BRIEF ARTICLES

Widespread Erythematous Plaques in a Multigravid Female

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ABSTRACT

When dermatological eruptions present diagnostic uncertainty, there is cause for concern, especially when treating the mother of an unborn child. While cutaneous outbreaks in pregnancy can be benign, suspicious disseminating rashes should warrant further investigation. We present a case of generalized pustular psoriasis of pregnancy that manifested in an atypical fashion and could have been overlooked without the utilization of biopsy and histopathological analysis. Due to reports of unfavorable maternal-fetal outcomes, prompt evaluation and treatment is critical.

INTRODUCTION

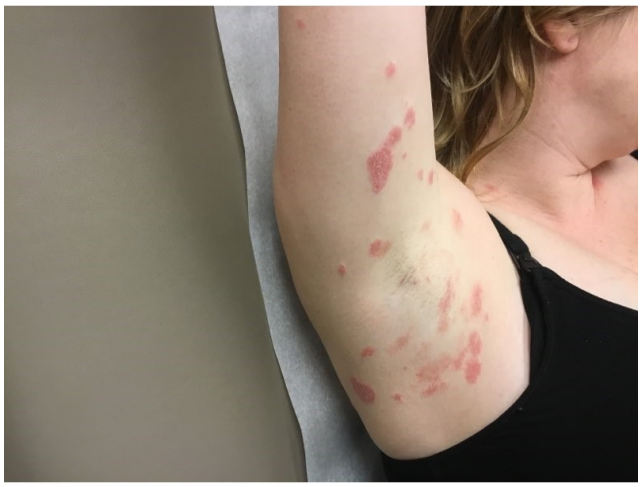
Dermatologic conditions in pregnancy can be attributed to physiological adaptations, changes in pre-existing skin diseases, or development of new dermatologic conditions specific to pregnancy.[1,2] Although controversy exists, there is an ill-defined group of pruritic conditions that are considered pregnancy-specific skin dermatoses, which include pemphigoid gestationis, polymorphic eruption of pregnancy (PEP), intrahepatic cholestasis of pregnancy, atopic eruption of pregnancy, and generalized pustular psoriasis of pregnancy (GPPP).[3] Excluding intrahepatic cholestasis of pregnancy, which presents with secondary skin lesions like excoriations, prurigo nodularis, and occasionally jaundice, these conditions can often manifest as widespread erythematous plaques and papules.

When these conditions occur simultaneously with chronic dermatologic conditions, such as herpes labialis, correct diagnosis can be elusive. Because pemphigoid gesationis and GPPP carry potential risk to the fetus, cutaneous eruptions in pregnancy should prompt immediate consultation and evaluation by a dermatologist.[4] Prognosis is typically favorable with early recognition and management. Delayed or misdiagnosis may result in placental insufficiency, preterm delivery, growth restrictions, miscarriage, and stillbirth.[4,5,6]

CASE REPORT

A 35-year-old G2P1 Caucasian female at 36 weeks gestation with chronic hypertension and history of herpes labialis presented with a pruritic eruption for two weeks. The eruption began on her abdomen involving the striae and subsequently generalized. She reported dysphagia but no other systemic complaints. Physical exam showed erythematous plaques and papules without

Figure 1: Erythematous plaques present on the axilla.



bullae, pustules or scales distributed on upper and lower extremities, truncal and intertriginous skin (Figure 1), and the left labia majora. The rash included palmoplantar involvement. A small superficial erosion was also noted on the soft palate.

The clinical differential diagnosis included erythema multiforme, PEP, herpes gestationis, and psoriasis. The patient completed a prednisone taper and was started on topical steroids with no improvement over two weeks.

Two punch biopsies were obtained, and histopathology revealed psoriasiform epidermal hyperplasia with mounds of parakeratosis containing neutrophils with slight spongiosis and no increase of eosinophils (Figure 2). The other showed spongiform pustule formation with parakeratosis and minimal spongiosis with dilated tortuous blood vessels in dermal papillae (Figure 3).

The patient was monitored closely in conjunction with her obstetrician. Significant laboratory studies revealed mild hypocalcemia of 8.6 mg/dL (reference range 8.7-10.2 mg/dL), leukocytosis of 13.3 x10E3/uL (reference range 3.4-10.8 x 10E3/uL) and a neutrophilia of 9.2 x 10E3/uL (reference range 1.4-7.0 x 10E3/uL).

Considering the new evidence, a diagnosis of GPPP was made. The patient was treated with topical steroids and underwent cesarean section due to hypertension at 38 weeks gestation. A healthy baby girl was delivered. Over a two-month period, the treatment regimen consisted of oral prednisone 20mg, mometasone 0.1% topical cream, clobetasol 0.05% topical cream, and triamcinolone acetonide 0.1% topical cream. The pruritic eruption resolved eight weeks post-partum.

Figure 2: Psoriasiform epidermal hyperplasia with mounds of parakeratosis containing neutrophils with slight spongiosis and no increase of eosinophils.

