



## Pemphigoid Gestationis: A Rare Disease

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### ABSTRACT

*Pemphigoid gestationis or herpes gestationis is a subepidermal blistering disease that occurs in women in the second or third trimesters of pregnancy or even during puerperium. Its incidence has been estimated at around 1 case in every 40,000 to 60,000 pregnancies. Pruritus is an important symptom associated with the onset of disease. The lesions usually arise on the abdomen, often involving the umbilicus, and spread centrifugally. Women with a history of pemphigoid gestationis are at higher risk of developing Graves' disease. Its clinical course is variable. The gold standard to diagnose pemphigoid gestationis is direct immunofluorescent of perilesional skin, which shows bright linear deposition of C3 along the basement membrane in 100% cases. The main differential diagnosis is polymorphic eruption of pregnancy (PEP) which occurs later in the third trimester of pregnancy. Treatment of pemphigoid gestationis starts involves application of topical corticosteroids and/or lotions.*

**Keywords:** pemphigoid, autoimmune, pruritis, pregnancy.

### INTRODUCTION

Pemphigoid gestationis or herpes gestationis is a subepidermal blistering disease that occurs in women in the second or third trimesters of pregnancy or even during puerperium. It is a rare skin disease whose incidence has been estimated at around 1 case in every 40,000 to 60,000 pregnancies. It is more common in patients with HLA DR3 and DR4 haplotypes. (Castro LA *et al*, 2006)<sup>[1]</sup> It is a rare autoimmune disease that most probably results from the breakdown of the protective immunity of the fetoplacental unit from maternal allogenic recognition, with auto-antibodies developed against two hemidesmosomal proteins BP180 and less frequently, BP230.

(IADVL Textbook of Dermatology)<sup>[2]</sup> Classically, PG presents as erythematous urticarial plaques that can subsequently develop into tense blisters. The periumbilical area is usually the first site involved. Pruritus is an important symptom associated with the onset of disease. (Jenkins RE *et al* 1999)<sup>[3]</sup>

### CASE REPORT

A 21 year old female gravida 1, nulliparous with 24 weeks period of gestation, with hypothyroidism since 8 years on tab Eltroxin 125 ugm OD, normotensive, non-diabetic came with complain of black pruritic papules on whole body since 3 days. Lesions first started from periumbilical

region, later, spread to whole body. There was no relevant family history and history of any medication. There was no history of oral contraceptive intake and there were no premenstrual flares. All routine investigations haemogram, liver function test, renal function test, thyroid profile (on treatment) were normal. Patient was diagnosed as Pruritic urticarial papules and plaques of pregnancy (PUPPD) now known as Polymorphic eruption of pregnancy (PEP). She was on tab dexchlorpheniramine, tab levocetirizine and mometasone ointment. Patient had no flares after the start of treatment. At 35 weeks 4 days of POG patient came with decreased fetal movement. Immediate ultrasound was done which showed:

single live intrauterine pregnancy with normal cardiac and somatic activity

cephalic presentation

BPD = 8.1cm=32week 5d,

FL=5.8cm=30week 2d,

Liquor adequate

Placenta upper posterior,

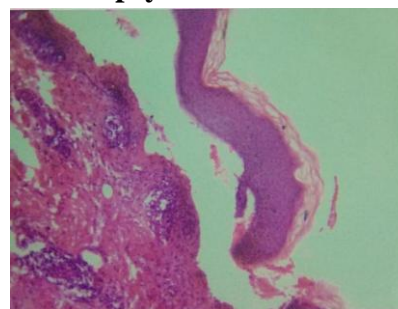
BPP=10/10 Non –stress test –Reactive.

Patient's relatives were explained about the risk of Intra-uterine growth retardation and prematurity and took their patient to higher centre for better neonatal care. Patient lost follow. Now again reported in OPD as gravid 2, para 1 live 0 at 8weeks 6 days period of gestation for regular ANC visit when she gave us history of her previous pregnancy, that, she delivered a single dead male baby two days going from Rajindra Hospital Patiala. On post-partum day 4, patient had blisters all over her body, associated with severe pruritus. Dermatologist opinion was taken. Skin biopsy was taken from lesion.

