Photo Quiz

Pruritic Rash in Pregnancy

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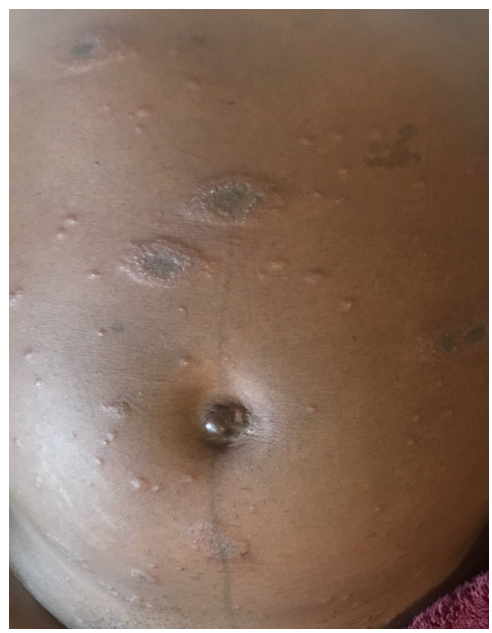

Figure 1.

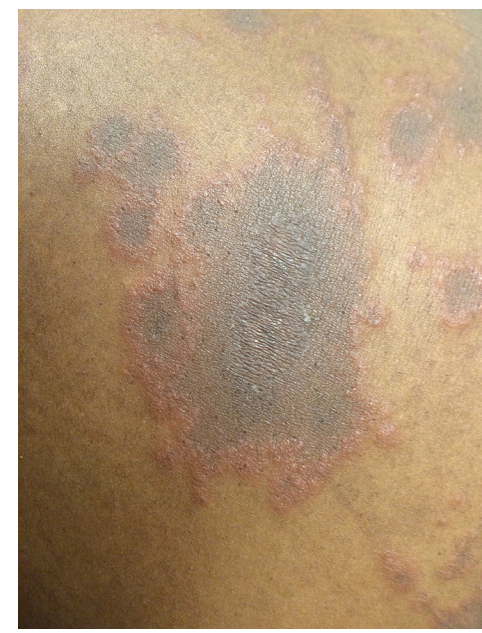

Figure 2.

A 29-year-old woman (gravida 4, para 2) presented at 29 weeks' gestation with the sudden appearance of scattered periumbilical and lower extremity pruritic papules. Despite treatment with topical hydrocortisone valerate and oral diphenhydramine (Benadryl), the rash spread to her entire abdomen and all four extremities. Physical examination revealed ovoid plaques with targetoid features and erythematous nodules *(Figures 1 and 2)*. Her face and mucous membranes were not affected.

The pruritus intensified, and her symptoms did not respond to an increased dose of topical or oral corticosteroids. Her medical and obstetric histories were unremarkable, including no history of similar rashes. She had no new exposures. A skin biopsy revealed prominent linear staining of the epidermal basement membrane for C3 and lesser staining for immunoglobulin G (IgG).

Question

Based on the patient's history, physical examination, and histologic findings, which one of the following is the most likely diagnosis?

- ❑ A. Intrahepatic cholestasis of pregnancy.
- ❑ B. Pemphigoid gestationis.
- ❑ C. Prurigo gestationis.
- ❑ D. Pruritic urticarial papules and plaques of pregnancy (PUPPP).

See the following page for discussion.

