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Pemphigoid Gestational and the Importance of Differential Diagnosis: A Case Report

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Abstract

Pemphigoid Gestational (PG), an uncommon autoimmune bullous dermatosis of pregnancy, stands out between the specific dermatological diseases of the gestational period. Despite being self-limited and benign, it has a significant impact on women's health, both physiologically and psychologically. In addition, it promotes pathological changes that can be complicated during pregnancy. This paper reports the case of a pregnant patient with blistering lesions throughout the whole body. The diagnostic hypothesis initially involved PG and the specific treatment with corticosteroids was initiated, but after the anatomopathological result of the biopsy there has been diagnostic confusion with drug hypersensitivity, since clinical and histological manifestations were exuberant for both pathologies. As the lesions recurred in the puerperium, due to the corticoid abstention that was administered for the treatment of the differential diagnosis, a diagnostic review with immunofluorescence was made and confirmed PG. It is important to establish a correct diagnosis therefore, in order to improve the quality of care offered to the mother-baby binomial.

Keywords

Pemphigus, Pregnancy, Dermatology

1. Introduction

Pregnancy is related to several changes in the female organism, some physiological and others pathological. Pemphigoid Gestational is an uncommon autoimmune disorder with vesico-bullous and intensely pruritic manifestations between the dermatological pathologies. It also has a self-limited evolution and commonly occurs in more advanced periods of the pregnancy.

This case report is discussed clinical aspects of the pathology and is related to the case of an affected patient. The importance of differential diagnoses and an adequate investigation is also highlighted. The description is based on the rarity of the case, in order to establish greater understanding about the pathology.

2. Case Report

Female patient, 42 years old, 36 weeks pregnant, with a previous vaginal delivery and c-section, seeks out care in the emergency department of a hospital. She had erythematous lesions, some of them bullous, on the back, upper and lower limbs. The eruptions progressively grew over a month on the right thigh. She also had intense local itching. The lesions spared the face and the mucous membranes.

On admission, the patient was using antibiotic therapy, previously prescribed by another health service. The management was probably taken considering the suspicion of impetigo, even in the absence of signs of infection. As soon as Gestational Pemphigoid was considered as a diagnostic hypothesis, corticosteroid therapy was started, antibiotic therapy was immediately discontinued and a consultation was requested from dermatologist.

Afterwards, a skin biopsy was performed. Besides diagnostic investigation, the pregnant woman was managed with parenteral corticosteroids and antihistamines to control symptoms. The patient was discharged from the hospital with an interview to the dermatologist in seven days, and she was instructed to use Prednisone 1 mg/kg daily.

The patient went to the Obstetric Center of the same service, before the scheduled appointment, because her water broke. At this moment, the skin lesions were in remission, with hyperchromic scars evidenced in the previous topography. C-section happened with 37 weeks and 6 days because the bag was ruptured with meconium amniotic fluid and the baby was in a breech presentation. The newborn showed vitality and no sign of bullous lesions.

The anatomopathological result of the biopsy indicated, during the hospitalization, an etiology probably related to a skin condition of hypersensitivity to drugs, instead of Pemphigoid Gestational, since an inflammatory process with an infiltrate of lymphocytes in blood capillaries of the upper dermis, eosinophils, extravasated erythrocytes, mild edema and epidermal spondylosis were founded. Therefore, corticosteroid therapy was immediately interrupted. In 48 hours postpartum, the patient was discharged, with referral for follow-up at a medical specialty center with a diagnosis suggestive of a hypersensitivity reaction.

In the puerperal follow-up, the dermatologist saw that the patient had lesions recurrence, since she has been abstinent from glucocorticoid therapy. This episode shows to be the contrary of the classic manifestations of hypersensitivity to drugs, which should recur when the medication is discontinued. Furthermore, PG lesions more commonly improve over longer periods, weeks to months, in the natural course of the disease after birth. Thus, the recurrence of lesions 48 hours postpartum was compatible with PG. It has been anatomopathological reavaliated

and corroborated by immunofluorescence. In this exam, nonspecific linear fluorescent deposits of C3 were identified in the basement membrane of the epidermis, definitively confirming the diagnosis of Pemphigoid Gestational (Figure 1 and Figure 2).

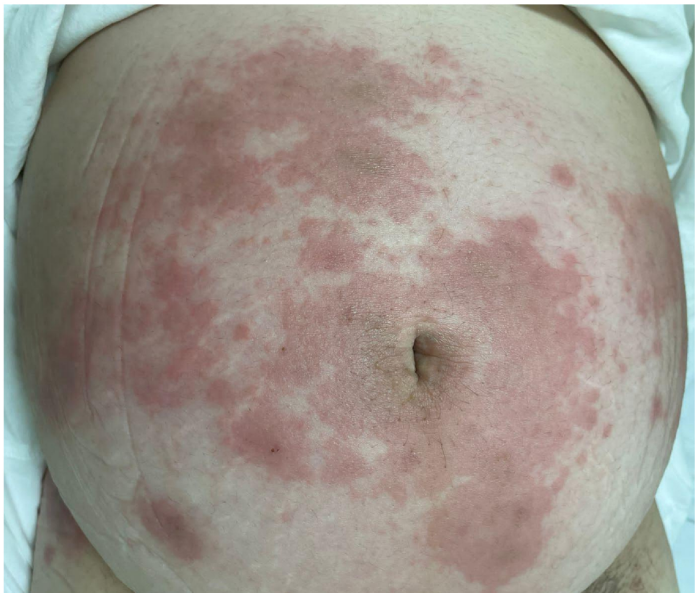

Figure 1. Urticarial lesions on the trunk.

