International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Singh D et al. Int J Reprod Contracept Obstet Gynecol. 2026 Apr;15(4):1387-1391
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20260904

Case Report

A rare presentation of pemphigoid gestationis in pregnancy

Divya Singh[1]*, Soumya P. Prasad[2]

[1]Department of Obstetrics and Gynaecology, Mansarovar Medical College, Sehore, Madhya Pradesh, India
[2]Maternal Fetal Medicine, Cloudnine Hospital, Kailash Colony, New Delhi, India

Received: 17 December 2025
Revised: 18 February 2026
Accepted: 19 February 2026

***Correspondence:**
Dr. Divya Singh,
E-mail: gynecdivya@gmail.com

ABSTRACT

Pemphigoid gestationis (PG) is a rare autoimmune subepidermal blistering disorder of pregnancy associated with maternal morbidity and potential adverse fetal outcomes. We report a 36-year-old gravida three, para two woman who developed non-pruritic erythematosquamous lesions at 24 weeks of gestation without prior dermatologic history. Dermatologic evaluation supported the diagnosis of PG. The patient responded favorably to topical corticosteroid therapy, with no maternal or fetal complications. Elective cesarean delivery was performed at 39 weeks, resulting in the birth of a healthy neonate. Postpartum recovery was uneventful, with near-complete lesion resolution by four months. This case highlights an atypical, non-pruritic presentation of PG and underscores the importance of early recognition, multidisciplinary management, and careful antenatal surveillance to optimize maternal and fetal outcomes.

Keywords: Pemphigoid gestationis, Autoimmune bullous disease, Pregnancy dermatoses, BP180, Corticosteroids

INTRODUCTION

Pemphigoid gestationalis (PG) is a rare autoimmune bullous dermatosis characterized by the formation of autoantibodies against adhesion molecules, ultimately leading to the loss of intercellular cohesion and the appearance of blisters on the skin and mucous membranes. Pregnancy has been observed to exacerbate, trigger relapse, or lead to the first-time onset of pemphigoid disorders. These manifestations may occur at any point during pregnancy or the puerperium but are most frequently noted during the second and third trimesters.[1]

The association between pregnancy and the relapsing-remitting course of the disease is thought to be influenced by fluctuating progestin levels.[2] Disease activity usually declines towards the end of the pregnancy when progesterone levels are high, and then increases immediately postpartum when progesterone levels decrease and estrogen levels rise; likewise, progesterone levels decrease premenstrually when PG can potentially flare.

Clinically, PG presents with numerous pruritic vesicles that may evolve into widespread erosions and ulcerations, which generally heal without scarring because the erosions are confined to the epidermis.[3] Lesions may be localized or generalized, with a predilection for the scalp, face, axillae, groins, and pressure-bearing areas. Oral and nasal mucosal involvement is common, and in some cases, other mucosal surfaces, including vulva, may also be affected.[4]

The diagnosis is predominantly clinical, supported by confirmatory tools such as direct immunofluorescence, light microscopy, serologic testing, and histopathological examination.[5] Given its rarity, PG must be distinguished from other more common gestational dermatoses, including atopic eruption of pregnancy, polymorphic eruption of pregnancy, and intrahepatic cholestasis of pregnancy.

Epidemiology

PG has an estimated incidence of 1 in 20,000-50,000 pregnancies.[6] In a small number of cases, the disease can

be a paraneoplastic manifestation of trophoblastic tumours, hydatiform mole, or choriocarcinoma. It sometimes relapses with the onset of menses or use of oral contraceptives postpartum but quickly remits after their discontinuation.[7,8]

PG is not significantly associated with other autoimmune disease except for hyperthyroidism (Grave's disease), from which 10-11% of PG patients suffer compared to 0.4% in the general female population.[9] This can partially be explained by the presence of HLA-DR3 and DR4.

