A Case of Pustular Psoriasis of Pregnancy With Positive Maternal-Fetal Outcomes

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PRACTICE **POINTS**

- Given its association with maternal and fetal morbidity/mortality, it is important for physicians to have a high suspicion for pustular psoriasis of pregnancy (PPP) in pregnant women with widespread cutaneous eruptions.
- Oral corticosteroids and close involvement of obstetric care is the mainstay of treatment for PPP.

Pustular psoriasis of pregnancy (PPP), also known as impetigo herpetiformis, is a rare condition that affects women in the third trimester of pregnancy through the postpartum period. The relative infrequency of PPP presents both clinical and pathologic challenges in the diagnosis and management of this condition. We report a case of a woman who presented at 32 weeks' gestation with a generalized rash demonstrating clinicopathologic features consistent with PPP. Based on prior reports of adverse maternal-fetal outcomes in PPP, coordinated efforts from our patient's dermatologic and obstetric teams ensured positive outcomes for the patient and the neonate.

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Pustular psoriasis of pregnancy (PPP), also known as impetigo herpetiformis, is a relatively rare cutaneous disorder of pregnancy wherein lesions typically appear in the third trimester and resolve after delivery; however, lesions may persist through the postpartum period. Pustular psoriasis of pregnancy may be considered a fifth dermatosis of pregnancy, alongside the classic dermatoses of atopic eruption of pregnancy, intrahepatic cholestasis of pregnancy, pemphigoid gestationis, and pruritic urticarial papules and plaques of pregnancy.[1]

As PPP is a rare disease, its effects on maternal-fetal health outcomes and management remain to be elucidated. Though maternal mortality is rare in PPP, it is a unique dermatosis of pregnancy because it may be associated with severe systemic maternal symptoms.[2] Fetal morbidity and mortality are less predictable in PPP, with reported cases of stillbirth, fetal anomalies, and neonatal death thought to be due largely to placental insufficiency, even with control of symptoms.[1,3] Given the risk of serious harm to the fetus, reporting of cases and discussion of PPP management is critical.

Case Report

An otherwise healthy 29-year-old G2P1 woman at 32 weeks' gestation presented to our emergency department with a 1-week history of a pruritic, burning rash that started on the thighs then spread diffusely. She denied any similar rash in her prior pregnancy. She was not currently taking any medications except for prenatal vitamins and denied any systemic symptoms. The patient's obstetrician initiated treatment with methylprednisolone 50 mg once daily for the rash 3 days prior to the current presentation, which had not seemed to help. On physical examination, edematous pink plaques studded with 1- to 2-mm collarettes of scaling and sparse 1-mm pustules involving the arms, chest, abdomen, back, groin, buttocks, and legs were noted. The plaques on the back and inner thighs had a peripheral rim of desquamative scaling. There were pink macules on the palms, and superficial desquamation was noted on the lips. The oral mucosa was otherwise spared (Figure 1).

Biopsy specimens from the left arm revealed discrete subcorneal pustules with mild acanthosis of the epidermis with spongiosis (Figure 2). The papillary dermis showed a sparse infiltrate of neutrophils with many

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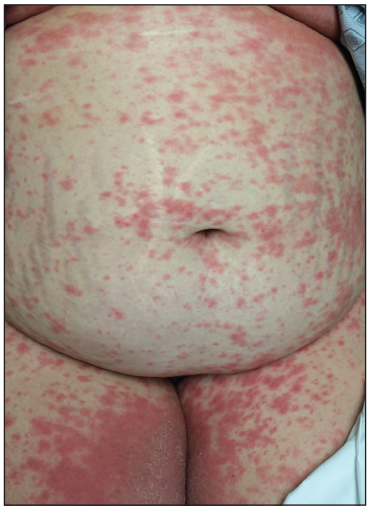

FIGURE 1. Pink plaques with rare pustules coalescing on the abdomen in pustular psoriasis of pregnancy.

