

# Copresentation of Common Variable Immune Deficiency and Sweet Syndrome

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## PRACTICE POINTS

- Suggested workup for Sweet syndrome includes ruling out connective tissue disorders and malignancies.
- Familial common variable immune deficiency is rare and can first manifest in adulthood.

## To the Editor:

A 38-year-old woman was diagnosed with common variable immune deficiency (CVID) by an immunologist at an outside institution 1 year prior to the current presentation. The diagnosis was based on history of severe recurrent sinopulmonary tract, inner ear, *Clostridium difficile*, urinary tract, and herpes zoster infections of approximately 6 years' duration, as well as persistently low IgG, IgA, and IgM levels of 530 mg/dL (reference range, 690–1400 mg/dL), 29 mg/dL (reference range, 88–410 mg/dL), and 30 mg/dL (reference range, 34–210 mg/dL), respectively, with failure to respond to vaccinations (ie, *Haemophilus influenzae* type B, *Streptococcus pneumoniae*, diphtheria IgG antibody, tetanus antibody). She was started on replacement intravenous immunoglobulin (IVIg) 40 g monthly (400 mg/kg) for CVID. She had a family history of CVID diagnosed in her son and sister.

One year after the CVID diagnosis, she was diagnosed with Sweet syndrome (SS) by a physician at our institution via biopsy of a lesion on the left arm (Figure 1) that showed dense dermal infiltrate of neutrophils with scattered background apoptotic nuclear debris without evidence of vasculitis (Figure 2). Gram stain and microbial

biopsy cultures were negative for mycobacterial, fungal, and bacterial organisms. Cutaneous lesions failed to respond to courses of intravenous antibiotics. Sarcoidosis workup was unremarkable and was pursued to exclude the association with SS. Other negative testing included antinuclear antibody, human immunodeficiency virus, rheumatoid factor, thyroid-stimulating hormone, Ro and La autoantibodies, cytoplasmic antineutrophil cytoplasmic antibody, perinuclear antineutrophil cytoplasmic antibody, antimitochondrial antibody, and urinalysis. Occult



**FIGURE 1.** Sweet syndrome painful erythematous nodule with central ulceration on the forearm.

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The authors report no conflict of interest.

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