	Photosensitive Sweet Syndrome	Bullous lupus erythematosus
Clinical presentation	Tender erythematous papules, plaques and nodules on photo-exposed sites. Can show pseudo-vesiculation	Blisters and bullae on photo-exposed (more common) and non-photo-exposed sites. May manifest initially as papules or plaques with erythema or urticaria
Predominantly affected age and gender	Females, aged 30–50 years	Females, aged 30–50 years
Associated systemic features	Mostly idiopathic, some cases can be associated with infections, malignancies and drug intake	May precede, occur simultaneously, or follow systemic lupus erythematosus
Laboratory investigations	• Peripheral leucocytosis with neutrophilia • Raised acute-phase markers such as erythrocyte sedimentation rate and C-reactive protein • Urinalysis is unremarkable • Antinuclear antibody negative • Serum complement levels are normal	• Peripheral leucocytosis with neutrophilia • Raised acute-phase markers such as erythrocyte sedimentation rate and C-reactive protein • Urinalysis may show haematuria, proteinuria and red blood cells casts • Antinuclear antibody positive • Serum complement may be normal or low
Histopathology	Diffuse, dense neutrophilic infiltrate in dermis, often with oedema of the papillary dermis. No evidence of vasculitis or mucin deposition	Subepidermal blisters with neutrophilic infiltrate in papillary dermis forming microabscesses. Mucin deposition in reticular dermis is characteristic
Direct immunofluorescence	Negative for any immunoglobulins or complement deposits	Linear or granular deposits of immunoglobulin G, immunoglobulin A, immunoglobulin M and/or complement C3 at the basement membrane zone

such as dapsone, colchicine and potassium iodide have also found to be effective.[6] Cutaneous lupus erythematosus is an important differential and should be excluded by appropriate investigations. Sweet syndrome like lesions (neutrophilic dermatoses on photo-exposed sites) have been described in association with lupus erythematosus in literature and can be associated with disease flare; though our patient did not have any features of lupus.[7-9] Some key distinguishing features are summarised in [Table 1].

In conclusion, this case illustrates a rare variant of Sweet syndrome, which should be considered in patients presenting with tender nodules and plaques on photo-distributed pattern. Biopsy is characteristic and once diagnosed, systemic corticosteroids induce rapid remission.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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