Case Report

Pyoderma gangrenosum: an atypical presentation*

*Pioderma gangrenoso: apresentação atípica**

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Abstract: Pyoderma gangrenosum is a rare neutrophilic disease of unknown origin that is associated with systemic diseases in 50% of cases. We report a case of atypical pyoderma gangrenosum in a 39- year-old man with psoriasis associated and optimal response to cyclosporin. This case report shows the diversity of clinical manifestations of this disease, the difficult diagnosis and the therapeutic options currently available.
Keywords: Cyclosporina; Psoriasis; Pyoderma gangrenosum

***Resumo**: O pioderma gangrenoso é doença neutrofílica rara, de etiologia incerta e que se associa a doenças sistêmicas em 50% dos casos. Relata-se um caso de pioderma gangrenoso com apresentação atípica em um homem de 39 anos com psoríase associada e ótima resposta à ciclosporina. Este relato de caso aborda as diversas formas clínicas da doença, a dificuldade diagnóstica e as opções terapêuticas atualmente disponíveis.*
Palavras-chave: Ciclosporina; Pioderma gangrenoso; Psoríase

INTRODUCTION

Pyoderma gangrenosum is a rare, relapsing and destructive neutrophilic disease. It was described for the first time in 1930 by Brunsting, Goeckermann and O'Leary, and remains with uncertain etiology to the present day.[1]

Most affected age range is 25 – 55 years, and lesions generally begin as pustules, nodules or hemorrhagic blisters that evolve to ulcers with undermined borders, which are usually located in lower limbs and trunk. Genitalia may be involved. Currently, four main clinical forms are described.[1,2]

• The ulcerated form is the most frequent, and corresponds to the classical picture of ulcerations with undermined borders, surrounded by erythema. It is associated to bowel inflammatory disease, arthritis and monoclonal gammopathy.

• The pustulous form is characterized by painful pustules with erythematous halo. It generally occurs in acute exacerbations of bowel inflammatory disease.

• The bullous form is occurs with superficial hemorrhagic blisters, which often leave scars and are associated to myeloproliferating disease.

• The vegetating form presents as a non-painful

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solitary superficial ulcer, not associated to any systemic disease.

There is an association with underlying systemic diseases in 50% of the cases, especially with bowel inflammatory diseases, poliarthrites and hematologic diseases.[1-3]

In this report, we describe a case of pyoderma gangrenosum associated to psoriasis with unexpected presentation, for which the initial treatment attempts failed, but which had optimal response to cyclosporin. Diagnostic difficulty and current available therapeutic options are also discussed regarding this pathology.

CASE REPORT

Thirty-nine-year-old black male patient, married, rural worker, who had been presenting for one year fissured lesion with bright red bed, covered by seropurulent secretion, with wine-colored undermined borders. The condition began in the inguinal region, later on evolving to bilateral ulcers of approximately 20cm (Figure 1).

Similar lesions appeared in axillae, perineum and anus, besides an exsudating ulcerated lesion in the glandis (Figures 2 and 3).

Patient had preserved general state and reported two previous hospitalizations because of his current disease, with various failed therapeutic attempts and with no conclusion on diagnosis. He also presented erythematous desquamative plaques of various sizes in almost the entire body surface, which had been diagnosed as psoriasis two years before. He denied drug use, alcohol drinking, risk sexual behavior or other diseases.

He underwent chest X-ray and laboratorial exams, such as blood count, biochemical profile, VDRL, FTA-ABS, HIV serology, IgG and IgA dosage and urinalysis, all of them normal, with the exception of HSV, which was slightly increased. AARB and fungal cultures were negative. Multisensitive beta-hemolytic *Streptococcos* sp grew in the wound secretion culture. Inguinal and axillary lesion biopsy revelaed a supurative granulomatous process, associated to pseudoepitheliomatous hyperplasia. There was an absence of parasites in all stains carried out.

DISCUSSION

Diagnostic impression was of pyoderma gangrenosum, with an unusual presentation: the patient had lesions in inguinal, axillary, perineal and penian regions, associated to psoriasis. Association between pyoderma gangrenosum and psoriasis was described for the first time in 1994 by Smith and White,[4] and up to this date there are only few published reports.

Initial therapy consisted of a steroid 1mg/kg/day and dapsone 100mg/day, with good response. However, after six months, his improvement halted, and therapeutic scheme was changed. Dapsone was suspended and azathioprine was begun at 100mg/day, which allowed for the reduction of the steroid and improvement of the lesions. Due to the associated psoriasis, azathioprine was replaced by cyclosporin, with excellent response. Initial dose was 5mg/kg/day of cyclosporin, with complete lesion remission after six weeks of treatment. Patient is currently using exclusively cyclosporin at 1mg/kg/day, and remains free of both pyoderma and psoriasis lesions. Renal function was monitored throughout the entire treatment, and presented no alteration.

