Case Report

Generalized pustular psoriasis induced by systemic steroid dose reduction*

Danielle Cristine Westphal[1]
Petra Pereira de Souza[1]
Carlos Alberto Chirano[1]
Antonio Pedro Mendes Schettini[1]
Jessica Castiel[1]
Mônica Santos[1,2]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20163804

Abstract: Generalized pustular psoriasis, or psoriasis of von Zumbusch, is an acute and severe clinical form of psoriasis, which usually occurs in patients with psoriasis undergoing aggravating factors. In this work, we report the case of a female patient, 70 years old, who developed generalized pustular psoriasis symptoms while reducing the dose of oral corticosteroids, improperly introduced for the treatment of alleged acute generalized exanthematous pustulosis. The differential diagnosis of generalized pustular psoriasis should be made with other pustular dermatoses, such as subcorneal pustulosis, IgA pemphigus and especially with acute generalized exanthematous pustulosis. Personal history of psoriasis and histopathological findings with psoriasiform changes and subcorneal pustule favored the diagnosis. She was treated with acitretin 30 mg / day, progressing to complete regression of the lesions.
Keywords: Diagnosis, differential; Psoriasis; Therapeutics

INTRODUCTION

Pustular psoriasis is an infrequent clinical variant of psoriasis, characterized by the presence of sterile pustules, non-follicular, that arise in an erythematous area. It may be classified as generalized, annular, exanthematous and localized. These clinical forms are differentiated by their extension, topography, configurations of lesions and by their response to the treatment.[1] Generalized pustular psoriasis (GPP), or psoriasis of von Zumbusch, is an acute and potentially grave clinical form, which occurs usually in patients with psoriasis who undergo aggravating factors, but which may arise in patients without previous history of psoriasis.[2] Among the aggravating factors the infections, sunburns, use of certain medications (lithium, salicylates, tar, chloroquine and beta-blockers) and, especially, the use and later interruption in the use of systemic corticosteroids stand out.[3] In this work, the case of a patient who developed grave symptoms of GPP while reducing the dose of prednisone is reported.

CASE REPORT

Female patient, 70 years old, was diagnosed with palmoplantar psoriasis in 2003. She was treated with topical medications and oral methotrexate, with regression of lesions. In 2012, she developed cutaneous symptoms with erythema and generalized pustules, accompanied by systemic symptoms. She searched another dermatology service, where she was diagnosed with acute generalized exanthematous pustulosis (AGEP). She underwent corticotherapy by injection, in the pulse therapy scheme, and then oral prednisone. There was improvement of clinical conditions, but, when she was in the process of reducing the dose of prednisone, symptoms worsened. At that moment, the dose of prednisone was increased to 80 mg/day, orally. However, during the reduction of prednisone dosage, there was worsening of clinical manifestations, then she decided to abandon treatment and search for another medical service. In July of 2013, she was attended to in our premises presenting generalized erythema and edema, followed by pustular lesions with some crusted areas (Figure 1). Additionally, there was confluence of pustular lesions, forming lakes of pus, mainly in the lower limbs

Received on 28.06.2014.
Approved by the Advisory Board and accepted for publication on 11.02.2015.
* Work performed at Fundação Alfredo da Matta (Fuam) – Manaus (AM), Brazil.
Financial Support: None.
Conflict of Interest: None.

[1] Fundação Alfredo da Matta (Fuam) – Manaus (AM), Brazil.
[2] Universidade do Estado do Amazonas (UEA) – Manaus (AM), Brazil.

©2016 by Anais Brasileiros de Dermatologia

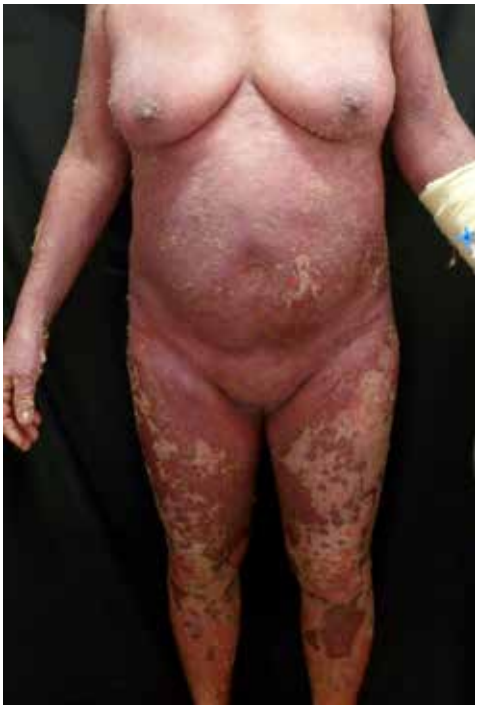

Figure 1: Generalized erythema and edema, accompanied by pustular lesions with some crusted areas

