Pyoderma gangrenosum associated with left iliac vein compression syndrome: presentation of difficult diagnosis*

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Abstract: Pyoderma gangrenosum is a rare neutrophilic dermatosis of unknown etiology, of which the most frequent clinical manifestations are ulcers. The diagnosis difficulty is, among other things, to rule out other causes of ulcers, since it is considered a diagnosis of exclusion. Skin ulcerations may also occur in the iliac vein compression syndrome, which, like pyoderma gangrenosum, mainly affects young women. Because they have such similar characteristics, the presence of vascular disease may hinder the diagnosis of concurrent pyoderma gangrenosum. Because of the clinical relevance of ulcerated lesions and scars, the early diagnosis and treatment of this condition is considered extremely important. We report a case in which the two diseases were associated, hampering the diagnosis of pyoderma gangrenosum.
Keywords: Pyoderma gangrenosum; May-Thurner Syndrome; Skin ulcer

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare neutrophilic condition, described by Brunsting *et al*. in 1930.[1] Its incidence is estimated to be around 3 cases per million persons per year.[2] Pathogenesis is still unclear, however, it is already known that it represents a multifactorial combination, including genetic predisposition, inflammatory mediators and neutrophilic dysfunction.[3] It preferentially affects adults between 20 and 50 years of age, with slight predominance for females.[4] It is strongly associated with inflammatory intestinal diseases, such as ulcerative colitis and Crohn's disease.[5] Pathergy phenomenon has been reported in 20% to 30% of PG patients and is characterized by the appearance of lesions after cutaneous trauma.[6] PG can also develop after surgical procedures, with the appearance of lesions mainly on incision sites.[5] There are 4 clinical variants: ulcerated (most frequent), pustular, bullous and vegetative.[7]

Iliac vein compression syndrome (IVCS), or May-Thurner syndrome, is an uncommon condition characterized by the extrinsic compression of the left common iliac vein by the right common iliac artery. It is more predominant in women between the second and fourth decades of life.[8] Clinical manifestations include pain, edema, unilateral varicosities, lipodermatosclerosis, deep vein thrombosis and ulcers.[9]

We report the case of an ulcerated PG with histopathological confirmation, associated to a chronic venous ulcer on the left lower limb resulting from IVCS in a female patient.

CASE REPORT

A 58-year-old female patient, phototype V, had a painful ulcer for 30 years on the left lower limb, that appeared after trauma to the medial malleolus, of progressive growth followed by edema (Figure 1). She underwent saphenectomy one year after the appearance of the ulcer due to the diagnosis of chronic venous insufficiency made at the time, associated to local dressings, with no

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resolution. In 2011, she was seen for the first time at this outpatient dermatology clinic. In 2012, she had a split-skin graft repair with the plastic surgical team at Hospital **São Paulo**; but she lost the graft. In 2013, she was assessed by the vascular surgery team, that diagnosed IVCS through phlebography of the limb and suggested venous angioplasty with stent to decompress the vessel — the procedure was performed on 31/07/2013. There was partial improvement of the ulcer and edema, and the patient underwent another surgery for skin graft placement on 23/09/2013, having 95% of the graft integrated after 1 month. In January 2014, the ulcerated area started to enlarge once again, with changes in the borders, associated to pain and smaller new ulcers on the same limb. Multiple dressings were used, with no improvement of the ulcerations, and 2 skin biopsies were taken, which were unspecific. Doppler ultrasound of the lower limbs did not show any vascular abnormalities consistent with the patient's pain. In September 2014, cultures for bacteria, mycobacteria and fungi were performed, and were all negative. Two months later, a third biopsy was taken, that showed a neutrophilic dermatitis with vasculitis and fibrosis — the histology corresponded to PG. The patient was admitted into hospital for clinical investigations of possible associated conditions and started treatment with prednisone and methotrexate. She had a good response in the PG lesions, but the initial ulcer persisted (Figure 2). She was discharged and maintained dressings as an outpatient. In the beginning of 2016, she started treatment with hyperbaric chamber, which is ongoing, together with follow-up for dressings (Figure 3).

DISCUSSION

PG is a rare condition, difficult to diagnose. The presence of a concurrent vascular condition that presents with ulceration makes its detection even more difficult. The case reported was in a female patient, in an age group similar to what is described in the literature for both conditions, since both PG and IVCS affect mainly young women.[2,8]

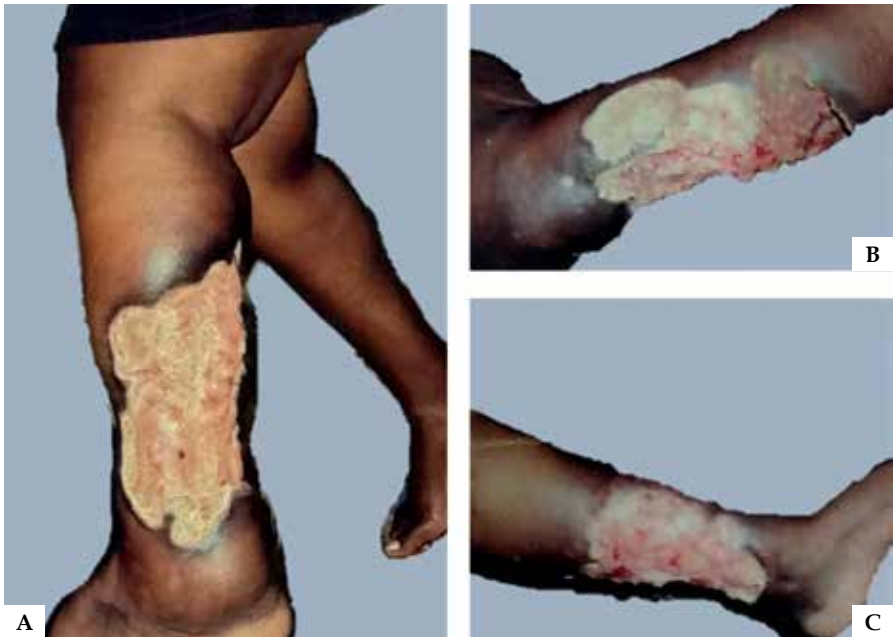

Figure 1: Initial clinical aspect: ulcer affecting almost the whole circumference of the left lower limb. **A:** Lateral view. **B:** Posterior view. **C:** Medial view

