

Sweet Syndrome Presenting With an Unusual Morphology

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To the Editor:

Sweet syndrome is a neutrophilic dermatosis that typically presents as an acute onset of multiple, painful, sharply demarcated, small (measuring a few centimeters), raised, red plaques that occasionally present with superimposed pustules, vesicles, or bullae on the face, neck, upper chest, back, and extremities. Patients are often febrile and may have mucosal and systemic involvement.¹ Although 71% of cases are idiopathic, others are associated with malignancy; autoimmune disorders; infections; pregnancy; and rarely medications, especially all-*trans*-retinoic acid, granulocyte colony-stimulating factor, vaccines, and antibiotics.^{1,2} We present a case of Sweet syndrome induced by trimethoprim-sulfamethoxazole (TMP-SMX) with an unusual clinical presentation.

A 71-year-old man with a medical history of nonmelanoma skin cancer initiated a course of TMP-SMX for a wound infection of the lower leg following Mohs micrographic surgery. Eight days later, he developed a painful eruption preceded by 1 day of fever, malaise, blurry vision, and myalgia. Trimethoprim-sulfamethoxazole was discontinued. Physical examination revealed ill-defined, discrete and coalescing, 1- to 6-mm edematous erythematous papules studded with pustules involving the scalp, face, neck, back (Figure 1), and extremities. The patient also had conjunctival erythema and an elevated temperature (38.3°C). Laboratory workup revealed an elevated white blood cell count (11,300/ μ L

[reference range, 4500–11,000/ μ L]), blood urea nitrogen level (33 mg/dL [reference range, 7–20 mg/dL]), and creatinine level (2.00 mg/dL [reference range, 0.6–1.2 mg/dL]). Liver function tests were normal. A biopsy demonstrated marked papillary dermal edema with a dense, bandlike, superficial dermal neutrophilic infiltrate (Figure 2). A few neutrophils were present in the epidermis with formation of minute intraepidermal pustules. The patient was diagnosed with Sweet syndrome and treated with intravenous methylprednisolone 60 mg 3 times daily (1.5 mg/kg body weight) tapered over 17 days and triamcinolone acetonide ointment 0.1% twice daily. His fever and leukocytosis resolved within 1 day and the eruption improved within 2 days with residual desquamation that cleared by 3 weeks.

Morphologically, our case resembled acute generalized exanthematous pustulosis (AGEP), which presents with edematous erythema studded with pustules.³ Although fever and leukocytosis are often present in both AGEP and Sweet syndrome, our patient's pain, malaise, and myalgia favored



Figure 1. Ill-defined, discrete and coalescing, 1- to 6-mm edematous erythematous papules studded with pustules on the back.

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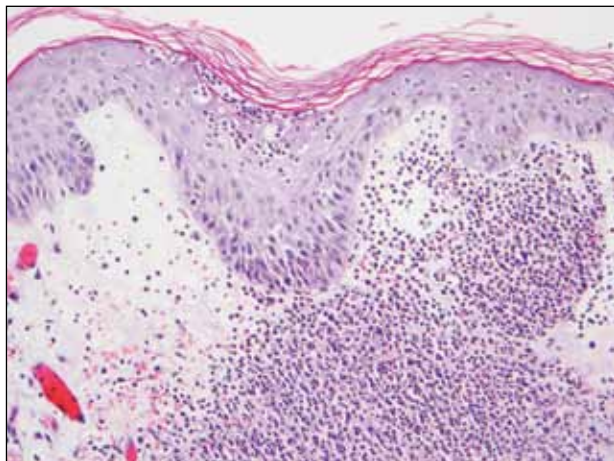


Figure 2. Marked papillary dermal edema, a dense superficial dermal neutrophilic infiltrate, and neutrophil exocytosis with formation of minute intraepidermal pustules (H&E, original magnification $\times 200$).

Sweet syndrome, as did his conjunctivitis, which is unusual in AGEP.^{1,3} Histologically, our case was characteristic for Sweet syndrome, which presents with marked papillary dermal edema and a dense neutrophilic dermal infiltrate with neutrophil exocytosis and spongiform pustules in 21% of cases.¹ Acute generalized exanthematous pustulosis, characterized by spongiform pustules and a perivascular neutrophilic infiltrate, does not exhibit the dense

dermal neutrophilic infiltrate of Sweet syndrome.³ Mecca et al⁴ also reported a case displaying overlapping features of Sweet syndrome and AGEP. The patient presented with photodistributed papules and pinpoint pustules on an erythematous base favoring a diagnosis of AGEP with histologic findings compatible with Sweet syndrome. The authors suggested a clinico-pathologic continuum may exist among drug-related neutrophilic dermatoses.⁴

In conclusion, we present a case of TMP-SMX-induced Sweet syndrome that morphologically resembled AGEP. It is important to recognize that Sweet syndrome may present in this unusual manner, as it may have notable internal involvement, and responds rapidly to systemic steroids, whereas AGEP has minimal systemic involvement and clears spontaneously.

REFERENCES

1. von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol.* 1994;31:535-556.
2. Kluger N, Marque M, Stoebner PE, et al. Possible drug-induced Sweet's syndrome due to trimethoprim-sulfamethoxazole. *Acta Derm Venereol.* 2008;88:637-638.
3. Sidoroff A, Halevy S, Bavinck JN, et al. Acute generalized exanthematous pustulosis (AGEP)—a clinical reaction pattern. *J Cutan Pathol.* 2001;28:113-119.
4. Mecca P, Tobin E, Andrew Carlson J. Photo-distributed neutrophilic drug eruption and adult respiratory distress syndrome associated with antidepressant therapy. *J Cutan Pathol.* 2004;31:189-194.