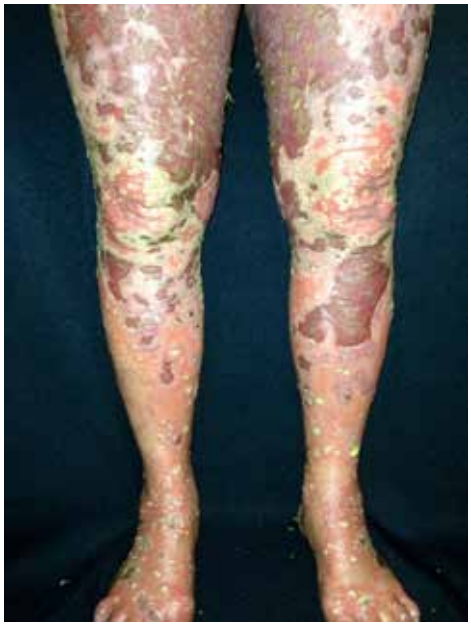

Figure 2: Confluence of pustular lesions in the lower limbs, forming lakes of pus

(Figure 2). There was also general involvement with anorexia, nausea, malaise, shivering and burning sensation at the site of lesions. Due to the seriousness of symptoms, hospitalization was indicated, with intensive care. As for the laboratory tests, hemogram showed leukocytosis and elevated erythrocyte sedimentation rate. Histopathological examination revealed subcorneal spongiform pustule, acanthosis and exocytosis in neutrophils. In the dermis, there was a mononuclear cell infiltrate and neutrophils (Figure 3). The patient was treated with acitretin orally at 30 mg / day and saw significant improvement of lesions, being discharged 21 days after hospitalization. She is still under care, using acitretin, with total regression of lesions and with no relapses to this day – 16 months after introduction of the oral retinoid (Figure 4).

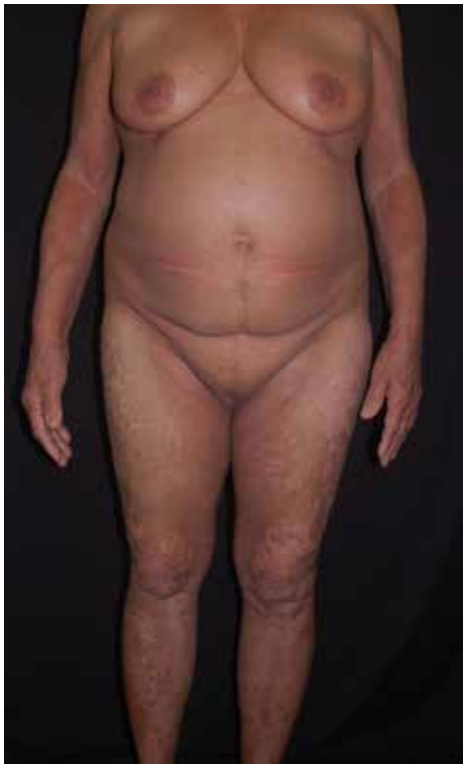

Figure 4: Total regression of lesions, without relapses, 16 months after the introduction of oral retinoid

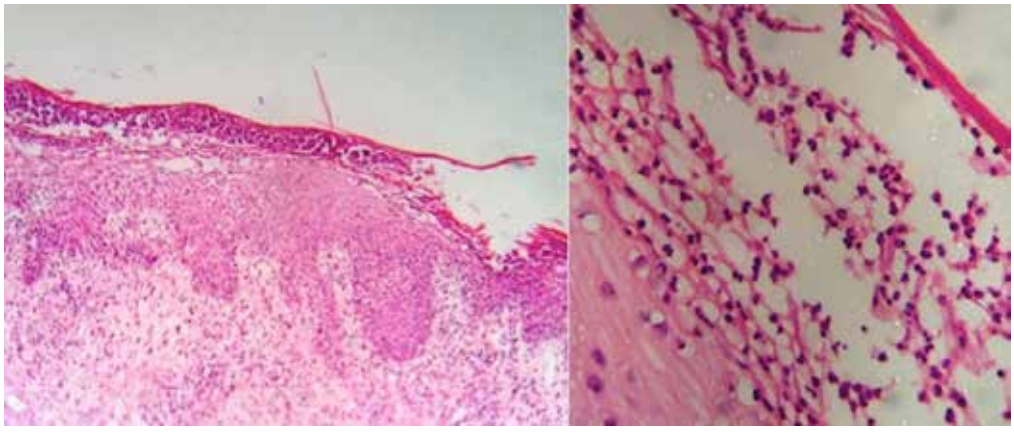

Figure 3: Subcorneal spongiform pustule, acanthosis and exocytosis of neutrophils. In the dermis, mononuclear cells and neutrophils infiltrate (HE, 100x and 400x)