Pathogenesis

The pathogenesis of PG is considered to be similar to that of bullous pemphigoid, which is characterized by deposition of autoreactive antibodies directed against two hemidesmosomal proteins, BP 180 and BP 230, within the dermoepidermal junction, resulting in the formation of bullae and skin erosions.[10]

In PG, the first immune response occurs within the placenta. Placental trophoblasts and amnio chorionic stromal cells show an abnormal expression of major histocompatibility complex (MHC) class II antigens allowing the presentation of BP180 (also known as BPAG1 or collagen XVII) protein to the maternal immune system.[11] BP180 is a key structural protein of hemidesmosomes linking the epidermis and dermis. BP180 is found in placental tissue, in fetal membranes, and also in the basement membrane zone of the skin.[12]

These specific proteins presented in the placenta are recognized as foreign, causing subsequent production of anti-placental IgG antibodies that cross-react with the same BP180 proteins in the skin. The binding of these antibodies to the basement membrane of the skin triggers an autoimmune response that consists of complement activation, deposition of immune complexes, consecutive chemoattraction of eosinophil granulocytes, and subsequent degranulation, resulting in tissue damage and blister formation.[13]

PG and pregnancy

Pregnancies affected by PG carry an increased risk of preterm birth and foetal growth restriction, which are believed to result from mild placental dysfunction associated with circulating BP180 antibodies.[14] The use of systemic corticosteroids, while often necessary to control disease activity, has been linked to complications such as foetal growth restriction, gestational diabetes, eclampsia, and preterm delivery.[15]

In most cases, PG lesions resolve spontaneously within 12-16 weeks postpartum without scarring. Corticosteroid therapy can generally be tapered and discontinued soon after delivery, although treatment may need to be resumed if disease flares occur.[16]

PG and the foetus

Most women with PG deliver healthy, full-term infants without complications. Neonatal pemphigoid, although rarely reported, is a transient autoimmune blistering condition resulting from the transplacental transfer of maternal antibodies. The condition is typically self-limiting and resolves within 2-3 weeks without specific treatment and without long-term sequelae.[17]

A few studies have estimated that out of 100 live-born children with available data, 83 (83%) were healthy without skin lesions, while 13 (13%) had skin lesions (urticarial and/or bullous). Two babies born from pregnancies complicated by anhydramnios died of sepsis few days after delivery and two had severe growth retardation.[18]

CASE REPORT

A 36-year-old gravida three, para two woman was admitted to the department of obstetrics and gynaecology at Lok Nayak Hospital, Delhi, at 34 weeks of a spontaneously conceived pregnancy. She presented with non-pruritic skin lesions that first appeared at 6 months of gestation over her hands and gradually progressed to involve the bilateral legs, arms, and abdomen. The lesions were confluent, erythematosquamous, and partially crusted.

Her previous two pregnancies, both delivered by caesarean section for fetal indications, were uneventful, and she reported no similar dermatologic complaints during previous pregnancies or the interconceptional period. Her current antenatal course had been uncomplicated, with regular supplementation of iron, folic acid, and calcium. Routine haematological and radiological investigations were within normal limits.

The patient was admitted for further evaluation and management. Following dermatology consultation, topical corticosteroids were initiated. Daily blood pressure monitoring and fetal growth assessments were conducted and were appropriate for gestational age. Routine laboratory investigations remained normal. After three days of treatment, no new lesions appeared, and existing lesions demonstrated significant healing; therefore, topical corticosteroid therapy was continued. No maternal or foetal complications occurred during hospitalization.

An elective caesarean section was performed at 39 weeks once no active lesions were present at the incision site and most lesions had healed.

The procedure was uneventful, and she delivered a healthy infant. Her postpartum course was smooth, with significant improvement in skin lesions by one month and near-complete resolution with minimal to no scarring by four months postpartum.

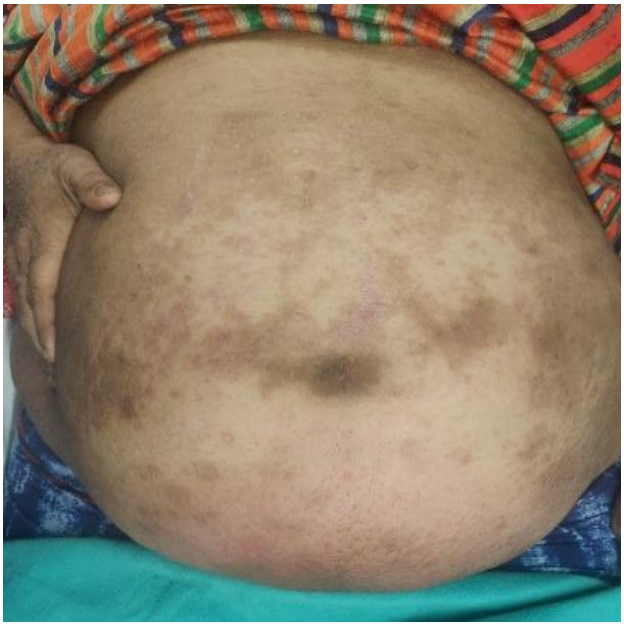

Figure 1: Lesions over abdomen.