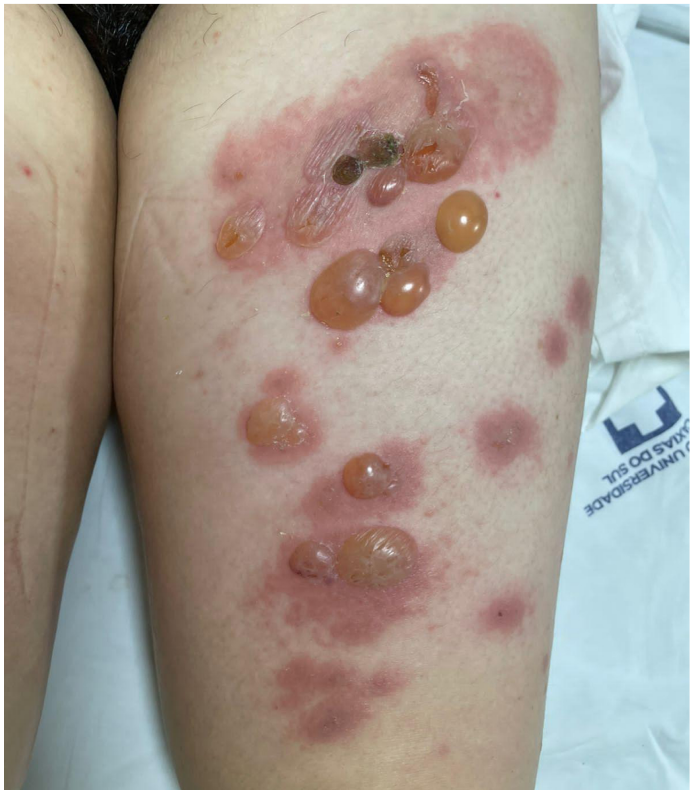

Figure 2. Image of abdomen (1) and left thigh (2), on first hospital admission.

3. Discussion

Pemphigoid Gestational, also known as Herpes Gestational, is a rare dermatosis [1] [2] [3] [4]. It is an autoimmune vesicobullous pathology associated with pregnancy [1] [2]. It occurs mainly in the second and third trimester of pregnancy [1] [2] [3] [4]. In the same way, the patient reported in the case presented her condition in the third gestational period.

It presents with intense skin itching [1] [2] [3] [4], associated with polymorphic papular or vesicular-erythematous deficiencies, which progress to generalized bullous eruptions [1] [2] [3]. After rupture, the blisters may leave ulcerated areas with melicero-hematic crusts and subsequent healing with residual hyperpigmentation [1] [2].

Usually, the involvement occurs initially in the periumbilical region, with dissemination to the extremities, back and thorax, sparing mucous membranes and face [1] [2] [3] [4]. Differently from what is described in the literature, the patient reported in this study started with lesions in the right lower limb, with subsequent dissemination to other areas. In addition, the pathology has a self-limiting character, with spontaneous regression after weeks or months of postpartum [3].

There is also a tendency towards a higher occurrence of prematurity, low neonatal weight and stillbirths [2] [3] [4] due to placental insufficiency associated with Pemphigoid Gestational [4]. Fortunately, the delivery of the patient reported was without difficulties or harm to the newborn, as well as the absence of similar lesions on her skin. Thus, it is described in the literature that there is a small occurrence of bullous lesions in the newborn of an affected mother, and in those who do, remission occurs without sequelae [2].

The diagnostic method of the pathology is based on a skin biopsy of the affected region, with visualization of sub-epidermal blisters on anatomopathological exams, and direct immunofluorescence [4]. The presence of edema in the papillary dermis is indicative of the diagnosis, in addition to an inflammatory infiltrate composed of lymphocytes, eosinophils, histiocytes and some neutrophils in the superficial and deep perivascular region. In addition, the dermis demonstrates spongiosis and basal cell necrosis in the dermal papilla type [1] [2]. In immunofluorescence, there are linear depositions of immunocomplexes on the basement membrane, being IgG, IgA and complement C3 [1] [2] [4].

Among the differential diagnoses, attention should be paid to pruritic urticarial papules and plaques of pregnancy, acute urticaria, and drug-induced bullous eruptions, scabies, and other dermatoses [1] [3]. The patient presented diagnostic confusion with drug hypersensitivity, given the clinical similarity between the pathologies, and a diagnosis of Pemphigoid Gestational was established after the natural evolution of the pathology and review of the investigative methods.

Finally, the proposed treatment is based on corticosteroid therapy and symptomatic drugs. In other words, topical or systemic corticosteroids, depending on the severity of the condition, with Prednisone being the main representative, and antihistamines for pruritus [1] [2] [3] [4], as it was made in the case.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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