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Case Report

Beyond the striae: an atypical case of polymorphic eruption of pregnancy with vasculitic features

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ABSTRACT

Polymorphic eruption of pregnancy (PEP), also known as pruritic urticarial papules and plaques of pregnancy (PUPPP), is the most common benign dermatosis of pregnancy. We report a case of a 26-year-old primigravida at 32 weeks gestation who developed an extensive pruritic rash initially on the abdomen and later spreading to the extremities, back, flanks, and palms, palpable purpura was observed on the lower extremities, suggesting a vasculitic component. Skin biopsy revealed features of both PEP and leukocytoclastic vasculitis, while direct immunofluorescence for IgA was negative. The patient was managed conservatively with complete recovery. This case highlights a rare presentation of PEP with vasculitic features, necessitating careful evaluation. Further research is needed to explore possible triggers, including immune mechanisms or viral associations, to refine diagnostic criteria and optimize management strategies.

Keywords: Polymorphic eruption of pregnancy, Atypical PEP presentation, Pregnancy dermatoses

INTRODUCTION

Polymorphic eruption of pregnancy (PEP) also known as 'Pruritic Urticarial Papules and Plaques of Pregnancy (PUPPP)' was first introduced by Lawley to describe the most common benign dermatosis occurring during pregnancy. PUPPP typically emerges in the third trimester or, in some cases, shortly after childbirth. It is characterized by the sudden onset of intensely itchy papules measuring 1–2 mm, which may merge to form plaques.

Some cases may also present with papulovesicular, urticarial wheals, target-like lesions, or eczematous changes. The rash predominantly appears on the distended abdomen, usually sparing the periumbilical region, and is often associated with striae. In more severe cases, the rash can extend to the extremities and, in rare instances, become generalized.[1] We report a case of PEP in a young, pregnant lady who had extensive involvement of body along with vasculitic lesions in the extremities. This case report is being reported for its rarity.

CASE REPORT

A 26-year-old primigravida, at 32 weeks of gestation with a singleton pregnancy, presented to our department with complaints of pruritic red lesions that had been progressively worsening over the past 10 days. The rash initially appeared on her abdomen and gradually spread to her extremities and back. She denied any recent history of fever, upper respiratory tract infection, abdominal pain, burning micturition, joint pain, or recent drug intake or any recent travel. She had a known history of hypothyroidism but no known drug allergies or other significant medical conditions. Her general and systemic examinations were essentially normal.

Dermatological examination revealed, multiple, erythematous papules of varying sizes, coalescing into plaques primarily over the abdomen, particularly within

the striae distensae, with sparing of the periumbilical region (Figure 1B) Similar erythematous papules, along with few vesicles, were also observed on the back, flanks, chest, thighs, and upper limbs, including the palms (Figure 1A, 1C).

Additionally, palpable, non-blanchable purpura was noted on the lower legs (Figure 1D), raising suspicion of a vasculitic component. There was no mucosal involvement.

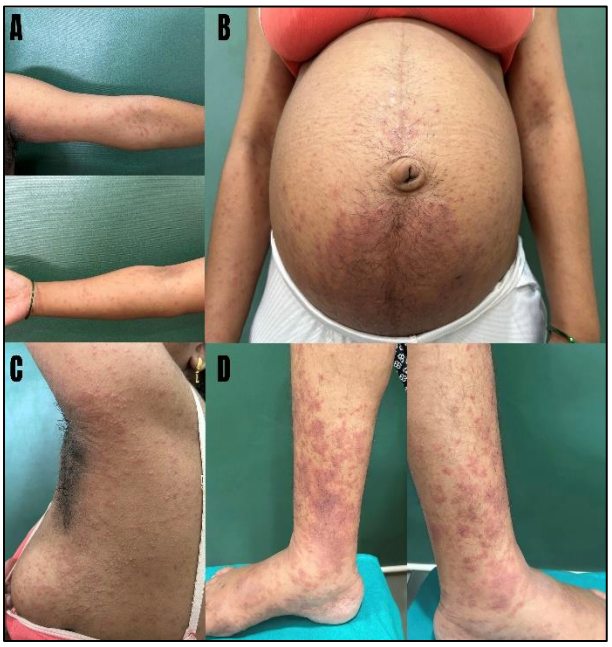

Figure 1: (A) Multiple, erythematous papules of varying sizes, coalescing into plaques, particularly within the striae distensae, with sparing of the periumbilical region. (B and C) Erythematous papules, along with few vesicles, over flanks, bilateral axilla, upper limbs with involvement of palms. (D) Discrete palpable purpura over lower extremities.

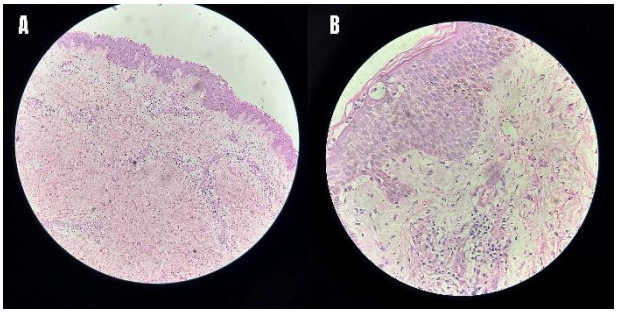

Figure 2 (A and B): Mild orthokeratosis, spongiosis, and sub-corneal vesicles with inflammatory cells. Moderate perivascular and peri-adnexal lymphoplasmacytic and eosinophilic infiltrate. Vasculitis with RBC extravasation and plump endothelial cells, without fibrinoid necrosis. Mild dermal oedema; subcutaneous tissue unremarkable.

