CASE REPORT

Pemphigoid Gestationis (Herpes Gestationis)

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Introduction

During pregnancy, alterations in the appearance of the skin are common, ranging from normal cutaneous changes to changes that are pathological, recurrent, and specific to pregnancy.

Pemphigoid gestationis (PG) is a specific dermatosis, which is a rare, pruritic, polymorphic, inflammatory, sub-epidermal, bullous dermatosis of pregnancy, and the post partum period. The incidence of PG is roughly estimated to be 1 in 10,000–60,000 deliveries [1, 2]. A definitive diagnosis can be made only with specific immunopathological studies [3].

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Case Report

A 26-year-old pregnant female (second gravida) presented with itchy skin lesions of 18 days' duration in her third trimester. Her first pregnancy and delivery were uneventful except that a caesarean section was performed as she was a case of post-polio residual paralysis.

On dermatological examination, there were multiple discrete hyperpigmented and skin colored, intensely pruritic annular papules, and plaques of varying sizes. The periphery of these lesions was studded with multiple tiny papules and papulovesicles. The lesions started on the abdomen and gradually involved all the four extremities including palms and soles. Face, scalp, and mucosa were spared. Initially, we treated her with systemic antihistamines and topical steroids (clobetasol propionate). (Fig. 1a–d) Though her general condition remained stable, itchy lesions continued to appear.

With the provisional diagnoses of pemphigoid gestationis, pruritic urticarial papules and plaques of pregnancy (polymorphic eruption of pregnancy) and erythema multiforme, skin biopsies were taken. Tissue from the periphery of annular plaque was sent for histopathological studies. Tissue from perilesional normal looking skin was sent for immunofluorescence. Routine blood and urine examinations were within normal limits. Serological tests for syphilis and HIV were negative, ANA was negative, and thyroid function tests were normal. The Tzanck smear from one of the vesicles showed plenty of lymphocytes and eosinophils, strengthening the possibility of PG, and we put the patient on 40 mg

Fig. 1 a–d Hyperpigmented and skin colored, annular papules and plaques

prednisolone. Dramatic improvement was observed within 3 days. Histopathology report: Epidermal spongiosis and marked perivascular edema, infiltration by lymphocytes, histiocytes, and eosinophils in the superficial dermis, suggestive of PG. (Fig. 2a–c) The Direct Immuno Fluorescence (DIF) report (the gold standard investigation) was consistent with the diagnosis of PG, as the tissue showed linear deposits of C_3 at the basement membrane zone of the normal appearing skin. (Fig. 3) Thus, the diagnosis of pemphigoid gestationis was confirmed. She underwent caesarean section at term and gave birth to a normal healthy male baby. Steroid was tapered off and withdrawn within 3 months. She remains symptom free after 12 months of cessation of treatment (Fig. 4a, b).

Discussion

PG usually presents as an extremely pruritic papulovesicular eruption on the abdomen, but may involve other areas, including the palms, soles, chest, back, and face. Mucosal surfaces are usually spared. Lesions may vary from erythematous, edematous papules to large, tense bullae. PG usually begins from the fourth to the seventh month of pregnancy. Despite being a self-limiting condition, after delivery, PG can get exacerbated during the first few menstrual cycles or by the use of oral contraceptives, or during subsequent pregnancies [3].

The etiology of PG is unknown, but there is an increased association with other autoimmune diseases [4–6] and HLA

antigens: B8, DR3, and DR4 [7, 8]. The PG antigen in 180 kD antigen (BPAG2) is present in the hemidesmosomes of the basement membrane [9]. Antibasement membrane zone (BMZ) antibodies, mainly IgG1 are pathogenic [10]. The antigenic trigger for their production is probably paternally derived since PG can occur in association with trophoblastic tumors hydatiform mole, and choriocarcinoma in which the chromosomes are paternally derived [11]. Aberrant expression of paternal MHC class II molecules in the placenta triggers off a maternal immunological response to placental (amnion) BMZ [12]. Cross-reaction with the skin is responsible for the cutaneous lesions [13]. Complement activation by antibodies deposited at the BMZ and subsequent inflammation results in basal cell degeneration and dermoepidermal separation [14].

In zones of erythema and edema, there is a perivascular infiltrate composed of lymphocytes and eosinophils. There is marked papillary dermal edema, spongiosis which may be eosinophilic in type [15]. DIF testing reveals linear deposition of C_3 in perilesional skin in virtually all patients. IgG is similarly deposited in 30–40 % [15].

Pruritic Urticarial Papules and Plaques of Pregnancy (PUPPP) is the disease most often confused with PG, but in PUPPP, there are no immunoreactants seen at the dermal-epidermal junction [3]. Dermatitis herpetiformis and erythema multiforme can be differentiated from PG by routine DIF.

The aim of treatment is to suppress blister formation and to relieve the intense pruritus. These aims can be achieved by giving 20–40 mg of prednisolone per day.

Fig. 2 a–c Epidermal spongiosis, perivascular edema, infiltration by lymphocytes, histiocytes, and eosinophils in the superficial dermis

Fig. 3 Linear deposits of C₃ at the basement membrane zone

The prednisolone is tapered gradually during the post-partum period, but exacerbations at the time of menstruation may demand a temporary increase in dosage. However, a few patients can be managed with antihistamines and topical steroids or emollients, but at the other extreme, some after parturition, require azathioprine in addition to prednisolone for control of their disease [3].

Infants born to affected mothers who have received high doses of prednisolone should be examined by a neonatologist for evidence of adrenal insufficiency. The cutaneous lesions noted in some infants are of transient nature requiring no therapy [3].

Fig. 4 a, b Post-inflammatory hyperpigmentation only

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