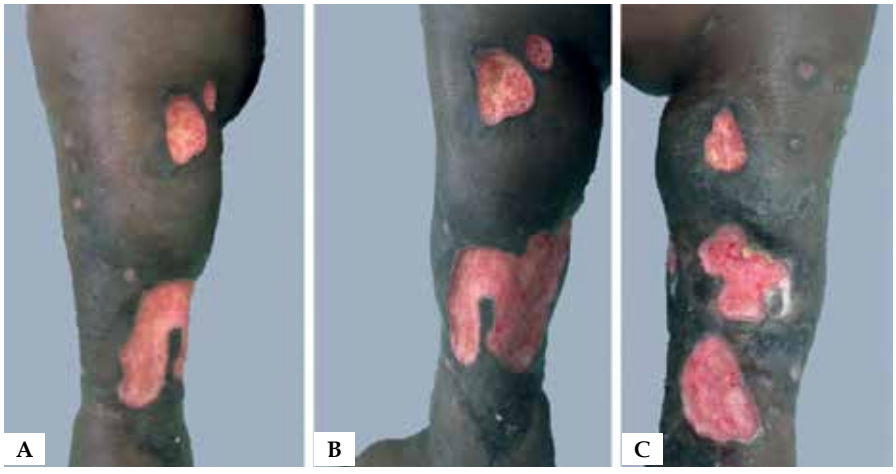

Figure 2: Healing of the smaller PG ulcers after starting treatment with methotrexate and prednisone. **A:** Anterolateral view. **B:** Lateral view. **C:** Medial view

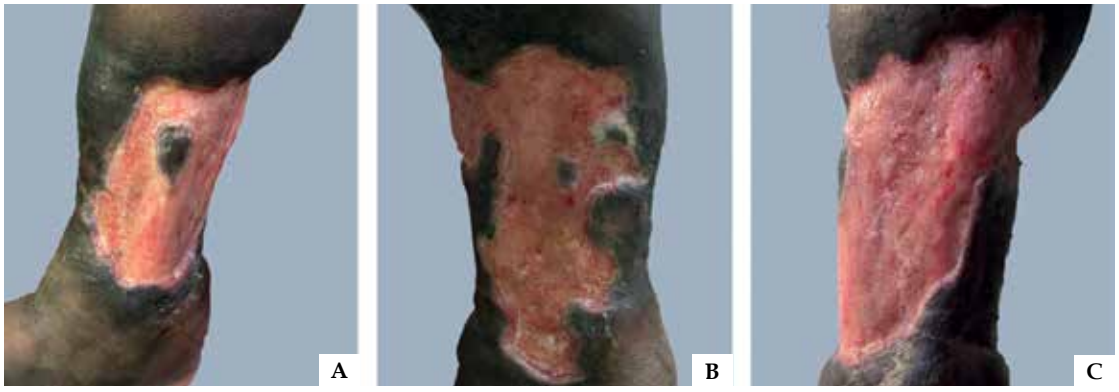

Figure 3: Current aspect, with increased area of ulceration due to PG activity in the border of previous ulcer. **A:** Lateral view. **B:** Medial view. **C**: Posterior view

Because PG is considered a diagnosis of exclusion and because it is less commonly considered in the context of another condition that can lead to lower limb ulcers, it is many times a late diagnosis. In the attempt to improve the recognition of PG, diagnostic criteria were proposed initially by von den Driesch in 1997 and subsequently by Su *et al*. in 2004.[7] These authors proposed very similar criteria, of which 2 majors and at least 2 minors are necessary for the diagnosis of PG. Major criteria include: 1) painful, irregular and violaceous cutaneous ulcer with rapid progression, necrosis and undermined borders; and 2) exclusion of other causes for the cutaneous ulcers. The minor criteria are: 1) history suggestive of pathergy; 2) cribriform scar; 3) associated systemic conditions; 4) consistent histopathological findings (inflammatory and sterile infiltrate in the dermis, with or without mixed inflammatory infiltrate or lymphocytic vasculitis); and 5) quick response to treatment with systemic corticosteroids.[10]

Although not specific, histopathology consistent with PG is crucial when this is suspected, which can demonstrate a neutrophilic inflammatory infiltrate, sometimes with microabscesses, vasculitis with leukocytoclasia and tissue necrosis with a mononuclear infiltrate.[4] Because it is one of the minor criteria, it was essential for diagnosing PG in the reported case.

In our case, the patient had IVCS as the trigger for the ulcer, therefore it was not possible, initially, to fulfill all the criteria needed for the diagnosis of PG according to Su *et al*. However, with the angioplasty, the vascular component was treated and could not be considered as the cause for the new ulcers.

It is possible that the vascular ulcer triggered PG lesions through the phenomenon of pathergy, or that these lesions appeared due to the grafting or angioplasty, since there are reports of post-operatory PG, with the development of ulcers on surgical sites and surgical incisions.[5] Thus, it is important to suspect of PG in association with previous ulcers when those stop responding to the treatment, always taking into consideration the clinical features of the lesions. ❑

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