Sweet syndrome associated with malignant disease

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A 45-year-old woman with relapsed acute myeloid leukemia and who was receiving palliative chemotherapy with monthly decitabine was admitted to the oncology ward with a 2-day history of fever (38.5°C) and painful skin lesions on her hands, arms and upper body. The lesions were red to violaceous coloured tender plaques of variable sizes (Figure 1). We did not observe any enlarged lymph nodes or splenomegaly on physical examination. The patient was anemic (hemoglobin of 7 g/dL [normal 12–16.5 g/dL], total leucocyte count of 11 000/μL [normal 4000–10 000/μL], 70% neutrophils [normal 40%–60%]), with an erythrocyte sedimentation rate of 40 mm per hour (normal 0–10 mm/h) and elevated C-reactive protein (27 mg/dL, normal < 5 mg/L). We prescribed empiric antibiotics as per protocol; these were stopped when her blood and urine culture results were negative. We biopsied a skin lesion on the back of her hand; the histopathology showed intense neutrophilic infiltration in the dermis with no evidence of infection or malignant disease (Appendix 1, available at www.cmaj.ca/lookup/doi/10.1503/cmaj.220827/tab-related-content). We diagnosed Sweet syndrome on the basis of her clinical presentation and histopathology and prescribed oral prednisone (1 mg/kg/d). Her fever subsided after 48 hours, and the skin lesions started improving after 1 week. Prednisone was tapered over 4 weeks and her skin lesions resolved without scarring.

Sweet syndrome is an uncommon autoinflammatory disorder that is characterized by abrupt onset skin lesions with fever and leukocytosis.[1] Diagnosis requires both major criteria (acute onset of painful erythematous plaques or nodules and histopathological evidence of a dense neutrophilic infiltrate) and 2 minor criteria (fever > 38°C; an association with malignant disease, inflammatory disease, pregnancy or a preceding infection; excellent response to systemic glucocorticoids; and abnormal laboratory values at presentation including erythrocyte sedimentation rate > 20 mm/h, elevated C-reactive protein, > 8000 leukocytes, > 70% neutrophils).[2] Differential diagnosis includes drug reaction, infection, leukemia cutis and vasculitis. Acute myeloid leukemia is the most commonly associated hematological malignant disease.[3]

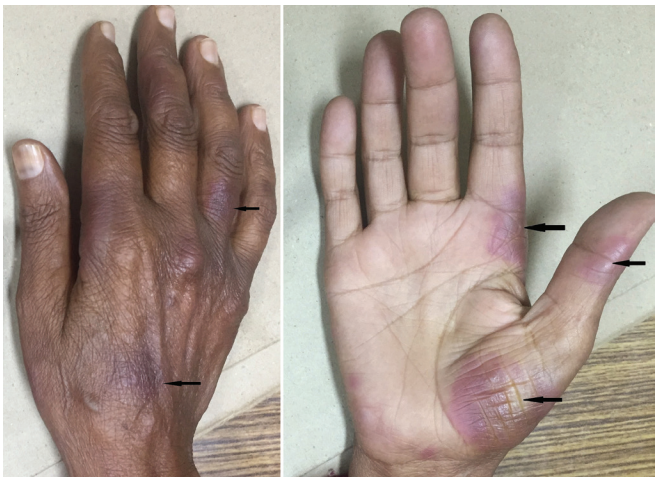

Figure 1: Photographs of the hand of a 45-year-old woman with violaceous plaques of varying sizes involving the back and palm of both hands.

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This article has been peer reviewed.

The authors have obtained patient consent.

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