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Summary Table

Condition	*Location*	*Characteristics*
Intrahepatic cholestasis of pregnancy	Pruritus usually begins on the palms and soles, and then becomes more widespread	Not a primary dermatologic condition; lesions may develop from intense pruritus and subsequent scratching; may cause linear excoriations and papules
Pemphigoid gestationis	Begins in the periumbilical region with subsequent spread to the remainder of the abdomen and extremities	Lesions vary in appearance, including erythematous papules, urticarial plaques, targetoid lesions, and bullae; may be associated with preterm delivery; biopsy required for diagnosis
Prurigo gestationis	May have typical atopic distribution (face, neck, and flexural regions of extremities) or it may be more widespread	Eczematous lesions; patients may have a history of atopic dermatitis; thought to be a flare-up of underlying atopic dermatitis
Pruritic urticarial papules and plaques of pregnancy	Typically begins on the abdomen within striae, and then spreads to the chest, legs, and arms; the periumbilical region is usually spared	Pruritic papules that coalesce into urticarial plaques

Discussion

The answer is B: pemphigoid gestationis, an uncommon skin disorder that occurs in one out of 50,000 pregnancies.[1] The condition initially presents as intense periumbilical pruritus, usually in the second or third trimester. Skin lesions develop after the onset of pruritus and may include erythematous papules, urticarial plaques, and targetoid lesions. Over the course of several weeks, the rash spreads to the remainder of the abdomen and extremities, and subepidermal bullae may form. The pathogenesis of pemphigoid gestationis is not well understood, but it is thought to be autoimmune and involve circulating IgG targeted at the epithelial basement membrane.[1,2] A definitive diagnosis of pemphigoid gestationis requires a biopsy demonstrating linear staining of the basement membrane for C3 deposition.[3]

Pemphigoid gestationis can affect the health outcome of the fetus, unlike PUPPP and prurigo gestationis. There may be an increased risk of spontaneous miscarriage and fetal demise, but the data are conflicting. One study of 87 pregnancies complicated by pemphigoid gestationis found no increased risk of miscarriage,[2] whereas a small study found that although the overall miscarriage rate was not increased, the rate of late miscarriages and fetal demise was increased.[4] Between 16% and 34% of patients with pemphigoid gestationis give birth prematurely[5]; the risk is higher when it presents in the first or second trimester or if blisters develop.[4]

Pemphigoid gestationis spontaneously regresses within six months of delivery; however, there is a risk of recurrence in subsequent pregnancies, with oral contraceptive use, and during menses.[2] Pemphigoid gestationis has been associated with autoimmune disease, particularly autoimmune thyroid diseases such as Graves disease and Hashimoto disease.[2,3]

Intrahepatic cholestasis of pregnancy is not a primary skin disorder, but skin lesions may appear secondary to the intense pruritus and subsequent scratching induced by cholestasis.[3] Pruritus usually begins on the palms and soles, and then becomes more widespread. Scratching may cause linear excoriations and papules.

Prurigo gestationis is the most common dermatosis of pregnancy and is thought to represent a flare-up of underlying atopic dermatitis. It presents as benign eczematous lesions. The lesions may be limited to the typical atopic distribution (face, neck, and flexural regions of extremities) or more widespread.

PUPPP may be distinguished from pemphigoid gestationis by the sparing of the periumbilical region. The benign, self-limited rash typically begins on the abdomen within striae and then spreads to the chest, legs, and arms. It appears as pruritic papules that coalesce into urticarial plaques.

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Author disclosure: No relevant financial affiliations.

REFERENCES

1. Bedocs PM, Kumar V, Mahon MJ. Pemphigoid gestationis: a rare case and review. *Arch Gynecol Obstet*. 2009;279(2):235-238.
2. Jenkins RE, Hern S, Black MM. Clinical features and management of 87 patients with pemphigoid gestationis. *Clin Exp Dermatol*. 1999;24(4):255-259.
3. Bergman H, Melamed N, Koren G. Pruritus in pregnancy: treatment of dermatoses unique to pregnancy. *Can Fam Physician*. 2013;59(12):1290-1294.
4. Chi CC, Wang SH, Charles-Holmes R, et al. Pemphigoid gestationis: early onset and blister formation are associated with adverse pregnancy outcomes. *Br J Dermatol*. 2009;160(6):1222-1228.
5. Huilaja L, Mäkikallio K, Tasanen K. Gestational pemphigoid. *Orphanet J Rare Dis*. 2014;9:136. ■