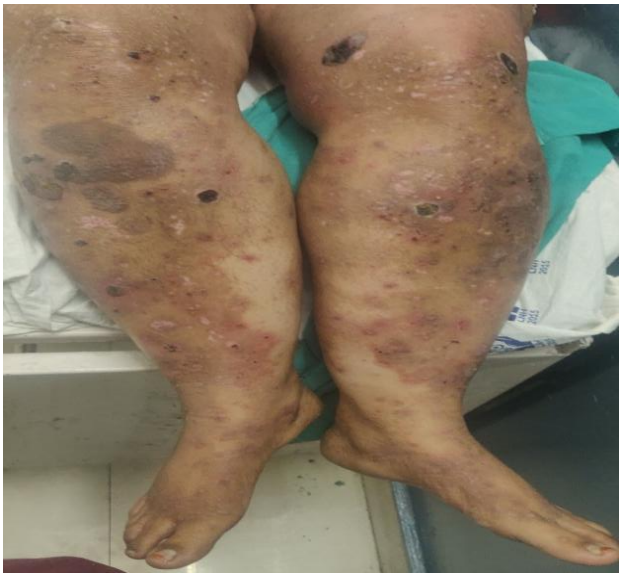

Figure 2: Lesions over bilateral legs.

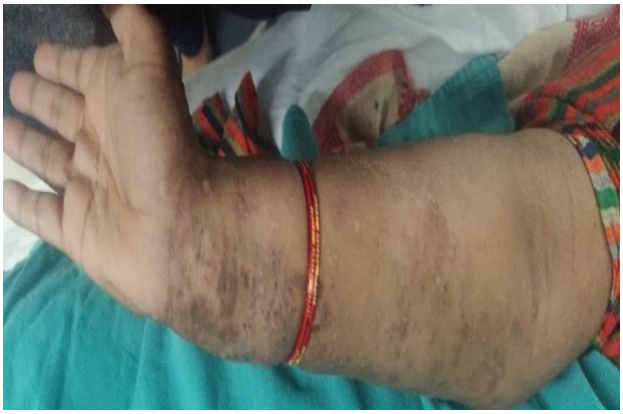

Figure 3: Lesions over right arm.

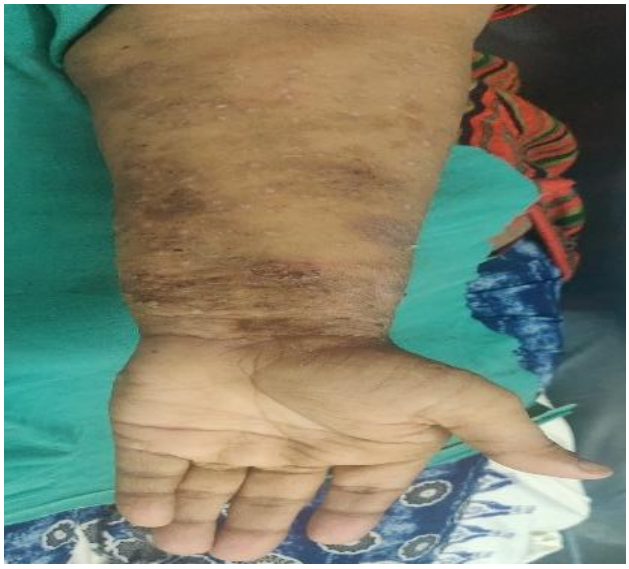

Figure 4: Lesions over left arm.

DISCUSSION

Due to the dynamic immunological and hormonal milieu of pregnancy, PG may worsen or first present during gestation.

