

Unusual presentation of a Dermatoses of pregnancy- Pemphigoid Gestationis

Adetoun Sanusi ¹ Ms Caroline Rice ²

1. Specialty Registrar in Obstetrics and Gynaecology, North Manchester General Hospital, UK. 2. Consultant Obstetrician and Gynaecologist, North Manchester General Hospital, UK.

INTRODUCTION

Pemphigoid gestationis (PG) is a rare (1 in 50,000 pregnancies) autoimmune skin blistering disorder of pregnancy with maternal and foetal implications such as scarring preterm delivery, neonatal pemphigoid gestationis and low birth weight. It was previously known as herpes gestationis although it has no association with the herpes virus.

It usually presents in mid to late pregnancy and immediate post-partum period with an itchy rash that develops into blisters. The lesions begin in the periumbilical region with the umbilicus being affected and spreads to limbs, palms and soles.

The eruptions can be intensely pruritic and there may be a variable delay between onset of itching and formation of vesicles. Therefore, a high index of suspicion is needed when pregnant patients present with excessive itching especially in the last trimester with normal LFT and bile acids and other common causes of itching without rash formation in pregnancy appear to have been ruled out.

We present a case of pemphigoid gestationis in which there was a significant delay between onset of pruritus and eruption of vesicles and bullae thereby causing a diagnostic dilemma.

PEMPHIGOID GESTATIONIS

1. Previously known as herpes gestationis
2. Incidence 1:50,000
3. Time of onset- 9 weeks GA to one week post partum, but usually in the third trimester.
4. Distribution- lesions begin in the peri umbilical region spreading the limbs, palms and soles.
5. Eruptions are intensely pruritic urticated erythematous papules and plaques which can form vesicles and large tense bullae.
6. Usually, a flare at delivery
7. Urticated plaques may persist for months after delivery and in a few may develop into bullous pemphigoid.
8. Recurrence during ovulation, menstruation, use of COCP, and in future pregnancies.
9. Lesions appear earlier and more severe in future pregnancies.
10. Fetal neonate risk- low birth rate, preterm delivery and still birth, neonatal pemphigoid rash in 10% of babies.
11. Pemphigoid gestationis is an autoimmune condition and is strongly associated with maternal MHC class II antigens haplotypes HLA-DR3 and HLA-DR4.
12. As in other autoimmune conditions, pemphigoid gestationis is associated with an increased risk of other organ-specific autoimmune diseases, in particular Graves disease and Pernicious anaemia.

CASE SUMMARY

A 22-year-old low risk primigravida with isolated pruritus at 34 weeks gestation.

She was started on antihistamines. Blood tests including liver function tests, bile acids, renal function test, coagulation and liver autoimmune screening were essentially normal.

Symphysio-fundal height measurement at 37weeks showed reduced growth trajectory necessitating an obstetric growth scan which showed severe intrauterine growth restriction. Immediate induction of labour was offered and accepted. She had an emergency caesarean section for suspected foetal distress in labour with delivery of a live male neonate-birth weight of 1920g (zero centile).

Placental histology showed mild hyper maturity with extensive chronic auto-immune villitis, which based on the distribution was likely to be a Villitis of Unknown Origin (VUO).

The patient had exacerbation of itching two weeks post partum and developed bullous rashes. She was referred to the dermatologists where she had skin biopsy with eventual diagnosis of Pemphigoid gestationis.

Management with oral corticosteroids led to resolution of symptoms albeit with scarring.



PEMPHIGOID GESTATIONIS..2

13.Diagnosis through skin biopsy and direct immunofluorescence test.

14. In 30-100% of cases the antibodies can also be detected by indirect IMF in the serum.

15. Commonest treatment modality is with potent and ultra-potent topical steroids, systemic steroids and sedative antihistamines.

16.Other treatment options include Intravenous immunoglobulin, azathioprine, dapsone, cyclosporine, plasmapheresis, pyridoxine, minocycline, immunoadsorption, nicotinamide, ritodrine, rituximab, doxycycline, erythromycin, methotrexate, and cyclophosphamide .

REFERENCES

1. Ambros-Rudolph CM. Dermatoses of pregnancy - clues to diagnosis, fetal risk and therapy. Ann Dermatol. 2011;23(3):265–275. doi:10.5021/ad.2011.23.3.265
2. Sävervall C, Sand FL, Thomsen SF. Pemphigoid gestationis: current perspectives. Clin Cosmet Investig Dermatol. 2017;10:441–449. doi:10.2147/CCID.S128144
3. Hallaji Z, Mortazavi H, Ashtari S, Nikoo A, Abdollahi M, Nasimi M. Pemphigoid gestationis: clinical and histologic features of twenty-three patients. Int J Womens Dermatol. 2016;3(2):86–90
4. Vanessa Ngan, Staff Writer, 2003. Updated: Dr Amy Stanway, Dermatologist, Tauranga, New Zealand. Copy edited by Gus Mitchell. September 2021, Pemphigoid gestationalis. Available online at <https://dermnetnz.org/topics/pemphigoid-gestationis>
5. Catherine Nelson-Piercy ;Dermatoses specific to pregnancy: Handbook of Obstetrics medicine 6th edition. CRC press 2020.
6. Iakov Shimanovich,Eva B. Bröcker,Detlef Zillikens, Pemphigoid gestationis: new insights into the pathogenesis lead to novel diagnostic tools. BJOG sept 2002 vol 109 pp 970-976

DISCUSSION

This case showed an unusual presentation of dermatoses in pregnancy. Isolated pruritus with characteristic rashes developing post natally- approximately 6 weeks from onset of pruritus.

This caused a delay in diagnosis and the first hint towards the possibility of autoimmune pemphigoid was severe IUGR and findings on the placental histology.

Presentation of PG varies from itching with hives or eczema like lesions to characteristically distributed bullous rashes. Mainstay of diagnoses is through histopathology features and **direct immunofluorescence (DIF) test** on skin biopsy. A case can be made for serology tests for D180 where no obvious cause of itching is identified.

Increased risk of recurrence means there should be clear advice regarding possible flares with menstruation or use of combined contraceptive pills. Also, due to extensive villitis in the case presented, the advice was to start oral corticosteroids in next pregnancy with multidisciplinary input from dermatologist, obstetricians and neonatologists.

CONCLUSION

Pemphigoid gestationis, albeit a rare condition should be considered in all cases of pruritus and vesiculo-bullous rash in a pregnant or postpartum patient.

There is need to reiterate the possibility of flare ups during ovulation, menstruation, use of COCP and in future pregnancies.

Steps should be taken to ensure a clear pathway of referral or communication between the obstetric and dermatology teams.

We advise early referral to the dermatologist to consider serology tests which may aid early diagnoses of PG in the absence of any skin lesions.