Pruritic urticarial papules and plaques of pregnancy: An unusual case

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Pruritic urticarial papules and plaques of pregnancy is considered the most common pruritic skin condition seen in pregnancy, and its classic presentation and treatment options have been well described. However, the exact etiologic factor of this condition remains obscure. This article presents an unusual case to demonstrate a variation in presentation of this disease as well as a unique course of treatment. The unusual progression of this case may render new insight regarding the etiologic factor of this disease.

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A 29-year-old primigravida in her 38th week of gestation presented to her doctor and complained of an intensely pruritic rash on her abdomen. Her prenatal course had been uncomplicated to this point. Her past medical history included mitral valve prolapse and an appendectomy at age 23 years. She had gained 27 pounds during the pregnancy.

The rash was described as erythematous papules that had begun below her umbilicus within an abdominal stria. For 3 days the rash remained localized, but then began to spread centrifugally outward on the abdomen. The papules coalesced to form plaques with halos of erythema. The intensity of the pruritus escalated. The papules and plaques spread in less than 1 week to cover the entire body diffusely, including the palms and soles, sparing only the face and scalp.

Based on this presentation, a tentative diagnosis of pruritic urticarial papules and plaques of pregnancy (PUPPP) was rendered. Initial treatment included low potency topical steroid cream, oral diphenhydramine, cool soaks, and emollients. Symptoms continued to increase on this regimen, and the patient had secondary excoriations. The pruritus was described as "incapacitating."

A dermatologist was consulted on treatment options. A skin biopsy was performed and the patient was placed on a category one topical steroid ointment. Skin biopsy was consistent with the diagnosis of PUPPP. No symptom relief occurred.

Because of the symptom severity and lack of response to treatment, labor was induced in this term patient. During labor the patient was placed on IV solutedrol and diphenhydramine with some relief of symptoms. Vaginal delivery of a 4500-g girl occurred without complication.

After delivery, the patient was placed on 40 mg of oral prednisone daily with tapering doses over 5 days, during which time the symptoms waned but never fully resolved. One week postpartum, the pruritus had significantly worsened and the patient was placed on 60 mg of oral prednisone daily for 5 days. Symptoms again improved on this regimen but still without complete resolution.

Ten days after completing the steroid, "burst" symptoms including pruritus and rash once again worsened, and the patient was placed on a 3-week tapering dose of oral prednisone starting at 60 mg daily. The pruritus and rash completely resolved on this regimen without reoccurrence.

Six weeks postpartum, the symptoms completely resolved.

Discussion

Pruritic urticarial papules and plaques of pregnancy, the most common pruritic skin condition seen in pregnancy,1 is predominately a disorder of primigravid women that occurs within the last 5 weeks of pregnancy.2 Clinically, the lesions of PUPPP involve red urticarial papules and plaques, which initially appear on the trunk and proximal regions of the extremities. The lesions generally develop in the abdominal striae and spread centrifugally with sparing of the periumbilical region. The papules and plaques are described as intensely pruritic and are occasionally accompanied by small vesicles. Sparing of the palms, soles, and face are typical of the rash's presentation.3

Pruritic urticarial papules and plaques of pregnancy produces no systemic symptoms and does not appear to be associated with adverse perinatal outcomes.4 Serum chemistries, liver enzymes, sedimentation rates, and white blood cell counts are all clinically normal in patients with PUPPP. Histologic examination reveals a nonspecific, perivascular mononuclear infiltrate with a variable number of eosinophils. Epidermal changes include focal spongiosis and parakeratosis with mild acanthosis.5

Initially, our patient presented a diagnostic dilemma due to the unusual presentation of her rash. Involvement of the palms and soles is unusual for PUPPP and is usually more typ-
ical of pemphigoid gestationis. Pemphigoid gestationis is a polymorphous, vesiculobullous eruption associated with intense pruritus that occurs in primagravidas in the second and third trimesters. Systemic symptoms and periumbilical lesions are common. Laboratory tests indicate leukocytosis with eosinophilia; histologic examination reveals subepidermal bullae.

