Pustular psoriasis of pregnancy (Impetigo herpetiformis) - Case report*

Psoríase pustulosa da gestação (Impetigo herpetiforme) - Relato de caso

Rogerio Nabor Kondo[1]
Fernanda Mendes Araújo[2]
Allamanda Moura Pereira[2]
Vivian Cristina Holanda Lopes[3]
Ligia Márcia Mario Martins[4]

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Abstract: Impetigo herpetiformis is a rare dermatosis of pregnancy with typical onset during the last trimester of pregnancy and rapid resolution in the postpartum period. Clinically and histologically, it is consistent with pustular psoriasis. This similarity has led some authors to name the disease "the pustular psoriasis of pregnancy". We report the case of a patient who developed impetigo herpetiformis in two sucessive pregnancies.
Keywords: Case reports; Impetigo; Pregnancy; Psoriasis

Resumo: Impetigo herpetiforme é uma dermatose gestacional rara que se inicia tipicamente durante o último trimestre e evolui com rápida resolução no período pós-parto. Clinica e histologicamente é consistente com psoríase pustulosa. Essa similaridade tem levado alguns autores a nomearem a doença como "psoríase pustulosa da gestação". Relatamos o caso de uma paciente que apresentou impetigo herpetiforme em duas gestações subsequentes.
Palavras-chave: Gravidez; Impetigo; Psoríase; Relatos de casos

INTRODUCTION

Impetigo herpetiformis is a rare gestational dermatosis with typical onset in the last trimester of pregnancy and rapid resolution in the postpartum period.[1,2] It is clinically and histologically consistent with pustular psoriasis. This similarity has led authors to name the disease "pustular psoriasis of pregnancy".[2]

The maternal-fetal prognosis is uneven. Maternal deaths are rare but there are risks of stillbirth.[3] We report two successive pregnancies of a patient with two different outcomes: stillbirth in the first pregnancy; and a healthy newborn in the second.

CASE REPORT

Patient, female, white, first pregnancy two years ago, at 17 years of age. First assessment at outpatient dermatology service in December 2010, after 27 weeks of pregnancy, due to the appearance of erythematous plaques in the sternal region in week 19, with rapid spread by skin surface. She denied having a history of psoriasis. Patient and fetus were stable from a gynecological standpoint.

Physical examination showed erythematous, scaly, figurative lesions, with pustules on the periphery (Figures 1A and 1B). A biopsy revealed spongiform pustules of Kogoj with neutrophils forming intraepidermal and multilocular microabscesses (Figure 2).

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[1] Dermatologist, Assistant Professor of Dermatology at the Regional University Hospital of the North of Paraná – State University of Londrina (HURNP-UEL) – Londrina (PR), Brazil.
[2] Resident doctor in the dermatology department at the Regional University Hospital of the North of Paraná – State University of Londrina (HURNP-UEL) – Londrina (PR), Brazil.
[3] Dermatologist – private clinic – Tupã (SP), Brazil.
[4] Dermatologist. Master's in Medical Science from the Health Sciences Center at the State University of Londrina (UEL), Assistant Professor of Dermatology at the Regional University Hospital of the North of Paraná – State University of Londrina (HURNP-UEL) – Londrina (PR), Brazil.

Laboratory tests showed leukocytosis of 24.800/mm^3, parathormone within reference values (PTH = 45pg/ml) and increased erythrocyte sedimentation rate (ESR = 80mm/h). Serum levels of calcium, sodium and potassium were normal.

Therapy was begun with oral prednisone 60mg per day. Since there was partial improvement, the decision was taken to reduce corticosteroid dose to 40mg per day. At 39 weeks of pregnancy, the patient spontaneously sought gynecological care, worried about decreased fetal movements. Clinical examination and ultrasound proved there was no fetal heartbeat. After finding the stillbirth, labor was induced vaginally. Systemic corticosteroid was reduced gradually during postpartum. However, persistent cutaneous lesions had motivated introduction of oral cyclosporine, with good response. After resolution of cutaneous symptoms, our patient lost dermatological follow.

In June 2012, a woman was referred by the attending physician of her city of origin for obstetric care in our hospital, 37 weeks pregnant and with skin lesions. On that occasion, dermatology was again called. It was found to be the same patient, now aged 19, G2P1 (1 stillborn), with skin lesions that started two weeks before (Figure 3).

In view of poor fetal outcome in the first pregnancy, we instituted supportive treatment with antibiotics and systemic steroids, and suggested delivery resolution to obstetric team. After failure in inducing vaginal delivery, a healthy child was born by cesarean two days after admission. Skin lesions improved progressively and rapidly during postpartum and steroid dosage was gradually reduced.

