Case for diagnosis*

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CASE REPORT

A 27-year-old woman at 28-weeks gestation presented with a widespread, pruritic eruption of macular, confluent lesions with tense vesicles and some blisters in the arms and thighs (Figure 1). The lesions initially presented at 26 weeks of gestation on the legs and spread to the abdomen, arms, and back. Past medical history included one prior abortion due to sickle-cell disease, without any history of similar symptoms. The patient had been previously treated with methylprednisolone cream and oral cetirizine, with persistence of the skin lesions. A cutaneous biopsy was performed in lesional skin, showing the presence of multiple vesicles in the dermal-epidermal junction, filled by serosity and eosinophils. In the underlying dermis, a marked edema outlined a dermal-epidermal detachment, with a dense inflammatory infiltrate (predominantly with eosinophils) extending to the dermis (Figure 2). Direct immunofluorescence in perilesional noninvolved skin showed linear deposists of C3 at the basement-membrane zone (Figure 3).

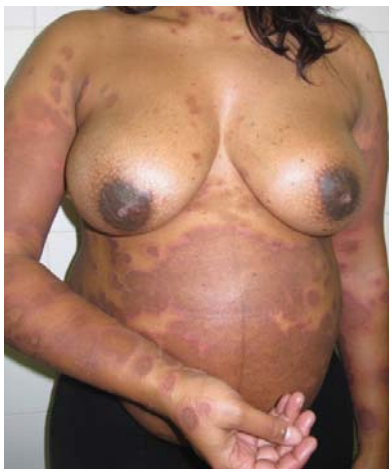

FIGURE 1: Macular, confluent, pruritic lesions in the abdomen and arms of a 26 weeks pregnant woman

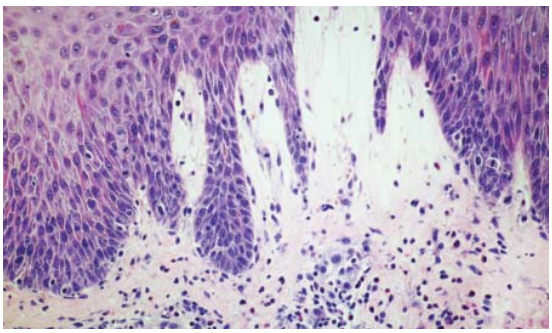

FIGURE 2: Cutaneous biopsy in lesional skin revealed a marked edema that outlined a dermal-epidermal detachment, with a dense inflammatory infiltrate (predominantly with eosinophils) extending to the dermis

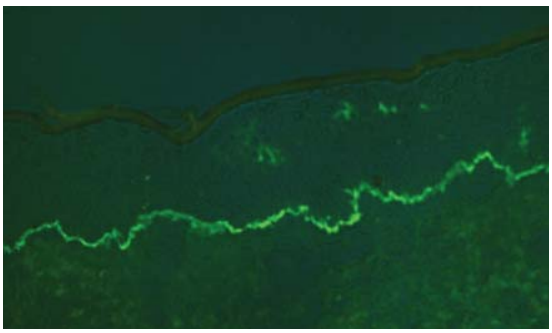

FIGURE 3: Direct immunofluorescence in perilesional noninvolved skin showed linear deposists of C3 at the basement-membrane zone

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DISCUSSION

Pemphigoid gestationis is a rare, autoimmune blistering dermatosis of pregnancy, with an incidence ranging up to 1:50.000~60.000 pregnancies depending on the prevalence of the HLA-haplotypes DR3 and DR4.[1,2] PG typically develops in the second or third trimester of pregnancy, with an abrupt onset, but may appear any time during pregnancy or even in the immediate postpartum period. Severe pruritus is followed by the appearance of erythematous, urticarial papules and plaques that progress to tense vesicles and blisters. The lesions usually arise on the abdomen, often involving the umbilicus, and spread centrifugally, sparing face, palms, soles and mucous membranes (< 20% cases). Flares have been observed at or immediately after delivery1 , pre-menses and with the use of oral contraceptives (25% of patients).[3-5]

The criteria for the diagnosis for PG include an appropriate clinical presentation and specific histologic findings of a subepidermal blistering process and a linear C3 deposition along the basement membrane in direct immunofluorescence, with or without deposition of immunoglobulin G (20-25% of cases).

Treatment depends on the severity of the disease and aims to prevent blister formation and control pruritus. Mild cases may be treated with topical corticosteroids and oral antihistamines.[1,4] Potent topical glucocorticoids, oral corticosteroids (prednisone 0.5~1 mg/kg/day), and oral antihistamines are reserved for more serious cases.[6]

In our case, clinical suspicion of PG was confirmed by histological and direct immunofluorescence findings and systemic treatment with methylprednisolone (0,5mg/Kg/day) was initiated during the pregnancy with gradual clinical improvement, despite a relapse after a first attempt to reduce the dosage, resulting in extension of the systemic treatment until delivery and 6 weeks after. A healthy, asymptomatic male infant was born without cutaneous lesions. To date the patient has not reported a flare with her menses. The present case corroborates the importance of a timely clinical and histopathological diagnosis of PG, thus preventing or minimizing the risk of adverse effects for the fetus. An interdisciplinary approach is also of crucial importance for the benefit of the pregnant woman and her pregnancy, and also for the infant, as well as during the postpartum period.[7] ❑

Abstract: Pemphigoid gestationis is a rare, autoimmune blistering dermatosis of pregnancy. No increase in fetal or maternal mortality has been demonstrated, but a greater prevalence of premature and small-for-gestational-age babies has been reported. Topical and systemic corticosteroids and antihistamines are the manstay of treatment. The authors report a case of a 27-year-old woman at 28-weeks gestation with sudden onset of pruriginous vesicles and blisters in the abdomen and limbs. Systemic corticosteroids were introduced and maintained throughout gestation to prevent flares and tapered after the birth of a healthy child.
Keywords**:** Pregnancy; Pruritus; Skin diseases, vesiculobullous

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