Diffuse Urticarial Rash in a Pregnant Patient

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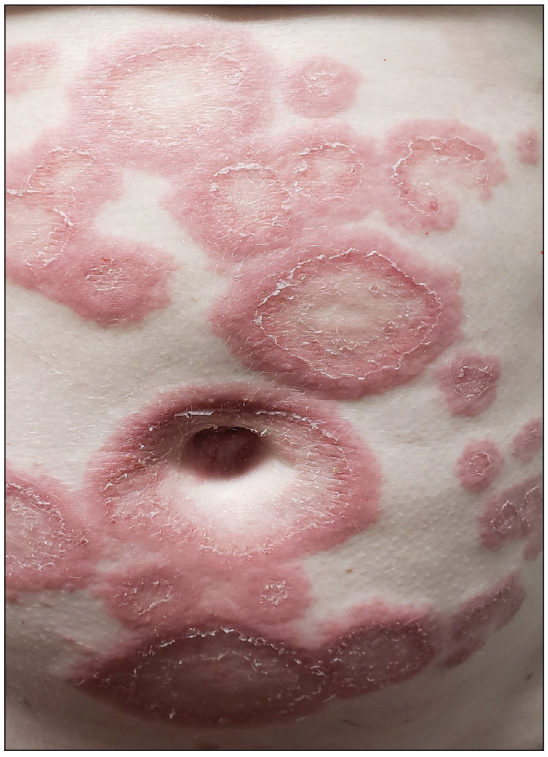

A 29-year-old pregnant woman at 18 weeks and 5 days of gestation presented with a diffuse, pruritic, blistering rash of 5 weeks' duration that started on the forearms and generalized to affect the trunk, legs, palms, and soles. Physical examination showed diffuse urticarial papules and plaques with small tense vesicles with an annular configuration on the abdomen and marked periumbilical involvement.

WHAT'S YOUR **DIAGNOSIS?**

a. atopic eruption of pregnancy
b. impetigo herpetiformis
c. intrahepatic cholestasis of pregnancy
d. pemphigoid gestationis
e. polymorphous eruption of pregnancy

PLEASE TURN TO PAGE E30 FOR THE DIAGNOSIS

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The authors report no conflict of interest.
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THE **DIAGNOSIS:** Pemphigoid Gestationis

A lesional biopsy showed a subepidermal split with eosinophils and neutrophils. Perilesional biopsy for direct immunofluorescence (DIF) showed linear deposition of 3+ C3 along the basement membrane zone. The clinical, histopathologic, and immunofluorescent findings were consistent with pemphigoid gestationis (PG). Prednisone 1 mg/kg daily was initiated. Her condition continued to worsen, and cyclosporine 250 mg daily was added while prednisone was tapered, with remission of disease.

Pemphigoid gestationis is an autoimmune bullous dermatosis that occurs in the second or third trimester of pregnancy, with an incidence of 1 in 50,000 to 60,000 pregnancies.[1] In terms of pathogenesis, aberrant expression of major histocompatibility complex class II molecules on placental tissues causes the loss of immune tolerance of the placenta, which leads to the production of antibodies against the placental protein bullous pemphigoid 180.[2] Bullous pemphigoid 180 also is a hemidesmosomal protein found in the skin of the mother; therefore, the presence of the circulating antibodies leads to separation at the dermoepidermal junction and vesiculation.

Pemphigoid gestationis is characterized by the sudden eruption of intensely pruritic urticarial papules and plaques, classically with periumbilical involvement. Tense vesicles and bullae can develop. Women with PG have an increased risk for development of Graves disease. Histopathology shows subepidermal vesiculation with a predominance of eosinophils. Direct immunofluorescence classically shows linear deposition of C3 along the basement membrane zone. Fetal complications include prematurity and small size for gestational age. Additionally, blisters can be seen in 5% to 10% of neonates due to placental transmission of autoantibodies.[3]

Frequently PG flares shortly postpartum. Pemphigoid gestationis resolves within 6 months postdelivery but frequently reoccurs in subsequent pregnancies. Mild disease can be treated with mid- to high-potency topical corticosteroids. Severe disease is managed with oral corticosteroids, most commonly prednisone. Refractory disease is managed with azathioprine, cyclosporine, intravenous immunoglobulin, or plasmapheresis.

The differential diagnosis of PG includes other pregnancy-associated dermatoses such as atopic eruption of pregnancy, impetigo herpetiformis, intrahepatic cholestasis of pregnancy, and polymorphous eruption of pregnancy. Atopic eruption of pregnancy is the most common dermatosis of pregnancy and is characterized by an eczematous eruption in patients with an atopic history, typically in the first trimester. Blisters are not seen, and DIF is negative. Impetigo herpetiformis, or pustular psoriasis of pregnancy, is a variant of generalized pustular psoriasis that occurs during pregnancy. Diffuse erythematous patches studded with pustules, rather than vesicles, are seen; DIF is negative. Intrahepatic cholestasis of pregnancy presents without primary skin findings and severe pruritus predominantly on the palms and soles, often with secondary excoriations. Polymorphous eruption of pregnancy presents as a polymorphous eruption of urticarial to erythematous papules and plaques commonly originating in striae. In contrast to PG, there is periumbilical sparing, vesiculation is rare, and DIF is negative.

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