Sweet Syndrome in Eosinophilic Granulomatosis with Polyangiitis

KATE MURPHY, MD, School of Medicine, Oregon Health & Science University; LYNNE MORRISON, MD, Department of Dermatology, Oregon Health & Science University; JENNA McGOLDRICK, MD, Division of Arthritis & Rheumatic Diseases, Oregon Health & Science University, Portland, Oregon, USA. Address correspondence to Dr. K. Murphy, School of Medicine, Oregon Health & Science University, 3181 SW Sam Jackson Parkway, Portland, Oregon 97239, USA. E-mail: murphka2020@alumni.ohsu.edu. The patient's written informed consent was obtained for the publication of this material. The Institutional Review Board at Oregon Health & Science University determined that this study is not human subject research and approval is not required (IRB ID: STUDY00020195). J Rheumatol 2020;47:1031–2; doi:10.3899/jrheum.190909

Sweet syndrome is a rare dermatologic disorder characterized by sterile neutrophilic infiltrate. Neutrophilic dermatoses have increasingly been recognized in antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis, with the notable exception of eosinophilic granulomatosis with polyangiitis (EGPA)[1].

A 46-year-old man with asthma presented with fever, dyspnea, difficulty ambulating, and a severe rash. Erythematous tender dermal plaques were present across his scalp (Figure 1). Labs were notable for leukocytosis with striking eosinophilia (absolute eosinophil count 7.75), high-titer myeloperoxidase antibody, and positive perinuclear ANCA. Computed tomography demonstrated large subpleural opacities. Nerve conduction studies were consistent with an asymmetric sensorimotor peripheral neuropathy.

His findings were consistent with EGPA; however, his rash was atypical. Skin lesions in EGPA are often subcutaneous nodules with biopsy demonstrating eosinophilic vasculitis, whereas the biopsy in this case showed a dense neutrophilic infiltrate with striking dermal edema consistent with Sweet syndrome (Figure 2)[2]. Sweet syndrome can be paraneoplastic, most commonly in association with acute myeloid leukemia. However, peripheral flow cytometry showed no evidence of hematologic malignancy and he had no symptoms or imaging findings to suggest a solid tumor[3].

Neutrophilic dermatoses have previously been reported in ANCA-associated vasculitis, suggesting a common underlying mechanism of neutrophilic dysregulation. To our knowledge, only 1 case has been reported in association with