PEP, IgA vasculitis, early form of pemphigus gestationis and exanthematous rash were considered in the differential diagnoses. Her routine haematological, biochemical tests along with urine analysis, viral markers, ANA, C3, C4, p-ANCA, c-ANCA, anticardiolipin antibody were essentially normal. A skin biopsy performed from the left leg lesion revealed mixed picture of PEP and leukocytoclastic vasculitis (Figure 2). Her direct immunofluorescence result for IgA was negative. The combination of clinical and histopathological findings suggested a mixed presentation, incorporating features of pregnancy-associated dermatoses and vasculitis.

The patient was managed with emollient, topical mometasone furoate 0.1% and tab levocetirizine 5 mg. She responded well to treatment, with a gradual reduction in symptoms. She delivered a healthy male infant at 35 weeks gestation, with a birth weight of 2700 grams. She had complete resolution of skin lesions after four weeks of childbirth.

Table 1: Specific dermatoses of pregnancy are classified as.[3]

Condition	Clinical features	Characteristics	Diagnosis
Pemphigoid gestationis	Erythematous urticarial papules and plaques	Involves the umbilical region, may generalise, autoimmune (BP180 antibodies)	Skin biopsy, DIF
Polymorphic eruption of pregnancy	Urticarial papules and plaques, initially on abdominal striae	Later becomes polymorphic, spares periumbilical area	Clinical exam, biopsy for differentiation
Atopic eruption of pregnancy	Appears earlier; associated with atopy	Type E: Eczematous (face, neck, flexures), Type P: Prurigo (trunk, limbs), Pruritic Folliculitis: Follicular papules, pustules (trunk)	Clinical history
Intrahepatic cholestasis of pregnancy	Generalised pruritic without primary lesions	Increased bile acids, secondary excoriation, possible jaundice	Serum bile acids

DISCUSSION

It is not uncommon to have dermatoses during pregnancy. At times it poses diagnostic challenge and apprehension on pregnant ladies. Traditionally, the dermatoses of pregnancy are specific and nonspecific. In year 2006, Ambros-Rudolph et al had classified the specific dermatoses of pregnancy into four groups namely pemphigoid gestation is, polymorphic eruption of pregnancy, atopic eruption of pregnancy, intrahepatic cholestasis of pregnancy.[2] The salient features of this are illustrated in Table 1.[3]

Non-specific dermatoses of pregnancy include physiological changes such as hyperpigmentation, stretch marks, and vascular changes. Pre-existing skin conditions like atopic dermatitis, psoriasis, acne, and lupus may persist or worsen during pregnancy. Additionally, infections such as candidiasis, bacterial infections (impetigo, cellulitis), and viral infections (herpes, varicella) can also occur.[4]

PEP is a self-limiting inflammatory disorder with an unclear cause due to limited research. Risk factors include first-time pregnancy, maternal weight gain, multiple pregnancies, and skin stretching. It is believed that connective tissue damage and collagen exposure may trigger an inflammatory response.[4] Hormonal imbalances, particularly reduced cortisol levels, and foetal DNA in the maternal dermis are potential contributors.[2,5] Possible links to IVF and male foetuses have been noted, but evidence is insufficient. Despite various hypotheses, a definitive causal explanation is still lacking.[6]

Histopathology findings of PEP are non-specific. Early stages show epidermal spongiosis, dermal oedema, and perivascular inflammation with T-helper lymphocytes, macrophages, and occasional eosinophils. DIF is negative. Older lesions may show acanthosis, hyperkeratosis, parakeratosis, and epidermal hyperplasia.[5]

PEP has a favourable prognosis for both mother and foetus, is self-limiting, and rarely reoccurs. Treatment is symptomatic, with mild cases responding to moisturizers, emollients, and low- to mid-potency corticosteroids like fluticasone. Sedating antihistamines (e.g., pheniramine, hydroxyzine) are safe in pregnancy.[1] Supportive measures include cold baths, menthol, urea applications, and cotton clothing.[2]

Severe cases may require systemic corticosteroids, though risks are rare. UVB therapy, early delivery, or autologous whole blood (AWB) injections have shown success in refractory cases.[1,7] AWB may modulate immune response and improve symptoms.[7]

IgA vasculitis (formerly Henoch-Schonlein purpura) is a small-vessel vasculitis affecting the skin, joints, GI tract, and kidneys. Though rare in pregnancy, it can lead to hypertension, proteinuria, and foetal complications. Symptoms include purpuric rash, arthralgia, abdominal pain, and haematuria. Close monitoring is crucial, especially for renal involvement. With timely care, maternal and foetal outcomes are generally favourable.[8]

This case report describes a rare presentation of extensive PEP with vasculitic features, requiring careful evaluation for differentiation. Most previously published case reports of PEP primarily involved the abdomen (striae) with periumbilical sparing, with occasional mild extremity involvement. In our case, the polymorphic lesions initially appeared on the abdomen and striae in the third trimester, consistent with typical PEP. However, over time, the rash became widespread, affecting the extremities, back, flanks, palms, and eventually developing vasculitic-like features on the lower extremities. Given the presence of a vasculitic component in a PEP patient, further research is needed to explore potential external triggers or viral associations. Additional studies are required to better understand this phenomenon.

CONCLUSION

PEP is one of the commonest dermatoses of pregnancy. Mostly, it presents with erythematous papules, plaques and urticarial lesions on the abdomen with occasional involvement of extremities. Our case was unique to have extensive lesions along with palpable purpura suggestive of vasculitic lesions. This case highlights that PEP may have vasculitic lesions when there are extensive lesions. Given the self-limiting nature of PEP and the potential complications associated with vasculitis, a cautious approach with close monitoring is essential. Increased awareness and further documentation of such atypical cases in the literature will aid in refining diagnostic criteria and optimizing management strategies for pregnancy-associated dermatoses.

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