Case Report

Pyoderma gangrenosum as a initial manifestation of ulcerative proctocolitis

CARLA BORTOLIN FONSECA¹,2, GUILHERME LANG MOTTA¹,2, ALEXANDRE RAMPACZO¹, JOÃO CARLOS CANTARELLI JUNIOR¹,2, RENATO BORGES FAGUNDES²,³

¹University Hospital of Universidade Federal de Santa Maria (UFSM) – Santa Maria (RS), Brazil. ²Departament of Medical Clinic/Center of Health Sciences of UFSM – Santa Maria (RS), Brazil. ³Post Graduation Program: Gastroenterology Sciences of the Medical School at Universidade Federal do Rio Grande do Sul (UFRGS) – Porto Alegre (RS), Brazil.

ABSTRACT: pyoderma gangrenosum is a rare inflammatory skin condition characterized by progressive and recurrent skin ulceration of destructive course. It is usually associated with rheumatoid arthritis, paraproteinemia, myeloproliferative diseases and inflammatory bowel diseases, especially non-specific ulcerative proctocolitis. In these situations, skin lesions are described as concurrent with the intestinal condition. However, reports on pyoderma gangrenosum preceding intestinal findings are less frequent. The authors describe a case of a woman with febrile condition associated with skin lesions diagnosed by biopsy as pyoderma gangrenosum. Two weeks later, she developed diarrhea, arthralgia and sepsis, being diagnosed as ulcerative proctocolitis. After the administration of the treatment for ulcerative proctocolitis, she showed improvements in sepsis care, remission of diarrhea and regression of skin lesions. This case highlights the importance of considering pyoderma gangrenosum as a manifestation associated with inflammatory bowel disease, regardless of its timing in relation to intestinal symptoms.

Keywords: pyoderma gangrenosum; proctocolitis; pyoderma; colitis.

INTRODUCTION

Pyoderma gangrenosum (PG) is a type of inflammatory skin condition of unknown origin characterized by progressive and recurrent skin ulceration of destructive course¹³. Dermatosis is usually unpredictable, sudden and aggressive, but it can also be chronic, slow and insidious, presenting skin ulcers that expand centrifugally⁴. The aggressive form may cause painful ulcerative lesions, with necrotic and hemorrhagic base⁵. It is associated with systemic disease in about 50% of the cases, such as rheumatoid arthritis, inflammatory bowel disease, paraproteinemia and myeloproliferative disease⁶,⁷. It is described as one of the extraintestinal manifestations for the patients who have inflammatory bowel disease (IBD), especially the severe forms of unspecified ulcerative proctocolitis (UUP)⁸,⁹. PG affects people at any age group, especially young women with proctocolitis and diffuse compromise of the entire colon¹⁰,¹¹. It can appear in any part of the body, but is mostly described in the inferior limbs, especially the lower third of the legs. The skin lesion usually manifests during the two first years of inflammatory bowel disease, being more prevalent during the periods of UUP clinical exacerbation¹². Skin lesions that precede the onset of the intestinal picture are a less frequent situation¹³. The authors report the case of a patient with severe UUP whose initial manifestation was the pyoderma gangrenosum, preceding diarrhea in two weeks.

Study carried out at the University Hospital of Universidade Federal de Santa Maria, Santa Maria (RS), Brazil.
Financing source: none.
Conflict of interest: nothing to declare.

Submitted on: 22/12/2010
Accepted on: 16/02/2011
CLINICAL CASE

A white 46-year-old female patient, who was previously healthy, presented with a 38.5°C fever and skin lesions in the face, limbs and vulva, characterized as deep and painful ulcers, with necrotic purulent center and small hemorrhagic blisters (Figures 1 and 2), five days prior to the first visit. At the doctor’s appointment, the patient was given antithermic drugs; skin lesions were biopsied. The anatomopathological examination of the lesions showed superficial and deep diffuse dermatitis with the prevalence of neutrophils (Figure 3) and epidermal ulceration, which are compatible with pyoderma gangrenosum and evidence of deep abscesses in the subcutaneous tissue (Figure 4).