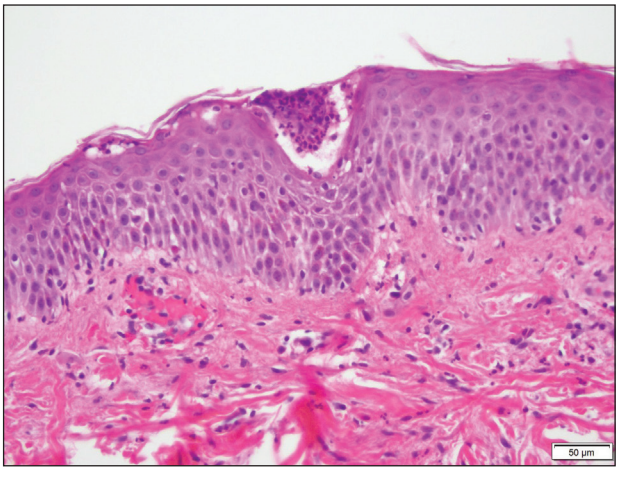

FIGURE 2. A biopsy of a lesion on the left arm in a patient with pustular psoriasis of pregnancy revealed discrete subcorneal pustules. The papillary dermis showed mild edema and a sparse infiltrate of neutrophils and eosinophils (H&E, original magnification ×100).

marginated neutrophils within vessels. Direct immunofluorescence was negative for human IgG, IgA, IgM, complement component 3, and fibrinogen. Laboratory workup revealed leukocytosis of 21.5×10^9/L (reference range, 4.5–11.0×10^9/L) with neutrophilic predominance of 73.6% (reference range, 56%), an elevated erythrocyte sedimentation rate (ESR) of 40 mm/h (reference range, 0–20 mm/h), and a mild hypocalcemia of 8.6 mg/dL (reference range, 8.2–10.2 mg/dL). The patient was started on methylprednisone 40 mg once daily with a plan to taper the dose by 8 mg every 5 days.

At 35 weeks' gestation, the patient continued to report pruritus and burning in the areas where the rash had developed. The morphology of the rash had changed considerably, as she now had prominent, annular, pink plaques with central clearing, trailing scaling, and a border of subtle pustules on the legs. There also were rings of desquamative scaling on the palms. During follow-up at 37 weeks' gestation, the back, chest, and abdomen were improved from the initial presentation, and annular pink plaques with central clearing were noted on the legs (Figure 3). Given the clinical and histopathologic findings, a diagnosis of PPP was made. It was recommended that she undergo increased fetal surveillance with close obstetric follow-up. Weekly office visits with obstetrics and twice-weekly Doppler ultrasounds and fetal nonstress tests were deemed appropriate management. The patient was scheduled for induction at 39 weeks' gestation given the risk for potential harm to the fetus. She was maintained on low-dose methylprednisolone 4 mg once daily for the duration of the pregnancy. The patient continued to have gradual improvement of the rash at the low treatment dose.

Following induction at 39 weeks' gestation, the patient vaginally delivered a healthy, 6-lb male neonate at an outside hospital. She reported that the burning sensation improved within hours of delivery, and systemic steroids were stopped after delivery. At a follow-up visit 3 weeks postpartum, considerable improvement of the rash was noted with no evidence of pustules. Fading pink patches with a superficial scaling were noted on the back, chest, abdomen, arms, legs (Figure 4), and fingertips. The patient was counseled that PPP could recur in subsequent pregnancies and that she should be aware of the potential risks to the fetus.

Comment

In our patient, the diagnosis of PPP was supported by the presence of erythematous, coalescent plaques with small pustules at the margins and central erosions as well as the histologic findings of subcorneal pustules with mild acanthosis of the epidermis with spongiosis and a sparse neutrophilic infiltrate into the dermis. Laboratory studies showing leukocytosis and an elevated ESR, which are often seen in PPP, also were noted.

