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Case Study

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PEMPHIGOID GESTATIONIS: DIAGNOSIS, FETAL PROGNOSIS AND TREATMENT: A CASE REPORT

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ABSTRACT

Pemphigoid Gestationis (PG) is a rare autoimmune dermatosis related to pregnancy, with an incidence of 1 in 20,000 to 50,000 pregnancies. it usually appears during the second and third trimesters, but can also occur at any stage of pregnancy and postpartum. Clinical manifestations of PG include intense pruritus, papules and annular plaques followed by vesicles and finally large, tense, bullae on an erythematous background that typically appear periumbilical and spread to the trunk and extremities. The diagnosis of PG is based on a deep clinical evaluation, the histological findings, direct and indirect immunofluorescence and the measurement of serum levels of anti BP180 antibodies using ELISA. PG can have an impact on pregnancy by increasing the risk of intrauterine growth restriction, prematurity and obstetric complications. We report the case of a 36-year-old multigravida, diagnosed with Pemphigoid Gestationis, in week 26 of gestation. The patient was treated successfully and delivered a healthy child without any symptoms of congenital pemphigus.

KEYWORDS: Pemphigoid Gestationis, Autoimmune, Pregnancy, Dermatosis.

INTRODUCTION

Pemphigoid Gestationis is a rare autoimmune blistering skin disorder that affects pregnant women.[1] Although the precise pathogenesis of PG remains unclear, it is considered an autoimmune disorder that involves the production of autoantibodies against bullous pemphigoid antigens 180 and 230.[2] These autoantibodies cause significant damage to the skins basement membrane, leading to noticeable pruritus and blisters on the abdomen and extremities.[2]

The aim of this report is to analyze the epidemiological and clinical aspects of PG, and to highlight the importance of early therapeutic management and fetal monitoring during pregnancy to avoid potential complications.

CASE REPORT

Mrs. NH, 36 years old, multigravida, mother of two children by c section, with no known comorbidities or family history of autoimmune diseases, admitted to our hospital at 26 weeks of gestation for skin Rash associated with intense pruritus, the patient reported that the skin lesions first appeared in the periumbilical area, which later spread to hands, forearms, feet and lower legs. the clinical examination objectified annular erythemato-papular lesions, with vesicles and tense bullae in some places, associated to multiples vascular lesions in rosette all over the body, sparing the face, scalp, palms and soles. obstetric examination was normal, with an obstetric ultrasound corresponding to the term. the fetal heart rate recording didn't show any anomalies. Complete blood tests were all within normal limits. The diagnosis was based on a skin biopsy with direct immunofluorescence and serum level of antibodies using ELISA. The direct immunofluorescence of the biopsy sample indicated the presence of linear deposits of component C3 along the basement membrane. The patient was daily treated with 0.5mg/kg of oral prednisone and local corticosteroid therapy with good clinical remission. The patient had an elective caesarean section at 38 weeks of gestation, giving birth to a male infant without any skin lesions. The patient didn't have any recurrence during the postpartum.

DISCUSSION

Gestational pemphigoid was first described in 1872 under the name Herpes Gestationis, because of the morphology of the skin lesion.[1] later, the similarity between bullous pemphigoid and herpes Gestationis became clearer, which prompted the transition in nomenclature to gestational pemphigoid.[3]

Gestational pemphigoid is a pruritic dermatosis that appears during pregnancy, where pruritic urticarial papules and plaques are the primary lesions that transform into blisters.[3,4] the lesions often start from the periumbilical area and extend to flexion areas[4], but in our case, the lesions began in the trunk and limbs, then spread to the entire body. Recommended tests include direct immunofluorescence and liver function tests to exclude intrahepatic cholestasis of pregnancy, in our patient case, liver function tests were normal.[5]

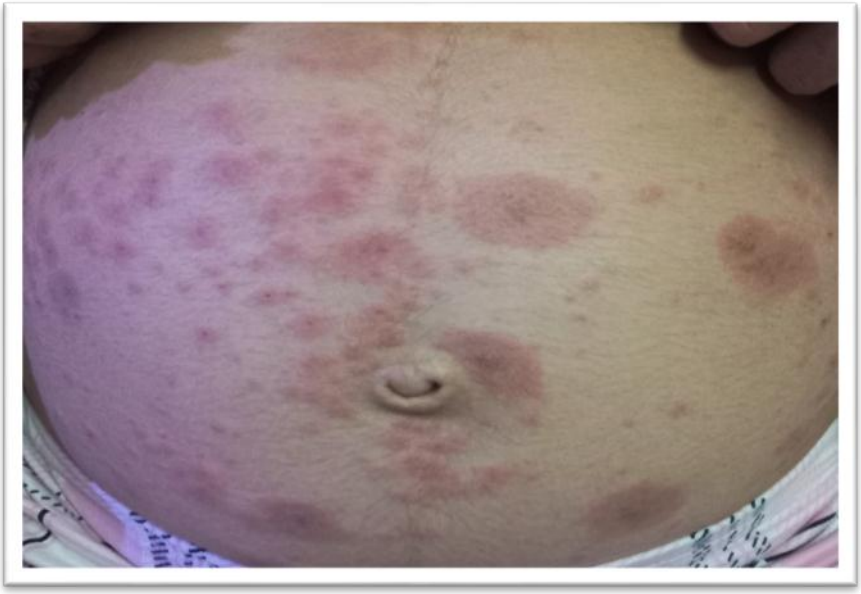

Figure 1: Periumbilical eruption typical of PG.