Two weeks after, she started presenting liquid diarrhea without mucus, suppuration or blood, followed by vomit and persistent fever. She reported the ingestion of untreated water, but denied having contact with sick people, trips, previous medications, insect bi-
Pyoderma gangrenosum as a initial manifestation of ulcerative proctocolitis

Carla Bortolin Fonseca et al.

Journal of Coloproctology
July/September, 2011
Vol. 31 Nº 3

Pyoderma gangrenosum is believed to be the skin manifestation of several systemic diseases. It is associated with: rheumatoid arthritis, myeloproliferative diseases, liver disease, monoclonal gammopathy, Wegener’s granulomatosis, diabetes mellitus and inflammatory bowel disease. In approximately 50% of the cases, no associated disease can be identified, thus being called idiopathic pyoderma. Among the systemic diseases, IBD is the most frequently found, corresponding to 27% of the cases. About 20% of the patients who present with skin lesions suggestive of PG can have IBD. According to other data, the established relation is 0.5 to 5%. The relation between PG and the extension, length and severity of IBD is controversial. PG is a relatively rare extraintestinal presentation of ulcerative proctocolitis, and its incidence ranges from 2% to 12%, affecting both genders and all age groups. It is also associated with Chron’s disease, however, the prevalence of such association is lower than the one observed for ulcerative proctocolitis.

It is believed that PG occurs due to a reaction against antigens of bowel disease. The presence of bacterial antigens in the bowel lumen and their absorption through the affected colonic mucosa could trigger and continue a local and systemic inflammatory reaction, which would be a result of the stimulation of cells in the immune system and

Figure 5. Colonoscopy showing edema, hyperemia and erosions throughout the colon, which is compatible with the active inflammatory bowel disease.

Figure 6. Colonic mucosa with architectural distortion. Detail: gland bifurcation, criterion of a chronic inflammatory process (H&E 20x).
of the production of proinflammatory cytokines\textsuperscript{22,23}. The existence of an antigenic relation between bacterial antigens and the colonic mucosa, biliary tract, skin and/or joints would turn these organs into real “target antigens”, which would explain the different manifestations\textsuperscript{24,25}. Deficiencies in immunoglobulin synthesis, the production of an inhibitory factor for mast cells, neutrophil dysfunction and skin allergies can also be involved\textsuperscript{8,5}. In most cases described in literature, PG presents during the active bowel disease\textsuperscript{12}. Bowel symptoms precede or are concomitant with PG, and exacerbations of the disease can be usually related with worse skin lesions\textsuperscript{26}. However, PG can occur in any stage of the disease, at the absence of active inflammation, even after total colectomy\textsuperscript{24,27,28}. In the described report, skin manifestations preceded the diagnosis of ulcerative proctocolitis in two weeks. PG preceded the intestinal picture in the described report, which reinforces how important it is to correlate both pathologies in order to conduct an early diagnosis.

Clinically, PG may be presented in four types: classic, pustulous, bullous and vegetative\textsuperscript{29}. The patient had the classic type, characterized by deep and painful ulcer, with violaceous border and necrotic purulent center. This type usually affects the legs, but it can also reach the head, neck and the genitalia\textsuperscript{13}, which was the case for this patient.

When PG is associated with IBD, the therapy should be directed to the bowel disease, whose remission is followed by the clinical improvement of the skin lesion\textsuperscript{30}. It is also important to have extra local hygiene care to avoid the secondary infection\textsuperscript{15}.

CONCLUSION

It is essential to consider the presence of IBD in patients with pyoderma gangrenosum, even at the absence of gastrointestinal symptoms, so that it is possible to have an early diagnosis. Thus, the treatment can be rapidly administered to avoid the development of the disease and further complications.

Correspondence:
Renato B. Fagundes, MD, PhD
Avenida Grécia 1000, ap. 1002 B
Passo d’Aria – CEP: 91350-070
Porto Alegre (RS), Brazil.
E-mail: fagundesrb@gmail.com