Skin biopsy showed a sub-epidermal blister with marked edema of the papillary dermis. There were perivascular and periadnexal inflammatory infiltrate in the dermis suggestive of Pemphigoid Gestanosis. Treatment with systemic steroids methylprednisolone 50 mg OD along with topical steroids was started. Later dose was gradually tapered. Patient showed improvement and was relieved of her symptoms.

During present pregnancy patient continued steroids throughout pregnancy and had a normal vaginal delivery on 22/7/2017 at 04:10 a.m. and delivered term live male baby with birth weight - 2700 gms and apgar 9, 9. Patient had no postpartum flares of pemphigoid during this pregnancy and was discharged in satisfactory condition.

#### Lesional skin biopsy HPE



Haemotoxylin and eosin staining

#### DISCUSSION

Pemphigoid gestationis may begin during the first or any subsequent pregnancy. It may develop between 9 weeks of gestation and one week postpartum, but most frequently it presents during the second and third trimesters. (Holmes RC 1983)<sup>[5]</sup> It may also be associated with hydatiform mole, trophoblastic tumors, and choriocarcinoma. Almost half of the cases develop during the first pregnancy (IADVL Textbook of Dermatology)<sup>[2]</sup>. Pemphigoid gestationis is an autoimmune blistering disease with autoantibodies of IgG1 and IgG4 subclass targeting BP 180 antigen and activate complement via classical pathway (Shornick JD. Dermatoses of pregnancy *et al* 1998)<sup>[5]</sup>. Women with a history of pemphigoid gestationis are at higher risk of developing Graves' disease. (Vaughan Jones SA 1999 *et al*)<sup>[6]</sup>. It is characterized with an acute onset of erythematous, urticarial papules and plaques that progress to tense vesicles and blisters, followed by severe pruritus. The lesions usually arise on the abdomen, often involving the umbilicus, and spread centrifugally. Recovery occurs generally in a few weeks after delivery but relapses are frequent in subsequent pregnancies.(Ambros-

Rudolph CM *etal* 2006)<sup>[7]</sup>, (Boulinguez S *etal*)<sup>[8]</sup>  
 The gold standard to diagnose pemphigoid gestanosis is direct immunoflourescent of perilesional skin, which shows bright linear deposition of C3 along the basement membrane in 100% cases. In addition, Ig G deposition can be seen in 25- 30 % of cases. Conventional indirect immunofluorescent demonstrates C3 binding to the basement membrane in over 90%of PG cases , but IgG is positive in less than 25 %. ELISA is more sensitive than indirect immunoflorescence. (IADVL Textbook of Dermatology)<sup>[2]</sup> Treatment of pemphigoid gestationis starts with application of topical corticosteroids and/or lotions to decrease itch. Antihistamines can also help to control itching. In patients with severe symptoms, systemic steroid therapy is needed. If these treatments are ineffective, plasmapheresis, cyclophosphamide, ciclosporin and dapsone are reported to be useful. However, their side-effects harm the mother and child. (Rare disease Pemphigoid gestationi Céline M J G Lardenoije *etal*)<sup>[9]</sup> Fetal prognosis is good, but early onset in 2nd trimester and blister formation are risk factors for prematurity and low birth weight. Rarely the newborn may be affected by transitory blisters. (Ambros-Rudolph CM *et al* 2006)<sup>[7]</sup> The increased risks small-for-gestational age babies and prematurity, suggests a low- grade placental insufficiency. (IADVL Textbook of Dermatology)<sup>[2]</sup>. The main differential diagnosis is polymorphic eruption of pregnancy (PEP) which occurs later in the third trimester of pregnancy. The skin eruptions in PEP usually begins at the striae, although vesicles may appear they almost never become bullous. In PG the neonate may have similar skin involvement as the mother, which is an useful distinguishing feature from PEP. Finally, immunofluorescence is the most definitive means for differentiating the two conditions because it is consistently negative in Polymorphic Eruption of Pregnancy.( Review on Pemphigoid Gestationi by Dr. K. H. Yeung)<sup>[10]</sup>

## CONCLUSION

Pemphigoid Gestationis is an autoimmune disease occurring almost exclusively during pregnancy. Its clinical course is variable but eruptions typically respond to steroid therapy. It is important to diagnose and treat Pemphigoid Gestationis early, not only to provide symptomatic relief to patients but to avoid fetal risks.

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