Walsall Hospitals NHS Trust

Pemphigoid Gestationis in Skin of Colour

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CASE

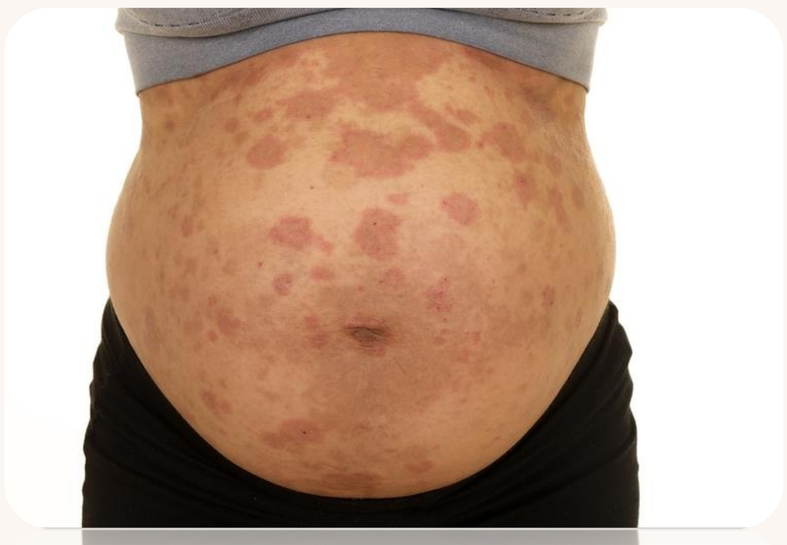
A 40-year-old, 22 weeks pregnant female presented with a pruritic rash, which began around the umbilicus. This evolved from small erythematous papules, to blisters and urticated plaques, which later spread to involve the abdomen, torso and limbs.
She was a multiparous woman of South Asian origin, with no significant dermatological history. Her first child had been born prematurely, and she was therefore taking aspirin.

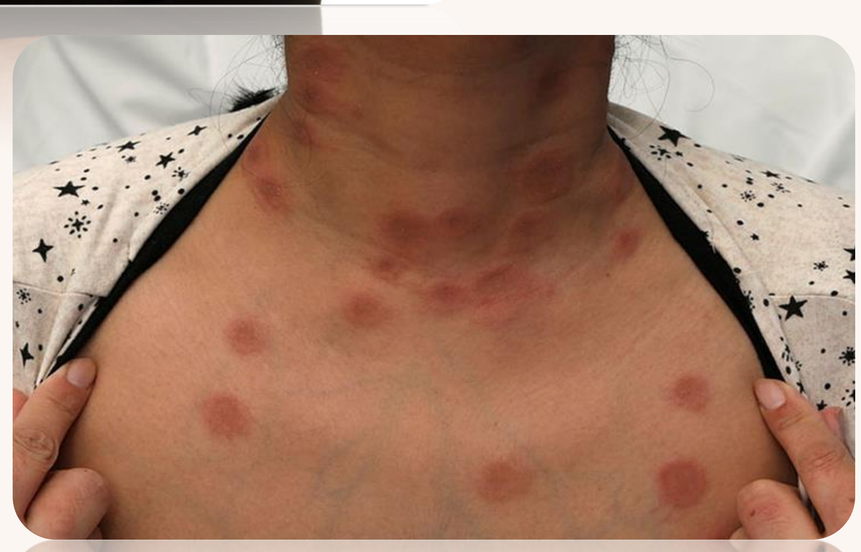
On examination she had widespread blisters and urticated targetoid plaques, affecting the body, palms and umbilicus. There was no mucosal involvement.