DISCUSSION

GPP is a severe form of psoriasis, which physiopathogeny is not completely clarified. Some authors propose that GPP without previous history of psoriasis has a different etiopathogenic mechanism than the GPP that arises in patients with previous diagnosis of the disease.[4] Despite the fact that the disease is considered polygenic, more recent studies demonstrate that mutations in genes *CARD14* and *IL36RN* may be associated to familial cases of GPP. GPP triggered by mutations in gene *IL36RN*, which codifies the interleukin 36 (IL-36) receptor antagonist, has also been named as IL-36 receptor antagonist (Ditra) and defined by some authors as a new hereditary inflammatory disease.[5] Other studies also point out that certain cytokines may be related to the severity of the disease. Elevated serum levels of IL-4, IL-8, CXCL-1 e CCL-3 were positively correlated with severity scores, while levels of IL-10 and IL-22 usually are diminished after start of treatment.[6] Differential diagnosis of GPP must be done especially with subcorneal pustulosis, IgA pemphigus and AGEP. Previous history of psoriasis and histopathological findings with psoriasiform alterations helps the diagnosis of GPP, and these factors were present in this case. The diagnostic difficulty between GPP and AGEP may be the cause of many cases of pustular psoriasis being treated with systemic corticotherapy, causing worsening of cutaneous symptoms and severe systemic involvement. It is important to emphasize that histological examination of AGEP usually reveals a predominantly eosinophilic infiltrate, with exocytosis of eosinophils, and, in some cases, necrosis of keratinocytes.[7] The histopathology of the present case showed a neutrophilic infiltrate with exocytosis of neutrophils, in addition to subcorneal pustule, which corroborated the diagnosis of GPP. The drug of choice for the treatment of GPP is acitretin, although good results have already been obtained with methotrexate, cyclosporine and immunobiologicals. Most recent studies report that GPP cases associated to mutations of gene *IL36RN* have presented good response to the use of *anakinra*, an IL-1 receptor antagonist.[8-10] ❑

REFERENCES

1. Borges-Costa J, Silva R, Gonçalves L, Filipe P, Soares de Almeida L, Marques Gomes M. Clinical and laboratory features in acute generalized pustular psoriasis: a retrospective study of 34 patients. Am J Clin Dermatol. 2011;12:271-6.
2. Varman KM, Namias N, Schulman CI, Pizano LR. Acute generalized pustular psoriasis, von Zumbusch type, treated in the burn unit. A review of clinical features and new therapeutics. Burns. 2014;40:e35-9.
3. Jaime TJ, Rodrigues BA, Jaime TJ, Barbo MLP, Simis DRC. Psoríase de von Zumbusch. An Bras Dermatol. 2009;84:299-301.
4. Iizuka H, Takahashi H, Ishida-Yamamoto A. Pathophysiology of generalized pustular psoriasis. Arch Dermatol Res. 2003;295:S55-9.
5. Sugiura K. The genetic background of generalized pustular psoriasis: IL36RN mutations and CARD14 gain-of-function variants. J Dermatol Sci. 2014;74:187-92.
6. Yamamoto M, Imai Y, Sakaguchi Y, Haneda T, Yamanishi K. Serum cytokines correlated with the disease severity of generalized pustular psoriasis. Dis Markers. 2013;34:153-61.
7. Kardaun SH, Kuiper H, Fidler V, Jonkman MF. The histopathological spectrum of acute generalized exanthematous pustulosis (AGEP) and its differentiation from generalized pustular psoriasis. J Cutan Pathol. 2010;37:1220-9.
8. Sbidian E, Maza A, Montaudié H, Gallini A, Aractingi S, Aubin F, et al. Efficacy and safety of oral retinoids in different psoriasis subtypes: a systematic literature review. J Eur Acad Dermatol Venereol. 2011;25:28-33.
9. Gallo E, Llamas-Velasco M, Daudén E, García-Diez A. Refractory generalized pustular psoriasis responsive to a combination of adalimumab and acitretin. Int J Dermatol. 2013;52:1610-1.
10. Hüffmeier U, Wätzold M, Mohr J, Schön MP, Mössner R. Successful therapy with anakinra in a patient with generalized pustular psoriasis carrying IL36RN mutations. Br J Dermatol. 2014;170:202-4.

Mailing address:
Mônica Santos
Avenida Codajás, 24
Cachoeirinha
69065-130 - Manaus - AM
Brazil
E-mail: m.n.souza.santos@gmail.com

How to cite this article: Westphal DC, Schettini APM, Souza PP, Castiel J, Chirano CA, Santos M. Generalized pustular psoriasis induced by systemic steroid dose reduction. An Bras Dermatol. 2016;91(5):664-6.