PG typically presents with severe pruritus preceding blister formation; however, our patient demonstrated an atypical non-pruritic presentation, making this case clinically significant. Ambros-Rudolph et al described abdominal and periumbilical predominance in 90% of cases, with later generalization.[19] In contrast, our patient initially exhibited lesions over the extremities, an uncommon primary distribution.

PG initially presents with intense pruritus and inflammatory skin lesions. Pruritus can remain the only symptom, but mostly it develops into eruptive polymorphic skin lesions. Eruptive skin lesions initially present as urticarial papules and annular plaques, followed by vesicles and finally large tense bullae on an erythematous background.[19] Skin lesions typically develop on the abdomen, characteristically involving the umbilical region. In 90% of the cases, it later spreads to the rest of the abdomen, and in some patients the involvement of thighs palms, and soles can be prominent.[20,21]

Hallaji et al in their series of 23 patients, reported extremity involvement in 100% of cases, though abdominal onset remained characteristic.[22] Our findings partially align with these observations, although the absence of pruritus represents a deviation from classical presentation.

Many patients experience remission during late pregnancy, sometimes followed by a flare immediately after delivery which in the present case did not have any flare up post delivery.

In the present case, the patient developed lesions for the first time during her third pregnancy at around 6 months of gestation. Her disease course remained uncomplicated, with no associated maternal or foetal morbidity, and she responded favourably to topical corticosteroids, achieving excellent postpartum recovery.

Management strategies for PG must be individualized and coordinated jointly by obstetricians and dermatologists. The primary goals include controlling the acute phase of the disease while ensuring maternal and foetal safety. In this case, topical corticosteroid therapy was sufficient, as no new lesions developed and existing lesions improved within 3-4 days, making systemic therapy unnecessary.

Systemic and topical corticosteroids are empirically recognized as a cornerstone of PG treatment, especially during the gestation period and in mild-to moderate cases. On the other hand, a wide variety of therapeutic approaches, including steroid-sparing agents such as intravenous immunoglobulin therapy, azathioprine, and dapsone, have been reported for persistent cases refractory to first-line regimens or patients with intolerance to (or medical inadvisability of systemic corticosteroids. Moreover, the management of this disease may be challenging owing to the safety concerns during pregnancy or lactation of some immunosuppressive drugs.[15]

This case underscores the importance of multidisciplinary management and emphasizes that atypical presentations without pruritus can delay diagnosis. Obstetricians must maintain a high index of suspicion in unusual dermatoses during pregnancy

CONCLUSION

Pemphigoid gestationalis is a rare and potentially debilitating blistering disorder of pregnancy. Given its association with adverse outcomes such as preterm birth and fetal growth restriction, careful and continuous monitoring is essential. Multidisciplinary management involving both obstetric and dermatologic specialists is crucial.

In conclusion, high-potency topical corticosteroids may be recommended as first-line agents either in mild and in moderate-to-severe PG. Systemic corticosteroids may be a useful approach for recalcitrant disease. Intravenous immunoglobulin therapy alone or in combination with systemic corticosteroids may be considered as further line of treatment. Although most patients achieve complete remission, a considerable proportion of them experience refractory/persistent disease requiring multiple lines of therapy. Randomized controlled trials are needed to better define the effectiveness of different treatments in PG.

Comprehensive counselling should be provided regarding the natural course of the disease, potential maternal and foetal complications, and the risk of postpartum relapse, recurrence in subsequent pregnancies, and flare-ups associated with hormonal contraception.

