PUPPP: Demonstration of Deranged Liver Enzymes during Pregnancy

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Abstract

PUPPP, a Polymorphic Eruption of Pregnancy (PEP), is a chronic hives-like rash that strikes some women during pregnancy. The condition typically develops in the third trimester of pregnancy or immediately in the postpartum period. Clinically, it appears as an intensely pruritic (itchy) rash consisting of raised, edematous areas of small papules coalescing into larger plaques. The case report concerns a primigravida woman (period of gestation – 36 weeks) diagnosed with pruritic urticarial papules and plaques of pregnancy (PUPPP). She had been married since 10 months and this is her first pregnancy. The patient was admitted to labor room of Hakeem Abdul Hameed Centenary (HAHC) Hospital, New Delhi, with chief complaints of fever since 2 days, cellulitis since 7 days. During physical examination, rashes were seen on the face, abdomen and legs. Per abdomen examination revealed cephalic presentation of the fetus with FHR as 142 bpm. Routine blood investigations revealed that the patient had gestational hypertension (albumin trace in urine) and the serum bilirubin total was also raised (1.58 mg/dL). The Liver Function Test (LFT) results were, SGOT (200 IU/L), SGPT (144 IU/L) and ALP (262 IU/L). After all the required investigations, she was diagnosed with PUPPP with GHTN.

Keywords: Pruritic urticarial papules and plaques of pregnancy (PUPPP), Gestational hypertension (GHTN)

Introduction

A 25-year old primigravida woman came to labor room of Hakeem Abdul Hameed Centenary (HAHC) Hospital, New Delhi, with chief complaints of fever since 2 days, cellulitis since 7 days. The patient was having 36 weeks of gestation and was also diagnosed as gestation hypertension. The patient visited the hospital regularly during the pregnancy. There was no history of any previous pregnancy and miscarriage. The patient weighed 86 kg and had been married for 10 months. There was no history of consanguineous marriage of parents and there was no family history of any obstetrical or gynecological disease in her family. Her age at menarche was 15 years and her menstrual cycles were regular. She had no significant past history of diabetes, hypertension or any other medical illness. Physical examination revealed the presence of rashes on the face, abdomen and lower legs of the patient and per abdomen examination revealed cephalic presentation of the fetus with FHR as 142 bpm. Abdominal girth and fundal height were proportionate to the gestational age. Routine blood investigations revealed that the patient was a case of gestational hypertension (albumin trace in urine) and the serum bilirubin total was also raised (1.58 mg/dL). The Liver Function Test (LFT) results were (200 IU/L), SGPT (144 IU/L) and ALP (262 IU/L).

After all the required investigations, she was diagnosed with pruritic urticarial papules and plaques of pregnancy (PUPPP) with gestational hypertension (GHTN). PUPPP, known in United Kingdom as polymorphic eruption of pregnancy (PEP),

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Epidemiology

The incidence of PUPPP in a single pregnancy is estimated to occur in approximately one in 200 pregnancies (0.5%). PUPPP is predominantly a disorder that occurs in a patient's first pregnancy (primigravida).[2] Polymorphic eruption of pregnancy also known as PUPPP, occurs with an incidence of one in 160 pregnancies and is the second-most common skin dermatosis in pregnancy after atopic eczema.[3]

Etiology

Despite the relative frequency of PUPPP, little is known about its etiology. There is usually no previous or family history of this type of eruption and studies of HLA types have revealed no consistent HLA association in women with PUPPP. The various etiological theories proposed can be considered under the following headings.

Abdominal Distension

The most striking and consistent clinical observation of PUPPP is that the eruption usually begins in and around abdominal striae. Localization of lesions to abdominal striae combined with a high frequency of multiple pregnancies in these patients has suggested to some authors that abdominal distension, or a reaction to it, might trigger PUPPP. In 90% of the patients with PUPPP studied by Holmes, the eruption was thought to have developed as a consequence of damage to connective tissue within the striae.[4]

Hormonal Etiology

Because PUPPP occurs in pregnancy with multiple associated hormonal changes, and is more common in twin pregnancies, a hormonal basis for this eruption has been explored by some authors. Multiple gestations, as frequently seen in patients with PUPPP, can be associated with high levels of progesterone, and increased progesterone receptor immunoreactivity has been demonstrated in skin lesions of patients with PUPPP.[4]

Autoimmune Hypothesis

Unlike pemphigoid gestations, there is no evidence to suggest that PUPPP is an autoimmune disease in the usual sense. There are no circulating autoantibodies in patients with PUPPP that have been detected to date. Unlike pemphigoid gestations, the condition generally does not recur in subsequent pregnancies as might be expected in a patient, whose immune system had been primed by a previous event, and if it does recur it is not necessarily more severe, nor does it occur earlier in the pregnancy.[4]

