

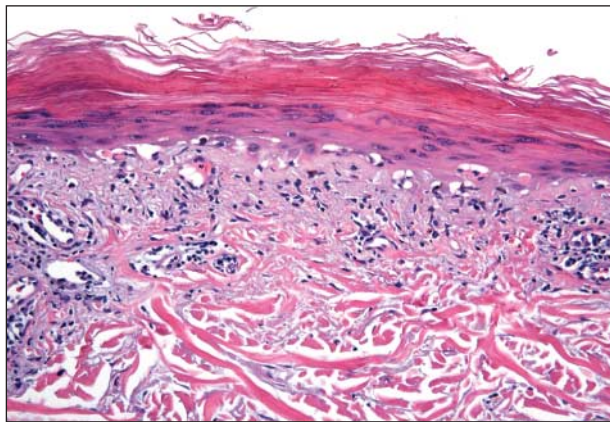


Figure 2. In a 4-mm punch biopsy specimen from the forearm, epidermal atrophy with lymphocytes is evident at the dermoepidermal junction along with vacuolar alteration and dyskeratosis. Perivascular lymphocytic infiltrate and mucinous ground substance in the dermis are also present (hematoxylin-eosin, original magnification $\times 100$).

ment of SCLE.¹ Capecitabine has been strongly associated with cutaneous reactions such as palmoplantar erythrodysesthesia syndrome, but not until recently has it been implicated as a causative agent of drug-induced lupus. In early 2008, Weger et al³ described SCLE induced by 5-fluorouracil and exacerbated by capecitabine. Merlin et al⁴ reported a case of capecitabine-induced discoid LE. Since capecitabine is bioenzymatically converted to 5-fluorouracil, which along with other fluorouracil agents has been reported to cause drug-induced lupus,⁵ the emerging evidence of capecitabine-induced lupus is not surprising. Our report of capecitabine-induced SCLE serves to remind physicians of the importance of understanding the mechanism of action of therapeutic agents when evaluating potential adverse effects.

Neil F. Fernandes, MD
Misha Rosenbach, MD
Rosalie Elenitsas, MD
Joseph M. Kist, MD

Correspondence: Dr Rosenbach, Department of Dermatology, University of Pennsylvania, 3400 Spruce St, Maloney Bldg, 2nd Flr, Philadelphia, PA 19104 (misha.rosenbach@uphs.upenn.edu).

Financial Disclosure: None reported.

1. Borchers AT, Keen CL, Gershwin ME. Drug-induced lupus. *Ann NY Acad Sci.* 2007;1108:166-182.
2. Walko CM, Lindley C. Capecitabine: a review. *Clin Ther.* 2005;27(1):23-44.
3. Weger W, Kränke B, Gerger A, Salmhofer W, Aberer E. Occurrence of subacute cutaneous lupus erythematosus after treatment with fluorouracil and capecitabine. *J Am Acad Dermatol.* 2008;59(2)(suppl 1):S4-S6.
4. Merlin F, Prochilo T, Kildani B, et al. Discoid lupus erythematosus (DLE)-like lesions induced by capecitabine. *Int J Colorectal Dis.* 2008;23(7):715-716.
5. Adachi A, Nagai H, Horikawa T. Anti-SSA/Ro antibody as a risk factor for fluorouracil-induced drug eruptions showing acral erythema and discoid-lupus-erythematosus-like lesions. *Dermatology.* 2007;214(1):85-88.

Autoimmune Progesterone Dermatitis

Report of Cases. Herein, we describe familial autoimmune progesterone dermatitis in 3 sisters and possibly

their mother. To our knowledge, no familial cases have been previously reported.

Case 1. A 41-year-old white woman first experienced intermittent urticarial eruptions at around age 25 years. She described the cyclic eruptions on her extremities and trunk as brief urticarial episodes that would resolve spontaneously then reappear several weeks later. A 3-month course of oral steroids administered at around age 35 years temporarily suppressed the episodes, but once treatment was discontinued, the eruptions returned with greater severity. An intradermal skin test was performed with both progesterone and the progesterone vehicle. The patient had a positive reaction to progesterone only. It was characterized by a wheal and flare initially followed by 2 weeks of indurated erythema. The patient discontinued taking triphasic oral contraceptive pills (OCPs) and began treatment with Seasonique (Duramed Pharmaceuticals, Cincinnati, Ohio) (levonorgestrel/ethinyl estradiol combination tablets and ethinyl estradiol tablets), an extended-cycle OCP. The patient's symptoms markedly improved thereafter.

