Autoimmune Progesterone Dermatitis Presenting With Purpura

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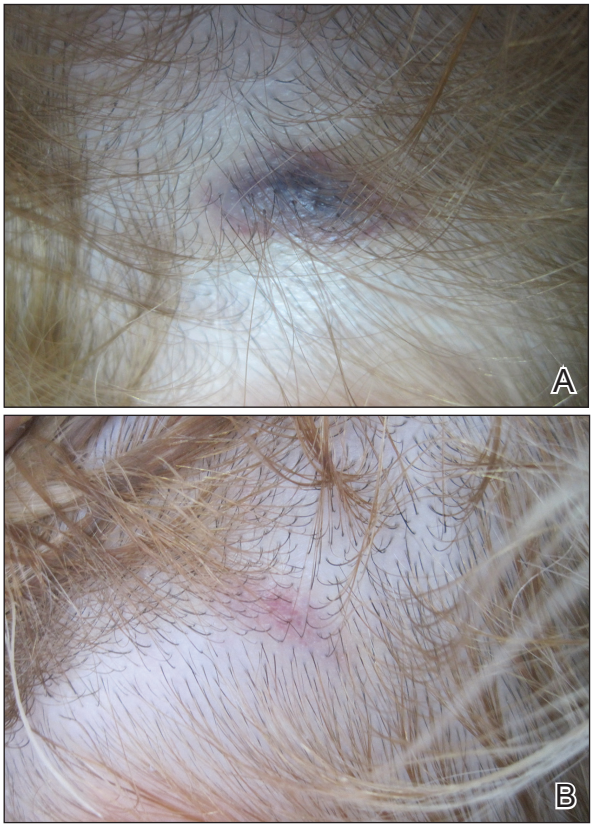
PRACTICE **POINTS**

- Autoimmune progesterone dermatitis is characterized by cyclical skin eruptions, typically occurring in the second half of the menstrual cycle.
- Autoimmune progesterone dermatitis is thought to be an autoimmune reaction to endogenous or exogenous progesterone.
- This condition should be considered in female patients with recurrent skin lesions related to their menstrual cycle.

To the Editor:

A 32-year-old woman presented with a recurrent painful eruption on the scalp of 1 year's duration. The lesion occurred on the left temporal region 1 week prior to menstruation and spontaneously resolved following menses; it recurred every month for 1 year. She had no notable medical history. She had taken oral contraceptive pills for 4 years and stopped 2 years prior to the development of the lesions. Dermatologic examination revealed a purple-colored, violaceous, centrally elevated, painful plaque that measured 2 cm in diameter in the left temporal region of the scalp (Figure, A). Laboratory test results were within reference range. The lesion spontaneously resolved with mild residual erythema at a follow-up visit after menstruation (Figure, B).

Because the eruption occurred and relapsed with the patient's menstrual cycle, we suspected progesterone hypersensitivity. An intradermal skin test was performed on the forearm with 0.05 mL of



Violaceous colored, centrally elevated, purpura-like plaque on the scalp skin (A) that spontaneously resolved after menstruation (B).

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medroxyprogesterone acetate, and saline was used as a negative control. An indurated erythematous nodule occurred on the progesterone-treated side within 6 hours. Based on these findings and the patient's history, she was diagnosed with autoimmune progesterone dermatitis (APD). We recommended her to use gonadotropin-releasing hormone agonists as treatment, but the patient refused. At 6-month follow-up she had recurrent lesions but did not report any concerns.

Autoimmune progesterone dermatitis is a rare condition that is characterized by cyclical skin eruptions, typically occurring in the luteal phase of the menstrual cycle with spontaneous resolution after menses.[1,2] It was first described by Geber[3] in a patient with cyclical urticarial lesions. In 1964, Shelley et al[4] characterized APD in a 27-year-old woman with a pruritic vesicular eruption with cyclical premenstrual exacerbations. Although it is believed there is no genetic predisposition to APD, a case series involving 3 sisters demonstrated that genetic susceptibility might play a role in the etiology.[5] The etiology of APD is still unknown. It is thought to represent an autoimmune reaction to endogenous or exogenous progesterone.[1] Our patient also had used oral contraceptives for 4 years and this exogenous progesterone might have played a role in the sensitization of the patient and the development of this autoimmune reaction.

The clinical features of APD usually begin 3 to 10 days prior to menstruation and end 1 to 2 days after menses. Autoimmune progesterone dermatitis can present in a variety of forms including eczema, erythema multiforme, erythema annulare centrifugum, fixed drug eruption, stomatitis, folliculitis, urticaria, and angioedema.[6] A case of APD presenting with petechiae and purpura has been reported.[7] There are no specific histologic findings for APD.[8] Demonstration of progesterone sensitivity with a progesterone challenge test is the mainstay of diagnosis. Immediate urticaria may occur in some patients, with others experiencing a delayed reaction peaking at 24 to 96 hours.[9] The main criteria of APD include the following: recurrent cyclic lesions related to the menstrual cycle; positive intradermal progesterone skin test; and prevention of lesions by inhibiting ovulation.[1] Two of these criteria were positive in our patient, but we did not use any medications to prevent ovulation at the patient's request.

Current treatment modalities often attempt to inhibit the secretion of endogenous progesterone by suppressing ovulation. Oral contraceptives and conjugated estrogens have limited efficacy rates.[8] Gonadotropin-releasing hormone agonists (ie, buserelin, triptorelin) have been used with success.[1,6] Tamoxifen and danazol are other treatment options. For cases refractory to medical treatments, bilateral oophorectomy can be considered a definitive treatment.[6]

Autoimmune progesterone dermatitis may present in many different clinical forms. It should be considered in the differential diagnosis in patients with recurrent skin lesions related to menstrual cycle both in women of childbearing age and in men taking synthetic progesterone.

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