The typical presentation of PPP is characterized by lesions that initially develop in skin folds with centrifugal spread.[3] The lesions usually begin as erythematous plaques with a pustular ring with a central erosion. The face, palms, and soles of the feet typically are spared with occasional involvement of oral and esophageal mucosae. Biopsy findings typically include spongiform pustules with neutrophil invasion into the epidermis. Typical laboratory findings include electrolyte derangements with elevated ESR and leukocytosis.[1]

Diagnosis of PPP is critical given the potential for associated fetal morbidity and mortality.[4] Anticipatory guidance for the patient also is necessary, as PPP can recur with subsequent pregnancies or even use of oral contraceptive pills (OCPs). Notably, a patient with recurrences of PPP with each of 9 pregnancies also experienced a recurrence

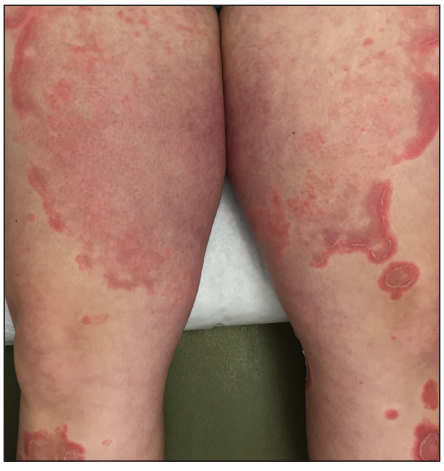

FIGURE 3. During follow-up at 37 weeks' gestation, improvement was noted in a patient with pustular psoriasis of pregnancy following a 40-mg methylprednisolone taper. Annular pink plaques with central clearing were noted on the legs.

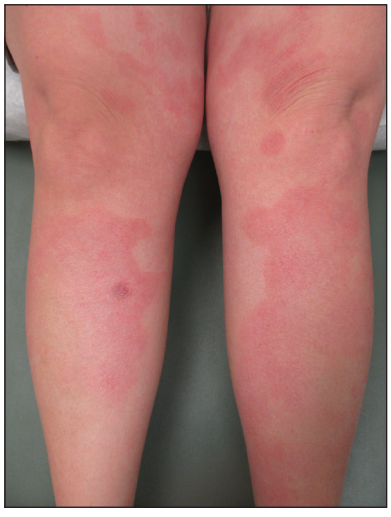

FIGURE 4. At a follow-up visit 3 weeks postpartum, considerable improvement of the rash from pustular psoriasis of pregnancy was noted on the legs.

when taking a combination estrogen/progesterone OCP, but not with an estrogen-only diethylstilbestrol OCP.[5] Although the pathophysiology is not entirely understood, the development of PPP is thought to be related to the hormonal changes that occur in the third trimester, most notably due to elevated progesterone levels.[2] The presence of progesterone in OCPs and recurrences associated with their use supports this altered hormonal state, contributing to the underlying pathophysiology of PPP.

Pustular psoriasis of pregnancy can occur in women without any personal or family history of psoriasis, and as such, it is unclear whether PPP is a separate entity or a hormonally induced variation of generalized pustular psoriasis. Recent evidence included reports of women with PPP who had a mutation in the IL-36 receptor antagonist, leading to a relative abundance of IL-36 inflammatory cytokines.[6] As the cytokine profile during pregnancy is physiologically altered to favor a T helper 2–cell state, the altered expression of cytokines is believed to trigger an inflammatory response conducive to the development of PPP.[2] This mutation in the IL-36 receptor antagonist also is found in individuals with generalized pustular psoriasis, suggesting an overlap of disease etiology related to these cytokine interactions.[2,7,8]

The mainstay of treatment for PPP is oral corticosteroids. Cases of PPP that are unresponsive to systemic steroids have been documented, requiring treatment with cyclosporine.[9] Antitumor necrosis factors also have been used safely during pregnancy.[10] Narrowband UVB phototherapy also has been proposed as a treatment alternative for patients who do not respond to oral corticosteroids.[11]

Conclusion

Pustular psoriasis of pregnancy is a rare dermatosis of pregnancy that, unlike most other common dermatoses of pregnancy, is associated with adverse fetal outcomes. Diagnosis and management of PPP are critical to ensure the best care and outcomes for the patient and fetus and for a successful delivery of a healthy neonate. Our patient with PPP presented with involvement of the body, palms, and oral mucosa in the absence of systemic symptoms. Close follow-up and comanagement with the patient's obstetrician ensured safe outcomes for the patient and the neonate.

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