Our patient had periumbilical sparing with involvement of her palms and soles. Skin biopsy revealed lack of bullae and a perivascular mononuclear infiltrate, and laboratory data were unremarkable. Clearly, these findings are more consistent with PUPPP. Initial spread of the classic papules and plaques of PUPPP to the palms and soles is a unique way in which this entity can manifest.

Treatment of PUPPP is generally successful with topical corticosteroids and antipruritic agents. Most cases respond to this regimen in 24 to 72 hours with clearing of the rash and resolution of the itching. Oral hydroxyzine or diphenhydramine and emollients provide symptomatic relief. Occasionally, oral corticosteroid therapy in rapidly tapering doses over 6 days is required for effective treatment of particularly severe presentations of this disease.

Finally, the unusually long duration of symptoms into the postpartum period coupled with the waxing and waning effect of systemic steroid entertains new thoughts as to the etiologic factor of PUPPP. Regardless of treatment response, PUPPP is generally described as spontaneously remitting within 10 to 14 days after delivery. Simultaneously, cases requiring systemic steroid therapy respond within 1 week. Our patient did not have complete symptom relief until 6 weeks postpartum after two short courses and one long tapering dose of prednisone. Symptoms did abate on the short courses of oral corticosteroid, only to reoccur with fervor once treatment was complete.

To this date the exact cause of PUPPP is unknown. Some have commented on skin distention as a result of excess maternal weight gain or twin pregnancy as an etiologic factor. Our patient had neither of these entities. The treatment course and duration of our patients’ symptoms suggest an immunologic process.

Pemphigoid gestationis is the only dermatosis of pregnancy with well-defined immunologic parameters. Direct immunofluorescence characteristically shows linear deposits of C3 along the basement membrane zone of the epidermis. An autoantibody reacts with a basement membrane antigen in the mother’s skin and the placenta, fixing C3 to the basement membrane. The autoantibody develops in response to aberrant expression of major histocompatibility complex class II molecules by the placenta during the second trimester. Histologically, one must also find subepidermal bullae on skin biopsy.

Pemphigoid gestationis was initially part of our differential diagnosis. However, our patient never had bullae formation, periumbilical lesions, leukocytosis, or an elevated sedimentation rate as is common in pemphigoid gestationis. Direct immunofluorescent studies for C3 were negative as well. Therefore, the diagnosis of PUPPP was confirmed. Literature search failed to find case reports or research clearly highlighting our inference. However, in a study performed by Zum and colleagues, a novel subtype of PUPPP was discovered. Indirect immunofluorescence performed on the sera of 111 pregnant women with pruritic dermatosis indicated that five of the women had circulating antibasement membrane IgM antibodies. Direct immunofluorescent studies performed to detect C3 were negative in these cases. None of these patients had vesicle or bullae formation, and histologic examination was consistent with a diagnosis of PUPPP. Unfortunately, only one of these patients was a primigravida and only one had a protracted course for 13 weeks. This patient had onset at 11 weeks of gestation and a past medical history significant for atopy. While this study does not provide a definitive answer, it lends evidence to the possibility of a subtype of PUPPP that clearly has an immunologic basis. Whether a protracted duration of symptoms requiring prolonged systemic steroid therapy is the normal course for this subtype will only be realized with further identification of these patients and analysis of their treatment regimens.

Pruritic urticarial papules and plaques of pregnancy is the most common skin condition seen in pregnancy. Unusual presentations of this entity must be recognized. Simultaneously, prolonged systemic steroid therapy may be an option for those patients unresponsive to topical corticosteroids. Finally, an immunologic mechanism may be the underlying cause of this illness, at least in some cases. Clearly, further studies are necessary to define the incidence, symptom patterns, and treatment outcomes of those patients with PUPPP found to have circulating IgM by indirect immunofluorescence.

References
CASE REPORT