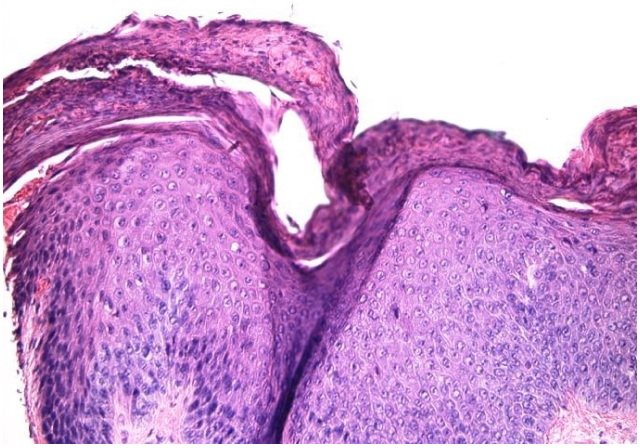
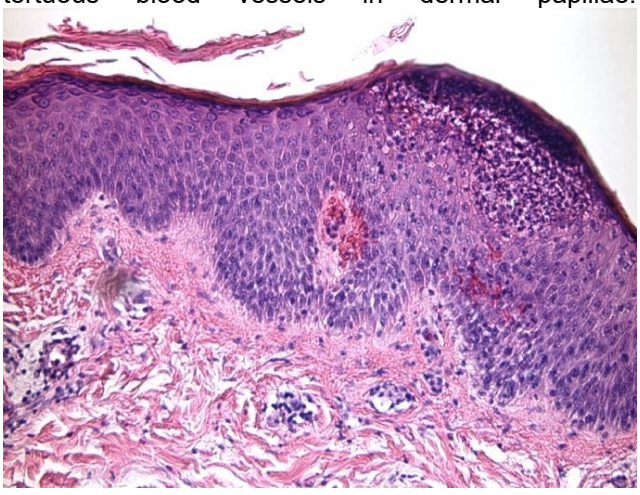


Figure 3: Spongiform pustule formation with parakeratosis and minimal spongiosis with dilated tortuous blood vessels in dermal papillae.



DISCUSSION

Generalized pustular psoriasis of pregnancy, formerly known as impetigo herpetiformis, is considered a rare variant of generalized pustular psoriasis that can occur anytime during pregnancy, but often presents in the third trimester. Although the etiology is unclear, elevated serum progesterone, hypocalcemia, stress and infections have been suggested as potential triggers.[7] An elevation in cytokines due to mutations in IL-36 receptor antagonist has also been suggested to contribute to the inflammatory response seen in GPPP.[8]

The rash of GPPP is characterized by sterile pustules at the periphery of erythematous plaques initially arising from intertriginous areas of the body with subsequent involvement of the trunk and limbs.[1] Our patient presented in a similar fashion in terms of progression, but interestingly never revealed pustules throughout the course of disease.

In addition to oral herpetic lesions, she displayed an acral eruption involving the palms and soles, which are usually spared in GPPP. Initially, erythema multiforme was considered given the history, distribution, and contemporaneous active herpes labialis. Polymorphic eruption of pregnancy was also possible given it is much more common and characteristically involves the striae. Despite the lack of bullae, pemphigoid gestationis is a diagnostic consideration that must be ruled out, as it too can present as urticarial plaques or papules surrounding the umbilicus and increases the risk of maternal-fetal harm.[4]

Unlike the other pregnancy-specific dermatoses, GPPP is often accompanied by systemic signs and symptoms of fever, malaise, vomiting, diarrhea, convulsions, delirium and elevated markers of inflammation.[7] Our patient did not exhibit any of these, except mild dysphagia.

When presenting with non-specific erythematous papules and plaques without prominent systemic symptoms, GPPP could easily be misdiagnosed and progress untreated. Clinical history and morphology can help differentiate GPPP from similar dermatoses, but ultimately punch biopsy with histopathologic analysis is recommended for definitive diagnosis.[4,9] The characteristic spongiform pustules containing neutrophils along with psoriasiform epidermal hyperplasia and parakeratosis were seen in our patient despite the absence of clinical pustules.

Current literature suggests early treatment with systemic corticosteroids or low dose cyclosporine for immunosuppression.[7] Infliximab, a TNF-α inhibitor, has also been used in a minority of cases as a first-line agent for severe forms.[7] Our patient gradually improved over a two-month period. Early in the course, she completed a prednisone taper but was subsequently managed with topical mometasone and clobetasol creams.

Numerous publications note rapid resolution of disease following delivery, but some reports suggest GPPP may recur in subsequent pregnancies.[10,11,12] In some instances it has been reported that the pustular form evolved into classic psoriatic lesions.[10] Differing opinions exist as to whether personal or family history of psoriasis confers increased risk of GPPP.[7,10,11,12] Our patient had no personal or family history and reported complete resolution eight weeks postpartum with only faint post-inflammatory erythema.

CONCLUSION

Generalized pustular psoriasis of pregnancy is an uncommon dermatosis of pregnancy often associated with obscure clinical and laboratory findings. Keeping GPPP on the differential of pruritic eruptions presenting with erythematous papules and plaques is important, even when no pustules are appreciable. Close management and follow up is recommended as fetal harm could be imminent.

Our case sheds light on GPPP, illustrating a rare condition in attenuated form. Utilizing biopsy and histopathological analysis is critical in the diagnostic pursuit, as we identified GPPP despite the absence of pustules and averted harm to mother and child.

Conflict of Interest Disclosures: None.

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