Case 2. A 37-year-old woman, the younger sister of patient 1, developed cyclic urticarial eruptions throughout her pregnancy that would last several days before spontaneously resolving. She had a normal delivery of a healthy baby, and during the postpartum period, a severe urticarial eruption on the trunk occurred. A biopsy specimen revealed mild inflammatory infiltrate in the superficial dermis composed of abundant eosinophils and vascular ectasia. Direct immunofluorescence did not demonstrate evidence supportive of pemphigoid gestationis. The clinical and histologic findings were most consistent with autoimmune progesterone dermatitis. Results of subsequent intradermal skin tests with progesterone were positive showing a wheal and flare initially, followed by 2 weeks of indurated erythema. After her pregnancy, only occasional mild urticarial eruptions during the luteal phase of her menstrual cycle occurred, and these were controlled with topical steroids.

Case 3. A 44-year-old woman, the older sister of patients 1 and 2, described intensely pruritic urticarial eruptions that began with her first pregnancy and redeveloped during each subsequent pregnancy. With each pregnancy, she had a normal delivery of a healthy baby. She also described several episodes between each pregnancy that were associated with her menstrual cycle. Her intradermal skin test with progesterone also showed positive results with a wheal arising soon after application but no prolonged erythema.

Case 4. A 69-year-old woman, the mother of patients 1, 2, and 3, reported a pruritic skin eruption that occurred at about age 25 years and lasted 2 to 3 years and resolved spontaneously. The patient was unwilling to undergo intradermal skin testing. Besides her 3 daughters, no other family members were tested or identified with similar skin eruptions.

Comment. Autoimmune progesterone dermatitis is a rare mucocutaneous disease characterized by cyclic urticarial or erythema multiformelike eruptions during the luteal phase of the menstrual cycle. Generally, the condition affects women, in their third and fourth decades of life, but

the disease has been described in a man taking exogenous synthetic progesterone,¹ in an adolescent female patient,² and during the postpartum period.³ The diagnosis is confirmed with a positive reaction to intradermal skin testing with progesterone. First-line treatment for the disease is with OCPs, though success with this therapy varies. Gonadotropin-releasing hormone and/or luteinizing hormone-releasing analogues, danazol, tamoxifen, and conjugated estrogens are other alternatives. Definitive treatment is with bilateral oophorectomy.

Sonya V. Chawla, MD
Catherine Quirk, MD
Steven J. Sondheimer, MD
William D. James, MD

Correspondence: Dr James, Department of Dermatology, 2 Maloney Bldg, Hospital of the University of Pennsylvania, 3600 Spruce St, Philadelphia, PA 19104 (william.james@uphs.upenn.edu).

Financial Disclosure: None reported.

1. Fisher DA. Drug-induced progesterone dermatitis. *J Am Acad Dermatol*. 1996; 34(5, pt 1):863-864.
2. Kakarla N, Zurawin R. A case of autoimmune progesterone dermatitis in an adolescent female. *J Pediatr Adolesc Gynecol*. 2006;19(2):125-129.
3. Stephens CJM, Wojnarowska FT, Wilkinson JD. Autoimmune progesterone dermatitis responding to tamoxifen. *Br J Dermatol*. 1989;121(1):135-137.

Atypical Linear IgA Dermatitis Revealing Angioimmunoblastic T-Cell Lymphoma

Angioimmunoblastic T-cell lymphoma (AITL) is frequently accompanied by dermatologic and autoimmune manifestations. We report an unusual case of AITL associated with atypical linear IgA bullous dermatosis (LAD).