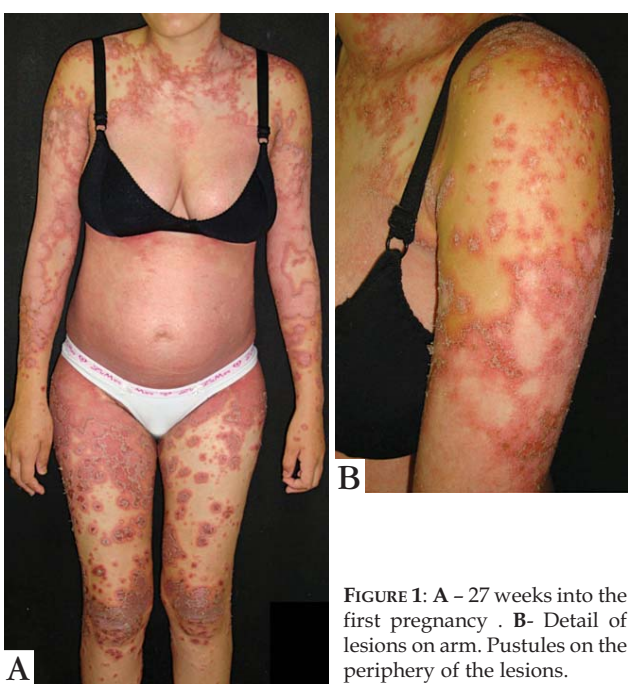

Figure 1: **A** – 27 weeks into the first pregnancy . **B**- Detail of lesions on arm. Pustules on the periphery of the lesions.

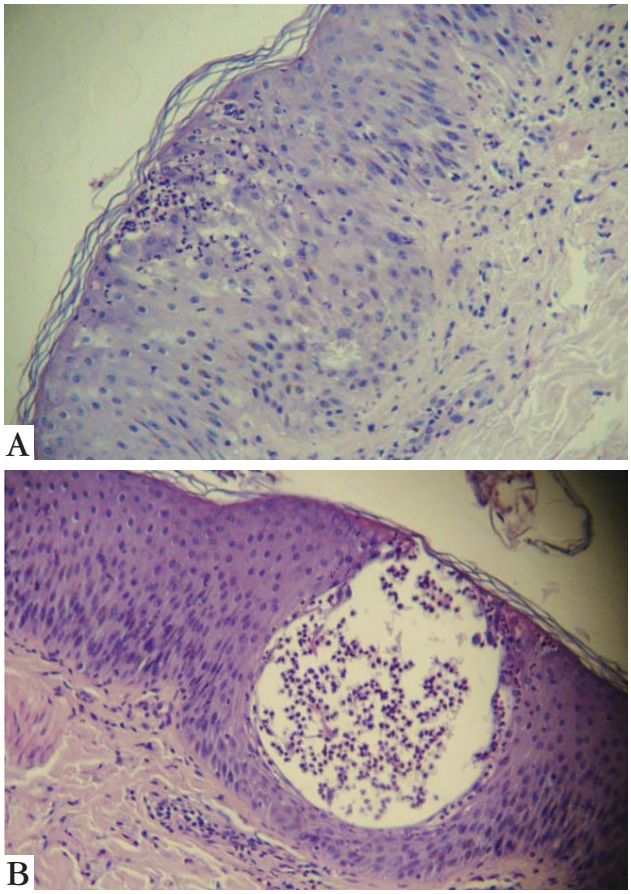

Figure 2: **A** – 27 weeks into the first pregnancy . **B**- Detail of lesions on arm. Pustules on the periphery of the lesions.