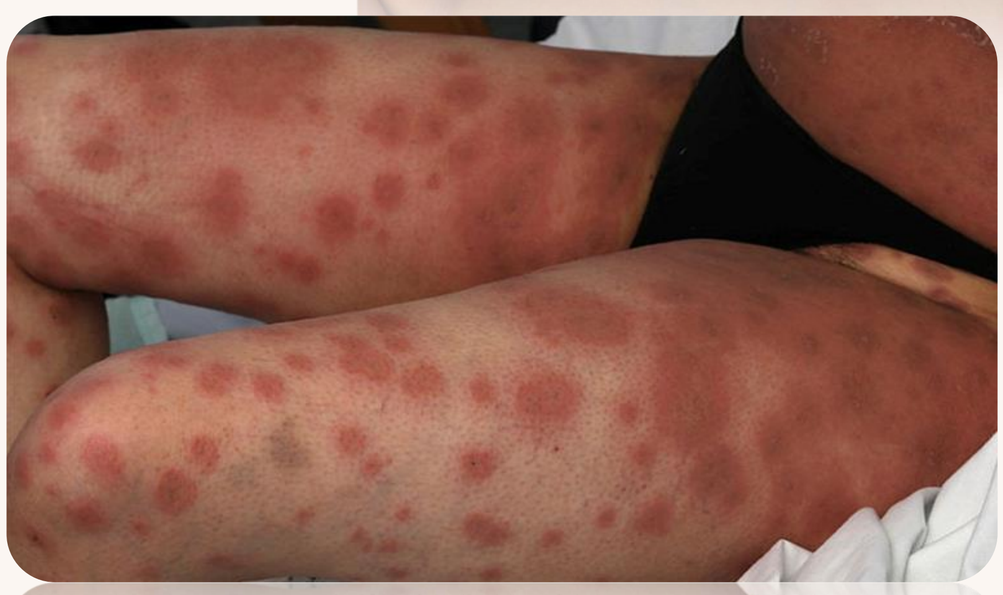
Biopsy demonstrated eosinophilic spongiosis of the epidermis, and dermal lymphoeosinophilic inflammatory cell infiltrate. Immunofluorescence was positive for IgG & C3 at the epidermal basement membrane. This was in keeping with the suspected diagnosis of pemphigoid gestationis.

She was managed with oral prednisolone, alongside topical steroids, emollients and antihistamines. The obstetric team arranged increased monitoring via regular ultrasound scans.

She gave birth prematurely at 34 weeks via emergency caesarean due to breech presentation and spontaneous rupture of membrane. The baby weighed 1.8kg and received 2 days of phototherapy for neonatal jaundice. Oral prednisolone was stopped 2 weeks prior to giving birth resulting in no major flare. Both mother and baby are well with the only remaining consequence being post-inflammatory hyperpigmentation in the mother.







DISCUSSION

Gestational pemphigoid is a rare subepidermal autoimmune blistering condition of the skin affecting pregnant women[1]. The term ‘herpes gestationis’, now known as gestational pemphigoid, was first coined by John Laws Milton in 1872 due to its similar appearance to herpetiform lesions[2]. It occurs in roughly 1 in 60,000 pregnancies due to the presence of autoantibodies against the hemidesmosal proteins BP-180 and BP-230 found in the basement membrane, resulting in subepidermal blisters[3].

Typically, it presents in late pregnancy or immediately postpartum[4], making our case unusual to present so early and with such severity. The main symptom is severe pruritis with a rash typically beginning around the umbilicus before spreading to the trunk and limbs, with associated plaques and blisters. There is no mucosal involvement.

Diagnosis can be made through a combination of clinical features, histological examination and immunofluorescence. Histological examination will demonstrate subepidermal vesicles, direct immunofluorescence will show linear complement deposition along the basement membrane in all cases, and indirect immunofluorescence microscopy may show circulating IgG in the blood[5].

The mainstay of treatment is topical/oral steroids and antihistamines[5]. The most common complications are prematurity and foetal growth restriction, hence MDT management between dermatology and obstetrics is vital to ensure safety of both mother and child.

There is emerging evidence correlating the severity of bullous disease to darker skin types. A recent study aimed to associate the effects of Fitzpatrick skin colour on the severity of bullous pemphigoid[6]. It found that severity scoring systems were higher in black patients compared to white patients, and more extensive disease correlated with darker skin types. Despite that, the urticaria and erythema scores were interestingly found to be lower in darker skin types. This illustrates the possible underscoring of bullous conditions in those with darker skin.

Further studies are needed to produce a validated scoring tool that can be used effectively and reliably in all skin types. We also propose more research is needed to evaluate if bullous conditions present with more severe disease in darker skin types.

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