Pyoderma gangrenosum is an exclusion diag-

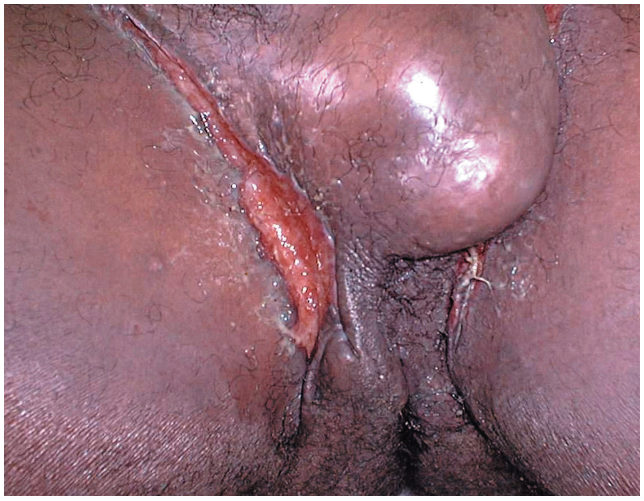

Figure 1: Fissured lesions with undermined borders and bright red bed, covered by seropurulent secretion, bilaterally in the inguinal region

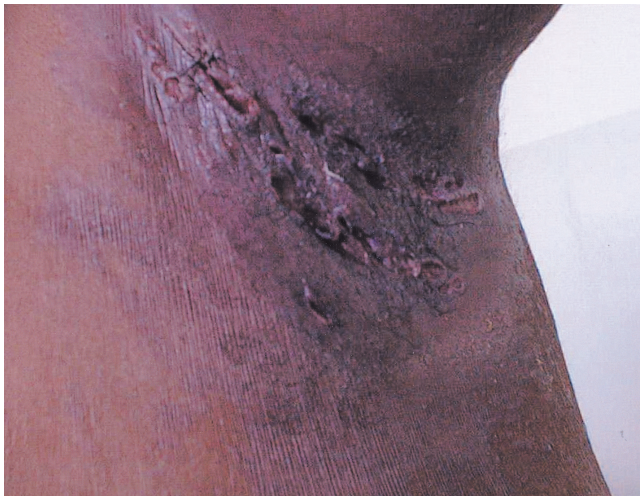

Figure 2: Fissured lesions with undermined borders and bright red bed, covered by seropurulent secretion in axillae

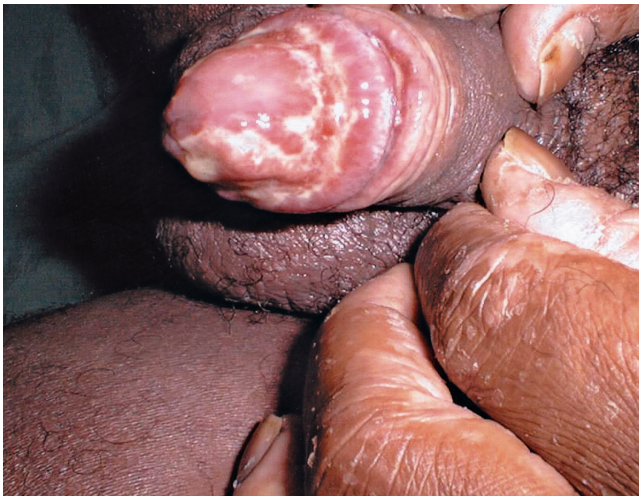

FIGURE 3: Exsudating ulcerated lesion in the glandis

nosis, based on clinical features.[3,5] Laboratorial exams are inespecífico, and, most of the times, only hemosedimentation velocity is increased.[3] Histopathological examination is likewise inespecific, and presents variable aspects, depending on the site of biopsy and disease duration time,[2] yet is fundamental to exclude other diagnoses.

Among differential diagnoses, should be especially born in mind: deep mycoses, vascular ulcers, insect bites, neoplasms and vasculites.[5]

Currently available therapeutic armamentarium is wide, but results are weak. There is no specific and really effective drug. Topical treatment is usually adjuvant and aids in pain relief and avoidance of secondary infections; when used isolated, it rarely controls the disease. Most commonly used drugs are intralesional triamcinolone and sodium chromoglicate.[5]

Systemic treatment is often necessary, and first choice drugs are still steroids,[6] although disease behavior is recurrent, unpredictable, and therapeutic response is individual. There are reports of successful use of other medications, such as clofazimine, dapsone, thalidomide and immussupressors: cyclosporin, azthioprine, cyclophosphamide and mycophenolate mofetil.[1,2,5,6] Even though these may be used isolated, they are generally associated to steroids, in other to reduce the dose of the latter, and as na attempt to control refractory cases. Articles describing the use of tumor necrosis factor (TNF) inhibitors, such as infliximab and etanercept, have been published;[7-9] however, these works are based on small patient sets, with no statistical proofs. There are still other therapeutic options described in the literature, such as hyperbaric oxygen therapy,[5] plasmapheresis and leukocytepheresis.[10]

It is important for the dermatologist to bear in mind the diagnosis of pyoderma gangrenosum, when he or she is confronted with an ulcerating process that has not responded to antibiotics and local care. This will certainly contribute for the improvement of the patient's quality of life, reducing costs, sparing suffering, and also avoiding tempestuous measures, such as unnecessary repairing surgery. ❑

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