Further prospective studies are required to better define optimal therapeutic strategies and long-term outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Huilaja L, Mäkikallio K, Tasanen K. Gestational pemphigoid. Orphanet J Rare Dis. 2014;9:1-8.
2. Semkova K, Black M. Pemphigoid gestationis: current insights into pathogenesis and treatment. Eur J Obstet Gynecol Reprod Biol. 2009;145(2):138-44.
3. Fainaru O, Mashiach R, Kupferminc M, Shenhav M, Pauzner D, Lessing JB. Pemphigus vulgaris in pregnancy: a case report and review of the literature. Hum Reprod. 2000;15(5):126-30.
4. Marren P, Wojnarowska F, Venning V. Vulvar involvement in autoimmune bullous diseases. J Reprod Med. 1993;38:101-7.
5. Kneisel A, Hertl M. Autoimmune bullous skin diseases. Part 2: Diagnosis and therapy. J Dtsch Dermatol Ges. 2011;9(11):927-47.
6. Schmidt E, Zillikens D. Pemphigoid diseases. Lancet. 2013;381:320-32.
7. Tindall JG, Rea TH, Shulman I. Herpes gestationis in association with a hydatidiform mole: immunopathologic studies. Arch Dermatol. 1981;117:510-2.
8. Djahansouzi S, Nestle-Kraemling C, Dall P. Herpes gestationis may present as a paraneoplastic syndrome of choriocarcinoma: a case report. Gynecol Oncol. 2003;89:334-37.
9. Shornick JK, Black MM. Secondary autoimmune diseases in herpes gestationis (pemphigoid gestationis). J Am Acad Dermatol. 1992;26:563-6.
10. Patel F, Wilken R, Patel FB. Pathophysiology of autoimmune bullous diseases: nature versus nurture. Indian J Dermatol. 2017;62(3):262-7.
11. Borthwick GM, Sunderland CA, Holmes RC, Black MM, Stirrat GM. Abnormal expression of HLA-DR antigen in the placenta of a patient with pemphigoid gestationis. J Reprod Immunol. 1984;6:393-6.
12. Powell AM, Sakuma-Oyama Y, Oyama N, Black MM. Collagen XVII/BP180: a collagenous transmembrane protein and component of the dermoepidermal anchoring complex. Clin Exp Dermatol. 2005;30(6):682-7.
13. Roth MM. Pregnancy dermatoses: diagnosis, management, and controversies. Am J Clin Dermatol. 2011;12:25-41.
14. Chi CC, Wang SH, Charles-Holmes R, Ambros-Rudolph C, Powell J, Jenkins R, et al. Pemphigoid gestationis: early onset and blister formation are associated with adverse pregnancy outcomes. Br J Dermatol. 2009;160(6):1222-8.

15. Murase JE, Heller MM, Butler DC. Safety of dermatologic medications in pregnancy and lactation: Part I. Pregnancy. J Am Acad Dermatol. 2014;70(3):401.e1-401.14.
16. Shimanovich I, Bröcker EB, Zillikens D. Pemphigoid gestationis: new insights into the pathogenesis lead to novel diagnostic tools. BJOG. 2002;109(9):970-6.
17. Bjarnason B, Flosadóttir E. Childhood, neonatal, and stillborn pemphigus vulgaris. Int J Dermatol. 1999;38(9):680-8.
18. Genovese G, Derlino F, Cerri A, Moltrasio C, Muratori S, Berti E, Marzano AV. A systematic review of treatment options and clinical outcomes in pemphigoid gestationis. Front Med (Lausanne). 2020;7:604945.
19. Ambros-Rudolph CM, Mullegger RR, Vaughan-Jones SA, Kerl H, Black MM. The specific dermatoses of pregnancy revisited and reclassified: results of a retrospective two-center study on 505 pregnant patients. J Am Acad Dermatol. 2006;54(3):395-404.
20. Al-Saif F, Elisa A, Al-Homidy A, Al-Ageel A, Al-Mubarak M. Retrospective analysis of pemphigoid gestationis in 32 Saudi patients-clinicopathological features and a literature review. J Reprod Immunol. 2016;116:42-5.
21. Intong LR, Murrell DF. Pemphigoid gestationis: pathogenesis and clinical features. Dermatol Clin. 2011;29(3):447-52.
22. Hallaji Z, Mortazavi H, Ashtari S, Nikoo A, Abdollahi M, Nasimia M. Pemphigoid gestationis: clinical and histologic features of twenty-three patients. Int J Womens Dermatol. 2017;3(2):86-90.
23. Kushner CJ, Concha JSS, Werth VP. Treatment of autoimmune bullous disorders in pregnancy. Am J Clin Dermatol. 2018;19:391-403.

Cite this article as: Singh D, Prasad SP. A rare presentation of pemphigoid gestationis in pregnancy. Int J Reprod Contracept Obstet Gynecol 2026;15:1387-91.