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Figure 1. Extensive progressive palpable painful purpura over the patient's trunk and upper extremities.



Figure 2. The confluence of purpura progressed to hemorrhagic bullae.

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A 47-year-old woman visited the emergency department (ED) because of extensive palpable painful purpura over her trunk and 4 extremities for 7 days (Figure 1). The purpura converged and progressed to cellulitis, with skin necrosis on the lower legs (Figures 2 and 3). One month before, she was diagnosed with recurrent Graves' disease and had been receiving propranolol for 30 days and propylthiouracil for 17 days. In the ED, she was afebrile with normal vital signs, exophthalmos, and a palpable goiter. Her initial ED laboratory results revealed a normal CBC count and serum electrolyte levels, a C reactive protein of 33 mg/L, and an erythrocyte sedimentation rate of 24 mm/hour.

For the diagnosis and teaching points, see page 268.

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Figure 3. The hemorrhagic bullae progressed to necrotizing vasculitis. Used with permission of Chih-Ming Huang, MD, Department of Emergency Medicine, National Cheng Kung University Hospital, College of Medicine, National Cheng Kung University, Taiwan, ROC.

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(continued from p. 260)

DIAGNOSIS:

Propylthiouracil-related ANCA-positive vasculitis. The patient was admitted to the hospital for further management of her vasculitis and treated with amoxicillin/clavulanate for her associated cellulitis. She had an excellent clinical response 9 days after she discontinued receiving propylthiouracil and was administered methylprednisolone. Skin biopsy showed leukocytoclastic vasculitis, and a serum perinuclear antineutrophil cytoplasmic antibody (ANCA) result was elevated. She underwent subtotal thyroidectomy for controlling Graves' disease 2 weeks after the cessation of propylthiouracil.

The diagnosis of drug-induced ANCA-associated vasculitis is based on the temporal relationship between clinically evident vasculitis and administration of the offending drugs, and excluding medical conditions that mimic vasculitis and other definable types of vasculitis.¹ ANCA positivity may range from 4.1% to 64% in patients receiving propylthiouracil, with very few developing associated vasculitis.² Prognosis is favorable with timely discontinuation of propylthiouracil and consideration of an alternate hyperthyroidism therapy such as radioiodine therapy or thyroidectomy.³

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