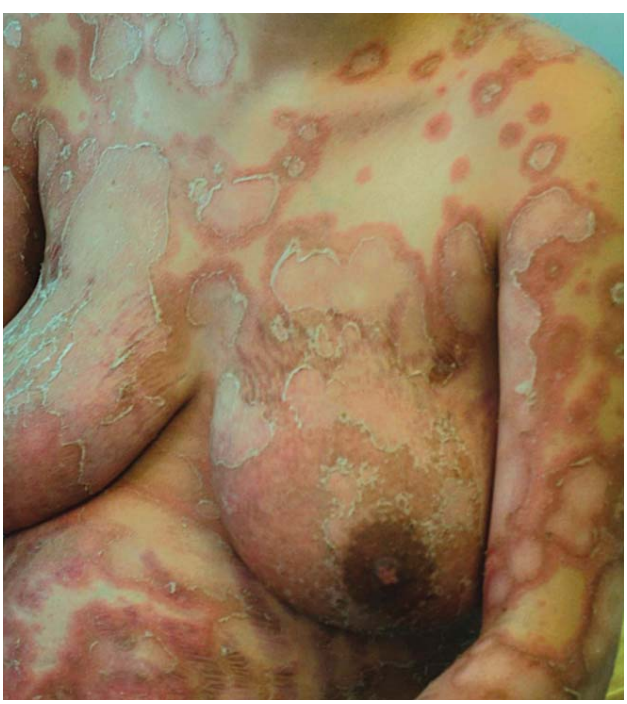

Figure 3: 37 weeks into the second pregnancy

DISCUSSION

Psoriasis is a chronic disease with a worldwide prevalence of 1-3%.[4,5] Clinical forms can be classified into: vulgaris, guttate, nail, inverted, keratodermic, pustular and erythrodermic. Pustular psoriases can be localized, for instance palmoplantar and continuous acrodermatitis of Hallopeau, or widespread, as in acute forms (von Zumbush), circinate and pregnancy (impetigo herpetiformis).[3,4,6]

Impetigo herpetiformis (IH) was first described by von Hebra *et al*, with a report of five pregnant women with pustular grouped lesions, with inflammatory nature and crust evolution, all of which evolved into fetal deaths, in addition to four maternal deaths.[3] Currently, some authors use the term "pustular psoriasis of pregnancy"(PPG), based on the fact that the pustules are sterile and do not present viral etiology.[2,3,7,8]

Some authors consider the IH/PPG as a variant of pustular psoriasis that occurs during pregnancy, due to clinical and histological similarities.[2] Former PPG criteria included: absence of personal and medical history of psoriasis; self-limited disease development, clearing spontaneously after delivery; and recurrence in subsequent pregnancies.[3] There are, however, reports of personal and familial psoriasis history and non-complete regression of lesions after delivery, as observed by Azulay-Abulafia *et al*.[3] Hence, PPG is assumed to be a simple variant of generalized pustular psoriasis, representing a pustular stage of the disease, as a result of the hormonal changes of pregnancy or other factors that are not yet understood.[2,3,7]

Its etiology is still unknown. Association with hypocalcemia and hypoparathyroidism has been suggested, although these findings do not feature in most cases. The role of hormonal contraception, stress, bacterial infection, seasonal variation and certain medications (activated charcoal, potassium iodide and salicylates), is questionable, and more research is needed on this topic.[3]

Clinical examination is characterized by lesions initiated in skin folds, with centrifugal spread, in some cases affecting the entire skin surface. There may be poor general condition, fever, diarrhea, dehydration, tachycardia and seizures. The following laboratory findings stand out: leukocytosis, increased ESR and negative bacterial culture of pustules and peripheral blood. Levels of calcium, phosphate and albumin may be reduced. The diagnosis is suggested by PPG clinical and laboratory elements, and is predominantly confirmed by predominantly neutrophilc inflammatory infiltrate, epidermal acanthosis and papillomatosis with focal parakeratosis, upon histological examination. There are neutrophils collections, forming intraepidermal multilocular microabscesses, called spongiform pustules of Kogoj.[3]

Differential diagnoses include: pustular psoriasis, dermatitis herpetiformis, erythema multiforme, pustular subcorneal dermatosis and gestational pemphigoid.[3,6]

The treatment of choice during pregnancy is systemic corticosteroids, with 30-60mg of prednisone per day. Cyclosporin may be used in refractory cases.[5] Methotrexate and retinoids are contraindicated during pregnancy.[6,7] Even if the pustules are sterile, some authors recommend adjuvant treatment with cephalospirin, especially in cases of slight improvement after the introduction of systemic steroids.[3] Replacement of calcium, fluids and electrolytes should be instituted as indicated.[3,6] Although lesions tend to disappear after delivery, there is a risk of recurrence in subsequent pregnancies, presenting earlier, with greater severity and worse maternal-fetal prognosis. This should be made clear to the patient regarding any future reproductive decisions.[3]

The current use of steroid and antibiotic therapy has dramatically reduced maternal deaths. However, the risk of stillbirth and perinatal mortality remains high, due to placental insufficiency, premature rupture of membranes, preterm labor and intrauterine growth restriction.[3] Dermatologists and obstetricians must work together to improve the quality of life of the mother and, of course, contribute to a favorable outcome for the fetus. ❑

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Mailing address:
Rogerio Nabor Kondo
Rua Paes Leme 1186
86010-610 - Londrina - PR
Brazil
E-mail: rkondo@onda.com.br

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