Report of a Case. A 47-year-old man with a history of epilepsy treated for many years with sodium valproate, gabapentin, and clonazepam developed generalized pruritus, night sweats, and urticarialike lesions 2 weeks after the introduction of citalopram to his drug regimen. These symptoms were first interpreted to indicate a drug reaction, but they did not resolve after the discontinuation of the new antidepressant and antihistamine therapy. Three months later, he showed low-grade fever, weight loss, and peripheral lymphadenopathy. A pale erythematous eruption on the upper body and large violaceous plaques with large bullae on the hands, feet (**Figure 1**),



Figure 1. Violaceous erythema and bullae of the foot.

elbows, and knees resolved spontaneously. Nikolsky sign was negative. Mucosal examination findings were normal.

The results of laboratory tests showed lymphocytosis (lymphocyte count, 8500/ μ L) with large atypical lymphocytes and abnormal phenotype, hyper-eosinophilia (eosinophil count, 2900/ μ L), macrocytic anemia (hemoglobin level, 0.68 g/dL), elevated β_2 -microglobulin levels (0.0031 mg/L), and polyclonal

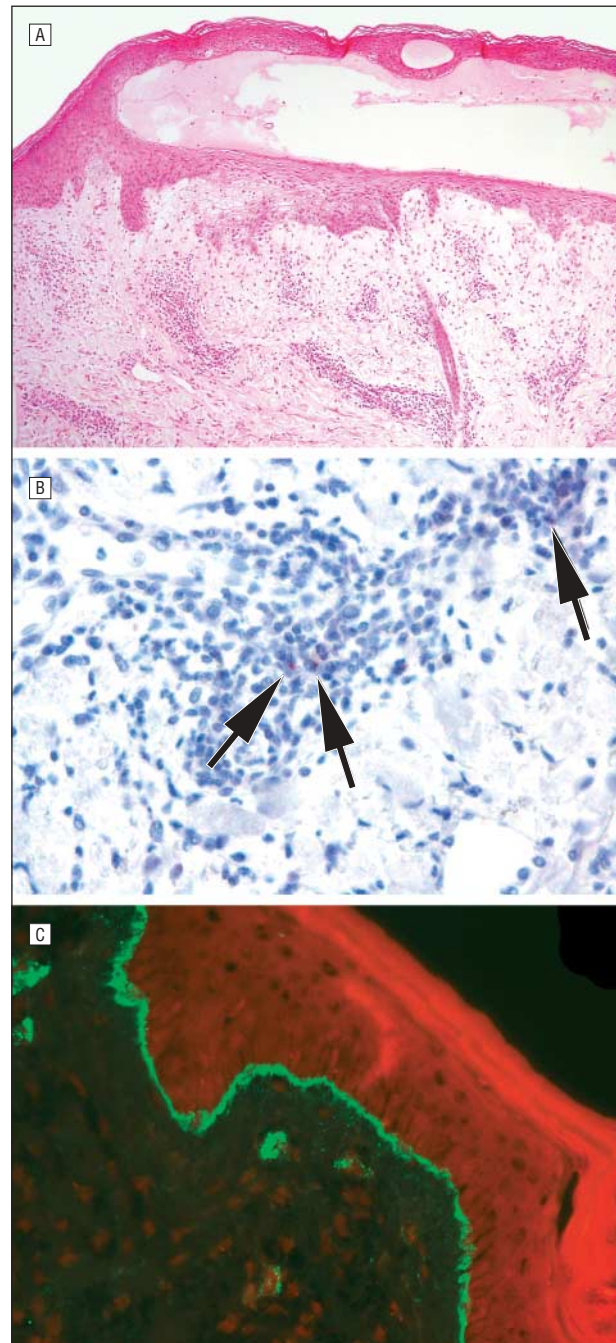


Figure 2. Histopathologic (A), immunohistochemical (B), and direct immunofluorescence (C) images from the present case. A, Subepidermal blister with secondary necrotic changes of the epidermis; in the blister floor, regenerative epidermis overlays a mild dermal lymphocytic perivascular infiltrate (hematoxylin-eosin, original magnification $\times 100$). B, Few scattered CXCL13⁺ lymphocytes are present within the dermal infiltrates (arrows) (original magnification $\times 200$). C, The basement membrane zone shows a linear pattern of staining for C3 (original magnification $\times 200$).