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Case Report

Severe Polymorphic Eruption of Pregnancy Developing in A Late Term Primigravida: A Case Report and Review of Literature

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ABSTRACT

Background: Polymorphic eruption of pregnancy [PEP] is a benign inflammatory dermatosis of pregnancy of unknown etiology developing in the third trimester as an intensely pruritic rash and inflamed striae with peri-umbilical sparing. It is a self-limiting condition which resolves after delivery and has no maternal or fetal adverse effects. There are no specific investigations. Diagnosis is based on history and typical clinical features. Emollients, topical corticosteroids and oral antihistamines provide symptomatic relief, though a few patients with severe symptoms may require a short course of oral or injectable corticosteroids. Autologous hemotherapy is a safe option and can be considered in severe conditions.

Case summary: This report describes an unusual patient with singleton pregnancy presenting at 41 weeks gestation with sudden onset intense itching and rash on abdomen, upper part of legs and arms. We have summarized current literature related to PEP and evaluated the variations in clinical presentations, relevant etiopathogenesis and treatment options.

Keywords: Polymorphic; Dermatosis; Pruritis; Peri-umbilical sparing; Pregnancy.

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INTRODUCTION

Polymorphic eruption of pregnancy [PEP] is a benign dermatosis of pregnancy which presents as an intensely pruritic rash with inflamed striae in the late third trimester[1]. It is also known as Pruritic urticarial papules and plaques of pregnancy [PUPPP]. It resolves spontaneously and completely near the time of delivery and is not associated with significant adverse maternal or fetal effects[2].

This report describes an unusual patient with singleton pregnancy presenting at 41 weeks gestation with sudden onset intense itching and rash on abdomen, upper part of legs and arms.

We have summarized current literature related to PEP and evaluated the variations in clinical presentations, relevant etiopathogenesis and treatment options.

Case Report

A 26-year-old primigravidae reported to our hospital at 41-week and 3-day period of gestation with complaint of rash with intense itching, mainly on the lower abdomen and upper part of legs for the past 2 days. There was no history of associated fever or medication. The woman was under our regular follow up. On examination, there were urticarial papules on the lower abdomen along the striae but sparing the umbilical area [Figure 1].

The eruptions were also present on the buttocks, thighs, legs and arms [Figures 2 and 3].

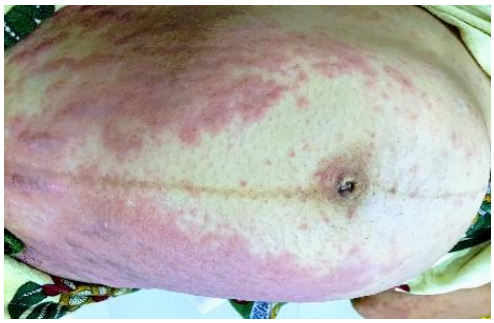

Figure [1]: Polymorphic rash on abdomen with peri-umbilical sparing

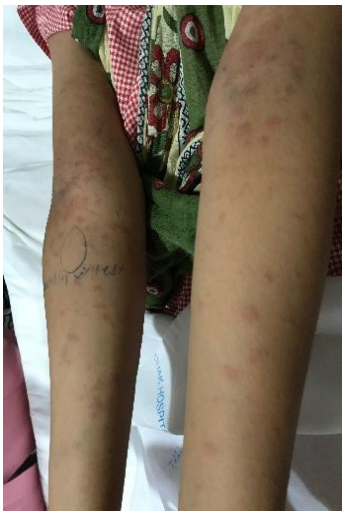

Figure [2]: Rash on bilateral arms

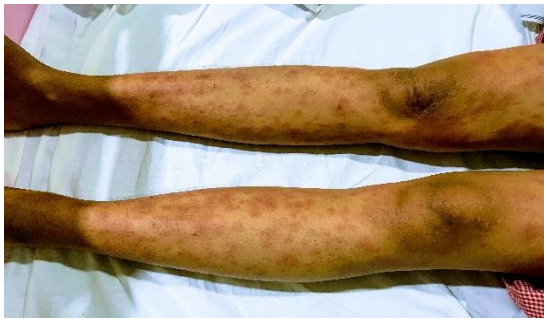

Figure [3]: Rash on both lower limbs

Per abdomen examination showed that the fetus was in transverse lie with a normal fetal heart rate. Her blood investigations including Liver Function Test and Renal Function Test were within normal limits.

Her obstetric ultrasound showed moderate oligohydramnios with normal Doppler indices. She was advised termination of pregnancy by Caesarean section in view of transverse lie with moderate oligohydramnios.

A healthy female baby of 3.25kg was delivered with APGAR scores of 8 and 9 at 1 and 5 min of life. The baby had no cutaneous lesions. She was given injectable corticosteroid in the immediate post-operative period as she was distressed by the intense itching. Otherwise, her post-operative period was uneventful. She was discharged on anti-histaminic, 1% topical steroid and emollients.

On follow up visit after 2 weeks, her urticarial rash had completely resolved [Fig – 4, 5 and 6].