Direct immunofluorescence confirms the diagnosis by demonstration eosinophilic infiltrates and a linear deposit of immunoglobulin G (IgG) and complement C3 along the basement membrane.[6,7] In our patient's biopsy, a linear deposit of IgG and complement C3 was identified. Direct immunofluorescence and the search for circulating antibodies are consistently negative in other pregnancy related dermatoses.[7]

PG can cause some fetal complications, including miscarriage, preterm birth, low birth weight, and transitory urticarial lesions or blisters,[8] that may appear in the newborn as a result of circulating maternal IgG antibodies crossing the placenta, but this occurs in fewer than 5% of cases. in our patient case, no intrauterine growth retardation was observed during the pregnancy monitoring.[8,9]

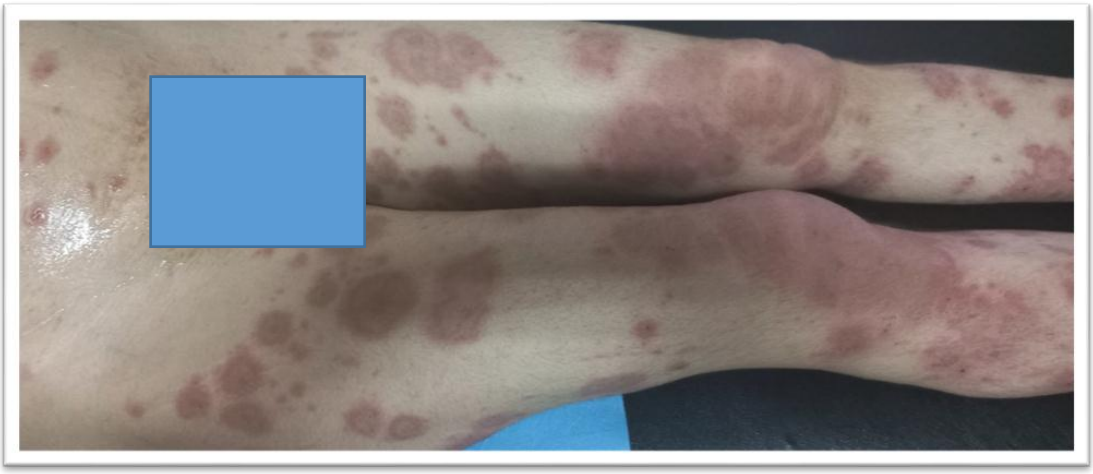

Figure 2: Inflammatory annular plaques on an erythematous background on the thighs and legs.

The goal of the treatment is to relieve symptoms, especially pruritus, control, or even suppress cutaneous signs, while avoiding any local or systemic complications. systemic corticosteroids remain the cornerstone of treatment.[10,11] Topical corticosteroids can be used to treat benign and moderate forms of PG. Prednisolone, at a dose of 0.3 to 0.5 mg/kg allows a rapid control of the diseas in most cases. we reserve higher doses (up to 1mg/kg) for very severe forms of the disease. Our patient was treated with systemic corticosteroids due to the extent of the lesions, associates to local corticosteroids. Immunosuppressant, especially

in the postpartum period, are reserved to the forms of the disease that are resistant to corticosteroid therapy.[12]

Pregnancy should be considered a high risk pregnancy, that needs close monitoring in a specialized obstetric environment. dermatological monitoring is recommended weekly at the beginning and then spaced according to the evolution.[13] potential local and systemic side effects of topical corticosteroids should not be overlooked. in our case, the patient didn't present any complications or side effects related to treatment, the patient gave birth to a healthy baby by a cesarean section. No skin lesions were detected after the delivery. Skin manifestations associated with PG typically regress within 2-6 weeks after delivery. the risk of recurrence in subsequent pregnancies is high, with more rapid and severe antibody accumulation and also when using hormonal contraception.[14]

CONCLUSION

The diagnosis of gestational pemphigoid can be challenging because of its rarity and its similarities with common skin lesions. Early identification and appropriate treatment of gestational pemphigoid is important to prevent complications in both the mother and the fetus. Therefore, it is crucial to establish a comprehensive collaboration between both gynecologists and dermatologists.

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