Pemphigoid Gestationis: A Case Series

Hengpoonthana R, Johansson C
Liverpool Hospital, SWSLHD, NSW, Australia

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Background: Pemphigoid gestationis is a rare autoimmune dermatosis which occurs in approximately 1 in 50,000 to 60,000 pregnancies.[1,2] Patients typically present with pruritic erythema in the periumbilical area which develops into papules, plaques, vesicles and bullae that may spread to the abdomen, trunk, legs and arms.[1] Diagnosis is made via skin biopsy with histopathology demonstrating perivascular lymphocytic infiltration and direct immunofluorescence showing linear C3 along the dermal-epidermal junction.[2]

Aims: This case series aims to raise awareness of pemphigoid gestationis and describe its diagnosis, management and complications.

Cases: We present three cases of pemphigoid gestationis. Diagnosis was confirmed on skin biopsy and the patients were treated with a combination of oral antihistamines, topical and oral corticosteroids, oral disease-modifying anti-rheumatic drugs, oral calcineurin inhibitors and intravenous immunoglobulin. Complications included gestational diabetes mellitus (GDM) requiring insulin secondary to oral corticosteroid therapy, preterm premature rupture of membranes (PPROM), preterm labour (PTL) and persistent postpartum pemphigoid gestationis.

Results: In the cases outlined, treatment in the antenatal and postnatal periods involved input from obstetric, dermatology and endocrinology specialties resulting in symptomatic relief, resolution of skin lesions and management of complications.

Discussion: Although rare, pemphigoid gestationis is an important differential diagnosis to consider in the assessment of rashes in pregnancy as this condition is associated with a recognised risk of complications requiring medical intervention and a multidisciplinary approach for optimal management.

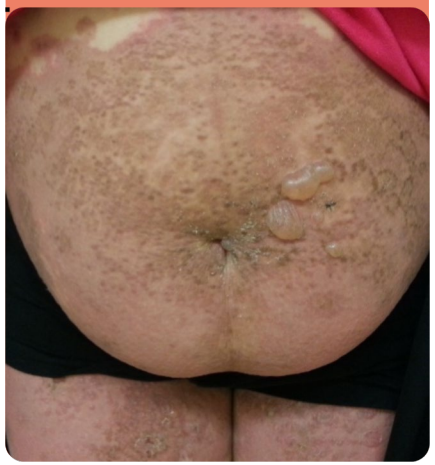
References

1. Pemphigoid Gestationis: Current Perspectives. Clinical, Cosmetic and Investigational Dermatology
2. Managing Pemphigoid Gestationis. European Medical Journal.

Case 1

Patient: 22 year old G2P1
P1 – uncomplicated pregnancy with spontaneous vaginal delivery of live 3.8kg baby at 38/40
Diagnosis: skin biopsy at 20/40
Complications: nil
Management:
- topical hydrocortisone and betamethasone, oral loratadine and dexchlorpheniramine, topical wet dressings, oral prednisone
- follow-up in high risk antenatal clinic with serial foetal growth and wellbeing ultrasounds, follow-up in dermatology clinic with postnatal prednisone weaning

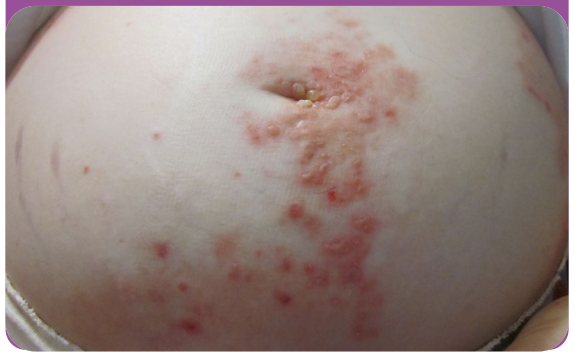
Outcomes: spontaneous vaginal delivery of live 3.6kg baby at 37/40



Case 2

Patient: 28 year old G2P1
P1 – uncomplicated pregnancy with emergency lower segment caesarean section for foetal distress in labour with delivery of live 2.9kg baby at 38/40
Diagnosis: skin biopsy at 31/40
Complications: GDM -> steroid-induced hyperglycaemia, PPROM, PTL, persistent postpartum pemphigoid gestationis
Management:
- topical hydrocortisone and betamethasone, oral loratadine, oral prednisone, topical wet dressings, subcutaneous insulin, intravenous ampicillin, oral erythromycin, 2x intramuscular betamethasone
- follow-up in high risk antenatal clinic with serial foetal growth and wellbeing ultrasounds, follow-up in endocrinology clinic with insulin titration, follow-up in dermatology clinic with monthly postnatal intravenous immunoglobulin infusion for 15 months

Outcomes: elective repeat caesarean section delivery of live 2.5kg baby at 36/40



Case 3

Patient: 27 year old G3P2
P1 – uncomplicated pregnancy with spontaneous vaginal delivery of live 2.9kg baby at 38/40 with suspected undiagnosed postpartum pemphigoid gestationis
P2 – pregnancy complicated with diet controlled gestational diabetes mellitus with suspected undiagnosed third trimester pemphigoid gestationis and spontaneous vaginal delivery of live 3.3kg baby at 39/40
Diagnosis: skin biopsy at 24/40
Complications: GDM -> steroid-induced hyperglycaemia, PPROM, PTL, peripartum pemphigoid gestationis flare
Management:
- topical hydrocortisone and betamethasone, oral promethazine, topical wet dressings, oral prednisone, subcutaneous insulin, intravenous ampicillin, oral erythromycin, 2x intramuscular betamethasone, oral cyclosporin
- follow-up in high risk antenatal clinic with serial foetal growth and wellbeing ultrasounds, follow-up in endocrinology clinic with insulin titration, follow-up in dermatology clinic with postnatal prednisone and cyclosporin weaning

Outcomes: spontaneous vaginal delivery of live 2.2kg baby at 33/40