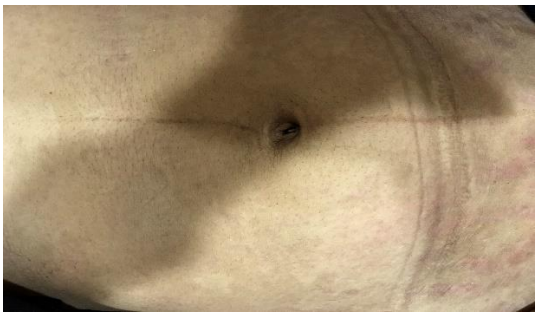

Figure [4]: Resolution of rash on abdomen after delivery

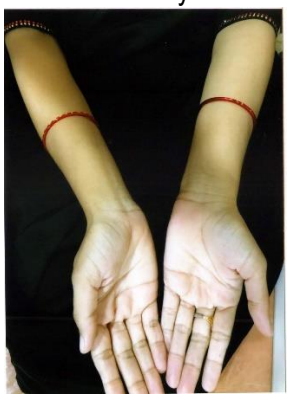

Figure [5]: Resolution of rash on arms after delivery

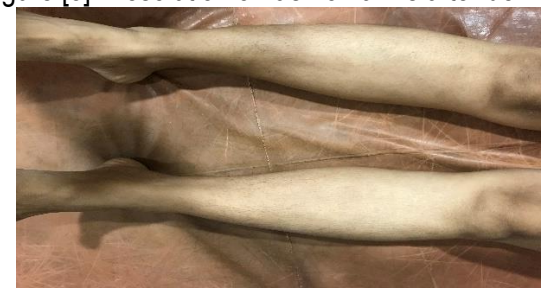

Figure [6]: Resolution of rash on lower limbs after delivery

DISCUSSION

Polymorphic eruption of pregnancy [PEP] is a self-limiting inflammatory pruritic dermatosis of pregnancy. It evolves in the third trimester and resolves rapidly and spontaneously postpartum[2]. It affects 0.5% of pregnancies[3], mainly primi-gravidae[4]. 15% cases may occur in the immediate postpartum period[2,5]. PUPPP is the usual term in the United States whereas PEP is the preferred term in the United Kingdom and Europe. The incidence of PEP in the Indian population is not known either because it is under reported or because the incidence is very low. The exact pathophysiology is not clear. Various studies have shown an association with excessive maternal weight gain and multiple pregnancies suggesting a possible relation with increased stretching of abdominal skin[2]. It is more commonly reported in women with white skin than in women with black skin[2]. One study has implicated low serum cortisol levels suggesting a hormonal influence[6].

Investigators identified fetal deoxyribonucleic acid [fetal DNA] in the skin of mothers with PEP, suggesting that chimerism may be relevant to the pathogenesis of this disorder[7].

Three clinical subtypes have been described[8]:
Type I- mainly urticarial papules and plaques.
Type II- non-urticarial erythema.
Type III- papules and vesicles and combinations of the two forms.

Diagnosis is on the basis of history and typical clinical features. Polymorphic eruptions are usually seen in later part of third trimester of pregnancy. There are reports of PEP developing after delivery, from 5 days to 3 weeks postpartum[3,9,10,11]. It usually involves abdominal striae, with peri-umbilical sparing. The rash spreads from the abdomen to the buttocks and thighs, and sometimes the arms and legs. The face, palms or soles may be rarely involved[12,13] and exclusive involvement of the extremities have also been reported[14].

The lesions consist of urticarial papules, plaques and polycyclic wheals. The lesions are intensely pruritic but excoriations are not seen. Small vesicles [<2mm] and target lesions may also be seen but bullae are absent[15,16]. Histopathology and immunofluorescent studies are non-specific and not required for diagnosis. Skin biopsy shows epidermal and upper dermal edema with a peri-vascular infiltrate of lymphocytes, histiocytes and eosinophils. Immunofluorescent studies are negative[17].

The main differential diagnoses are Prurigo of pregnancy, Pemphigoid gestationis, idiopathic urticarial and drug eruption. A pruritic rash early in pregnancy may indicate Prurigo of pregnancy or Atopic eczema. When the rash starts around the umbilicus, or bullae are present, a diagnosis of Pemphigoid gestationis must be considered. Pemphigoid gestationis can cause adverse fetal effects like low birth weight, preterm birth and stillbirth. 10% babies can develop similar transient bullous eruption when born to mothers with Pemphigoid gestationis.

General treatment measures include cool,

soothing baths, emollients, wet soaks and light cotton clothing. The patient should be reassured that it will resolve after delivery spontaneously within 4-6 weeks with no adverse maternal or fetal effects.

High potency topical corticosteroids [Class I and II] like Fluocinonide or Fluticasone are used for symptom alleviation of pruritis[18]. First generation oral Anti-histamines like Diphenhydramine help to alleviate pruritis and improve sleep. Systemic steroids are indicated in resistant cases[14]. Successful treatment with intramuscular autologous hemotherapy has been reported with a good safety profile[19]. There is early regression of skin lesions and pruritis, improving the quality of life. PEP generally does not recur with pregnancy. If it recurs, it is milder.

Conclusion: PEP is an intensely pruritic dermatosis of pregnancy with a variable clinical presentation. There are no specific investigations and the diagnosis is based on clinical polymorphic findings. It is self-limiting with no adverse maternal or fetal effects. Treatment is limited to emollients, antihistamines and topical or systemic corticosteroids. Autologous hemotherapy is a safe option in severe conditions

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