

Gestational pemphigoid

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Abstract

Pemphigoid gestationis is a rare, autoimmune blistering dermatosis. It is unique in that it is diagnosed primarily in association with pregnancy.

Autoantibodies against placental BP180 (also known as BPAG2 or collagen XVII) cause damage to the skin basement membrane, resulting in severe itching and blistering rash over the body and the extremities.

Keywords: pemphigoid gestationis, pregnancy

Introduction

Gestational pemphigoid (pemphigoid gestationis, PG) is a rare autoimmune skin disorder occurring characteristically during pregnancy.

It is a group of immune-mediated bullous disorders, which often cause blisters and extensive lesions of the skin. During pregnancy, its condition will become more complicated due to the change in the mother's hormone level and the effect of drug therapy on both the mother and her fetus.

We report a case of a pemphigoid gestationis in a 25-year-old woman.

Case report

A 25-year-old woman presented to our clinic at 32 weeks gestation with an intensely pruritic, bullous eruption of 10 days duration (Fig. 1). The patient was referred from obstetrics for her rash which was persistent despite treatment with a topical mid-potency steroid. It was her first

pregnancy. Physical examination revealed numerous erythematous, urticarial vesicles, bullae and pustules which began on the abdomen and rapidly spread to involve the extremities (Figure 1). No facial or mucous membrane involvement was appreciated. Given the presentation, a presumptive diagnosis of pemphigoid gestationis was appointed. Biopsies and immunofluorescence were taken. They revealed a subepidermal blistering dermatosis with perivascular and interstitial infiltrates of eosinophils and lymphocytes. Direct immunofluorescence (DIF) demonstrated linear deposits of complement component C3 along the basement membrane zone. These biopsies confirmed the diagnosis of pemphigoid gestationis.

The patient was put on dermocorticoides 30g per day with progressive decreases after improvement.

At 39 weeks gestation during an uncomplicated vaginal delivery, the patient delivered a healthy baby without any cutaneous involvement.



Fig 1: Numerous erythematous, urticarial vesicles, bullae and pustules which began on the abdomen and rapidly spread to involve the extremities

Discussion

Pemphigoid gestationis was first described in 1872 under the name "herpes gestationis". This was an elegant initial description that encompassed both the morphology of the skin lesion and the state of the patient in which the eruption occurred [1].

Studies looking for the epidemiology of PG are rare. Population-based studies have reported an annual incidence ranging between 0.5 and 2.0 cases per 1 million people in France, Kuwait [2] and Germany [3]. Based on the current epidemiological data PG is estimated to occur in one out of about 40,000-50,000 pregnancies [4] with no difference in racial distribution [5, 6]. Single cases have been described in association with molar pregnancies [7] and trophoblastic tumors [8].

Pemphigoid gestationis is a distinct disease of pregnancy which typically occurs late in gestation or occasionally in the immediate postpartum period. It is characterized by an intensely pruritic, vesiculobullous eruption which begins on the central abdomen and spreads centrifugally to involve the extremities. The face, mucous membranes, palms, and soles are usually spared. The differential diagnosis includes: bullous pemphigoid, drug eruption, and allergic contact dermatitis among others [9].

The diagnosis of PG is preferably made by a dermatologist, but all physicians treating pregnant women, general practitioners and obstetricians, should be able to consider PG. A biopsy for histopathology is not needed; the diagnosis is based on clinical picture, direct immunofluorescence microscopy and serology [10, 11].

Due to the rarity of PG, no randomized studies have been published and treatment recommendations are based on clinical experience and studies from treatment of other skin diseases. PG symptoms can be quite debilitating, but the condition does not constitute a direct health risk to the mother. When choosing a treatment, the benefit of the medication to the mother is critically weighed up against possible risks to the fetus. The aim of the treatment is to suppress the excessive itching and to prevent formation of new blisters [12].

Topical steroids and antihistamines may work in mild cases but are generally considered to be ineffective. Systemic steroids are the mainstay of therapy. Most patients respond to prednisone 0.5 mg/kg daily in divided doses. The dose is then tapered and occasionally discontinued as gestation progresses. Due to frequent postpartum flares, therapy can be increased or resumed after delivery [9].

Infants born with active disease usually resolve spontaneously in the first few days of life and do not require treatment. The consultation of a pediatrician is necessary to monitor for adrenal insufficiency in infants exposed to steroids throughout gestation [9].

Conclusion

In conclusion, pemphigoid gestationis is an interesting, rare autoimmune blistering skin disease.

The severe itching and blistering caused by the disease can be quite debilitating. The diagnosis of PG is made in a specialized care setting at a dermatology department. Since PG is associated with a risk of prematurity and fetal growth restriction, pregnancy monitoring by an obstetrician is recommended. Mothers with PG should be informed of the natural course of the disease, good fetal prognosis, the

Possibility of relapses after delivery, and the risk of relapses in subsequent pregnancies and with hormonal contraception.

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