Another Hypothesis

- Increased levels of fetal DNA that have been detected in the skin of PUPPP patients may contribute to the pathology.
- There is some evidence that patients with atopy may be predisposed to PUPPP, as well as patients who are hypertensive or obese.
- Strongly associated with maternal weight gain and multiple gestations.[4]

Clinical Presentation

The most common symptom of PUPPP is itching or pruritus which is present in the patient as discussed.[5]

Book Picture	Patient Picture
Very itchy rash	Present
Redness	Present
Small blisters	Present
Eczema-like lesions	Absent
Appears on abdomen first	Present
Is not found on the belly button	Absent
Stretching of the skin, stretch marks are generally the first sign	Present

Diagnosis

PUPPP is often a diagnosis of exclusion, and physicians may obtain laboratory testing in order to rule out other dermatoses. Lab tests may include complete blood count, comprehensive metabolic panel, liver function tests, serum human choriogonadotropin, serum cortisol, serum for indirect immunofluorescence, and biopsy of the lesions for direct immunofluorescence to rule out pemphigoid gestationis.[6]

Results of skin biopsy, if performed, reveal nonspecific changes with some inflammation and infiltrate around the vasculature, spongiosis and the presence of eosinophils. Direct immunofluorescence testing, when special stains are used to look for the presence of antibodies directed at specific structures in the skin, is usually negative in PUPPP.[6]

A 25-year-old primigravida underwent several tests, i.e., complete blood count, liver function test in which her liver enzymes were raised [SGOT (200 IU/L), SGPT (144 IU/L) and ALP (262 IU/L)], kidney function test, urine routine (albumin trace), ultrasonography of whole abdomen and ultrasound doppler (edema in both legs).

Management

- In most cases, symptoms are adequately controlled using a topical corticosteroid cream, such as Betamethasone Valerate 0.1% applied 2–4 times daily.
- Sedating first-generation antihistamines such as Pheniramine appear to be safe in pregnancy, as they are a category A drug, and can be used as adjunct therapy in PUPPP to provide relief from pruritus, which at times can be severe and disturb sleep, leading to exhaustion of the mother.
- General measures, such as cool soothing baths, frequent application of emollients, wet soak applied to the skin, light cotton clothing, etc., provide symptomatic relief.
- In occasional cases, either when topical corticosteroids are ineffective or when lesions are extensive, oral Prednisolone in doses ranging from 40 to 60 mg daily induces a prompt resolution of symptoms and the eruption. Systemic corticosteroids are frequently necessary in the treatment of maternal asthma, inflammatory bowel disease, and autoimmune disease. It has been suggested that the maternal-fetal gradient of Prednisolone is only 10:1 and fetal adrenal suppression is a rare sequel.[7]

Patient had been treated with Tab. Ursodeoxycholic acid (Udiliv) 600 mg twice a day which suppresses hepatic synthesis and secretion of cholesterol and Tab. Labetalol 200 mg twice a day which acts as a beta blocker.

Prognosis

The rash usually regresses within a week postpartum; however, it can also clear at any time before birth and up to the sixth week postpartum. In a few cases, postpartum exacerbation of PUPPP has been observed. PUPPP does not usually recur in future pregnancies. Most studies of women with PUPPP confirm the absence of associated maternal and fetal morbidity and mortality in patients, and it is thought that PUPPP is harmless to both the mother and the fetus.[4]

Nevertheless, in a few case studies, complications potentially associated with dermatosis have been observed. Moreover, spontaneous abortion in the ninth week was noticed in a patient whose eruption appeared in the seventh week of gestation. It was suggested possible association of PUPPP and severe preeclampsia with fetal death. They reported a case of a woman with PUPPP who had a stillbirth.[4]

On postoperative day 8 after LSCS, the pruritic rashes reduced and itching also reduced over the abdomen and lower limbs.

Conclusion

Though PUPPP is the most common skin condition seen during pregnancy, many questions pertaining to this disease remain unanswered. This review points to PUPPP as a well-defined entity, the diagnosis of which is based mainly on the clinical presentation (onset, typical localization, and appearance of changes). In women who have unusual presentations, laboratory investigation, histologic examination, and immunologic studies can be performed to exclude more serious pregnancy disorders. These persons should be referred to a dermatologist for diagnostic work-up.

Treatment of PUPPP focuses on the mitigation of pruritus. Antihistamines, skin emollients, and topical steroids are the primary agents used. In some cases, a short course of oral corticosteroids may be of value.

Conflict of Interest: None

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