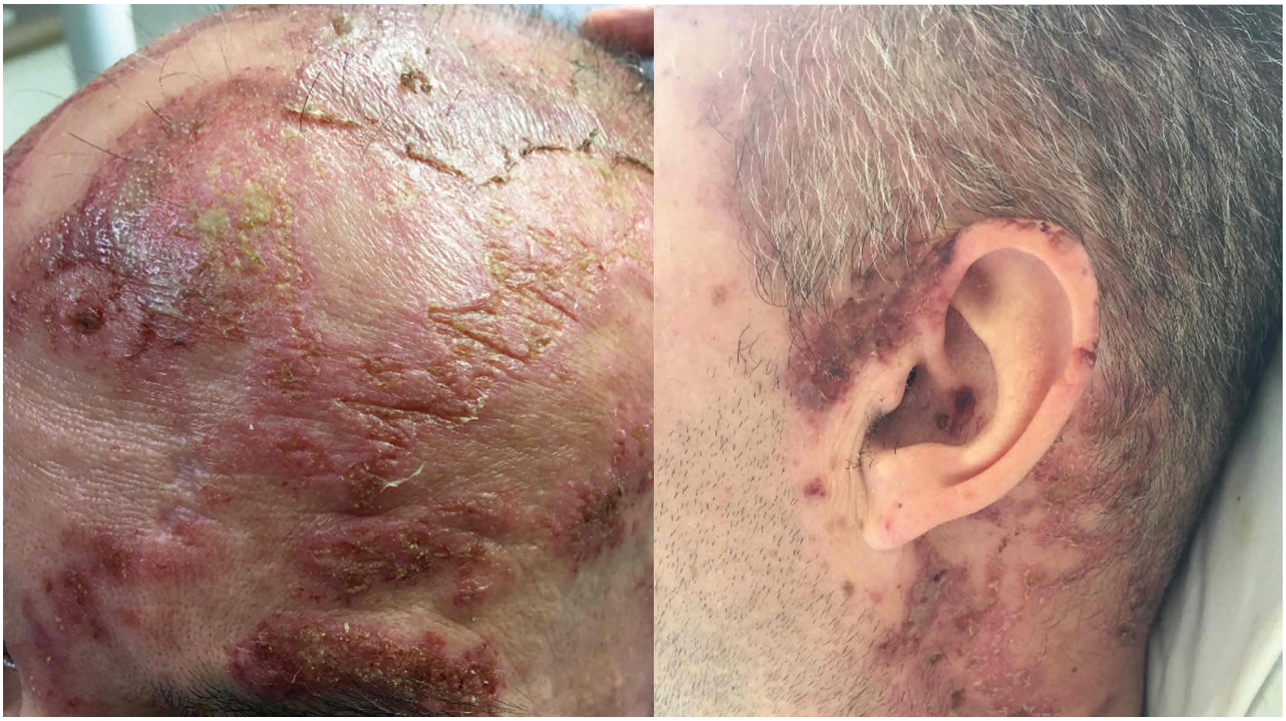

Figure 1. Skin examination on admission demonstrated erythematous, edematous plaques with yellow crusting on the patient's scalp, posterior neck, and ears.

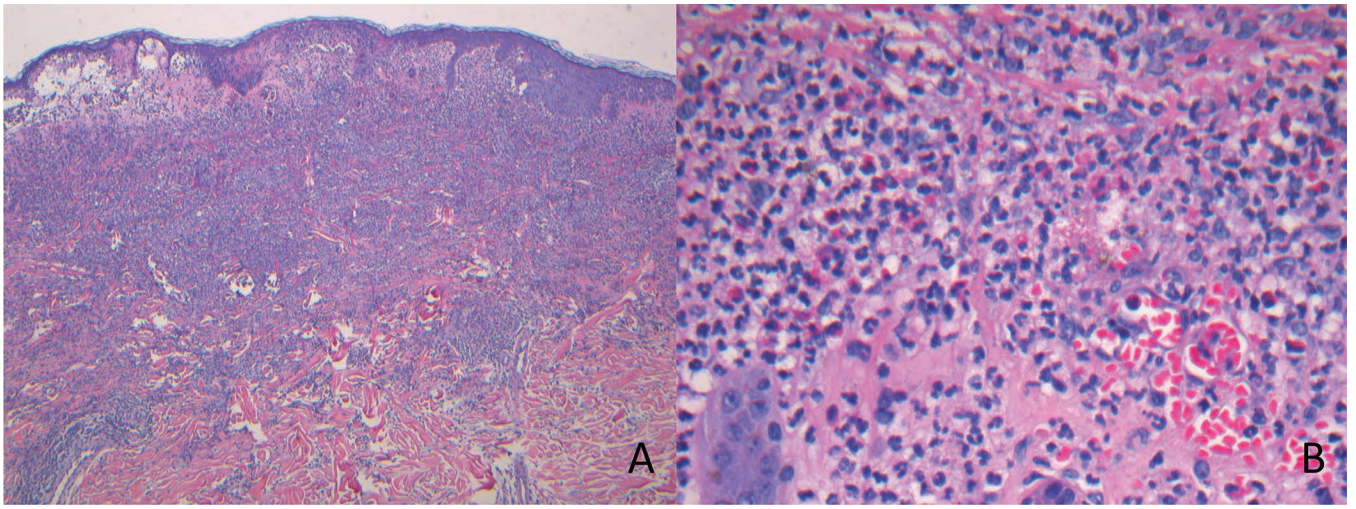

Figure 2. Skin biopsy showed a dense, mixed dermal infiltrate with neutrophils, occasional eosinophils, and lymphocytes, typical of Sweet syndrome. A. Massive papillary dermal edema results in epidermal separation. There is no vasculitis, no granulomatous infiltrate, and the direct immunofluorescence study was negative (50×). B. Higher power view (400×) showing a predominantly neutrophilic infiltrate, with occasional eosinophils and lymphocytes.

EGPA, a female patient with Sweet syndrome and cardiopulmonary symptoms[1].

Rapid response to systemic glucocorticoids is part of the diagnostic criteria for Sweet syndrome, as well as the mainstay of EGPA treatment[4]. Our patient's symptoms improved with intravenous methylprednisolone, followed by longterm oral prednisone and rituximab.

REFERENCES

1. De Boysson H, Martin Silva N, de Moreuil C, Néel A, de Menthon M, Meyer O, et al. Neutrophilic dermatoses in antineutrophil cytoplasmic antibody-associated vasculitis. A French multicenter study of 17 cases and literature review. Medicine 2016;95:e2957.
2. Kawakami T, Soma Y, Kawasaki K, Kawase A, Mizoguchi M. Initial cutaneous manifestations consistent with mononeuropathy multiplex in Churg-Strauss syndrome. Arch Dermatol 2005;141:873-8.
3. Cohen P. Sweet's syndrome – a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34.
4. Groh M, Pagnoux C, Baldini C, Bel E, Bottero P, Cottin V, et al. Eosinophilic granulomatosis with polyangiitis (Churg Strauss) (EGPA) consensus task force recommendations for evaluation and management. Eur J Internal Med 2015;26:545-53.