Autoimmune Progesterone Dermatitis

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Case Presentation

History of Present Illness

A 35-year-old Caucasian female presented with a long-standing history of a treatment resistant, relapsing-remitting urticarial rash diffusely over her entire body. She reported that the lesions had first developed five years prior after discontinuing depo-provera, which she had been taking for the previous 13 years. She noted that the rash would appear (# of days) before her menses and would resolve one to two days after. She also noted that the rash completely resolved during pregnancy and remained absent during lactation, only to recur after discontinuation of breastfeeding. The patient had been treated previously with antihistamines and topical corticosteroids that were ineffective at controlling symptoms, and oral prednisone that was only effective at high doses. She denied any family history of similar conditions. Her medical history was unremarkable and she was not currently on any medications. A review of systems was negative for preceding illness, recent weight loss, or constitutional symptoms.

Work-up

CBC with differential, C3, C4, CH50, and C1Q, TSH, ESR were performed and all laboratory results were within normal limits.

Physical Examination

Physical examination revealed diffuse erythematous wheals involving bilateral flanks and neck and mild swelling of the eyelids.

Discussion

Autoimmune progesterone dermatitis (APD) is a rare disease caused by an autoimmune response to endogenous progesterone.1 APD primarily occurs in women during their reproductive years, commonly around the luteal phase of menstrual cycle when progesterone levels reach their peak. In rare cases, men being treated with synthetic progesterone preparations have also been reported to be afflicted.2 Additionally, reports of familial APD have been described. To date, there have been approximately 60 previously reported cases.

The dermatological features of APD can vary morphologically, but the most commonly described are urticaria, eczema, and erythema multiforme.3 Other findings include angioedema, deep gyrate lesions, papulovesicular lesions, targetoid lesions, or anaphylaxis.4 Often patients with eczematous skin lesions are frequently misdiagnosed with eczematous dermatitis or allergic contact dermatitis, leading to delays in treatment.4

The pathogenesis of APD remains unclear. One theory proposes that after exposure to exogenous progesterone, sensitized presenting cells and T helper 2 cells begin to react with acute and delayed responses consistent with both type I and type IV hypersensitivity reactions.5 Additionally, reports of the presence of anti-progesterone antibodies suggests other pathogenic mechanisms, including type III hypersensitivity reaction to antigen-antibody complexes that are deposited in the skin, which could induce dermatitis as progesterone secretion increases before and after menstruation.4 However, this antibody is not detected in all patients, which only partially explains the pathogenesis.4

Course and Therapy

Based on history and clinical examination, a diagnosis of autoimmune progesterone dermatitis was made. The patient was initially given an intramuscular triamcinolone injection that resulted in minimal improvement of the rash. Based on the patient’s presumed diagnosis, a trial of oral contraceptives was prescribed. The patient began a daily progesteroidal agent (Mini Pill). With this treatment regimen the patient’s condition has been well controlled and limited to one outbreak over the past six months.

References

8. Baptist AP, Baldwin JL. Autoimmune progesterone dermatitis in a patient with endometriosis: case report and review of the literature. Clinical and Molecular Allergy, 2004;2, article no. 10