Journal of Clinical & Experimental Dermatology Research

Paganelli et al., J Clin Exp Dermatol Res 2017, 8:5
DOI: 10.4172/2155-9554.1000416

Case Report Open Access

Old but Gold: The Importance of Medical History in Diagnosing Neutrophilic Dermatoses Characterized by Pathergy. A Case of Pyoderma Gangrenosum

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Received date: August 10, 2017; **Accepted date:** August 21, 2017; **Published date:** August 24, 2017

Abstract

We describe the case of an 84-years old woman referred to our clinic without an established diagnosis in 2015 for the presence of multiple cervical ulcers since 2013, when she underwent vascular surgery of the carotid arteries. The ulcers had infiltrated and actively inflamed violaceous borders; they were itchy and showed signs of scratching. The patient had already been prescribed different antibiotic treatments, without any clinical improvement. Multiple biopsies had also been performed, but histology was not diagnostic, showing a non-specific dermal inflammatory infiltrate. Despite optimal wound care treatment, we observed a dramatic worsening of the skin lesions, spreading from the neck to the vertex, especially at sites of minor trauma (for example, starting from scratch lesions). The presence of pathergy, which consists in the occurrence of lesions at sites of trauma, suggested the diagnosis of pyoderma gangrenosum. Systemic glucocorticoid therapy was then prescribed, with quick improvement and nearly completes clinical remission. Our case confirms the importance of anamnesis and detailed collection of symptoms associated with the clinical manifestations in dermatological dermatoses such as pyoderma gangrenosum where imaging, histology and laboratory findings are often not very helpful for a correct diagnosis.

Keywords: Pyoderma gangrenosum; Pathergy; Ulcers; Medical history; Glucocorticoids

Introduction

Pyoderma Gangrenosum (PG) is a neutrophilic dermatosis which typically presents with painful exudating ulcers, with infiltrated and actively inflamed violaceous borders [1]. One helpful clinical feature for diagnosing active PG may be the presence of "pathergy" or the occurrence of lesions at sites of trauma, in up to 30% of the patients [2-4]. PG may be idiopathic, but it is associated with an underlying disease in 50% of patients [5].

Clinical Case

We describe the case of an 84-years old patient who was referred to our clinic in 2015 for the presence of multiple cervical ulcers evolving since 2013. The patient reported a previous left carotid endarterectomy in 2006, which in 2013 required a surgical re-intervention to substitute the arterial patch. Nearly 3 months later, a slow-growing nodule appeared in the left latero-cervical region and slowly started to grow and eventually ulcerated. Physical examination showed multiple ulcerated lesions with annular disposition at the base of the neck, apparently limited to the superficial layers of the skin (Figure 1). Those lesions were itchy and signs of scratching were present.

The principal diagnostic hypotheses were: neoplastic ulcers, vascular ulcers, infectious ulcers and a factitious disorder (auto-induced ulcers).

Blood tests, swab cultures, cervical soft tissues and carotid ultrasound (US) and magnetic resonance imaging (MRI) of the cervical region were requested as the patient was admitted to our department. Skin biopsies were also performed to rule out a possible malignancy underlying the disease. Microbial cultures were positive for meticillin-sensitive *Staphylococcus aureus*, therefore a specific antibiotic systemic treatment was started. Blood tests were in range, showing only a mild normocytic anaemia. The US showed thickening of the surrounding tissues, without any direct involvement of the carotid area, where previous surgery was performed, as confirmed by MRI.

The patient underwent multiple skin biopsies, but histology was not diagnostic, showing a non-specific dermal inflammatory infiltrate, with hyperparakeratosis and with lymphocytes, plasma cells, neutrophils and multinucleate giant cells both in the dermis and in the hypodermis. We also considered other uncommon possible diagnoses, such as a mycobacteriosis, an actinomycosis or a deep fungal infection, therefore skin samples were sent to our microbiology department, but microbial cultures turned out to be negative. Clinically sulfur granules were absent and Gram staining didn't show the typical gram-positive branching filaments, so we could exclude a possible actinomycosis. Moreover, typical granulomas and acid-alcohol-resistant bacilli (Ziehl Neelsen staining was performed) were absent and quantiferon test was negative.

The patient was then discharged from the hospital and underwent regular follow-up at our Wound Healing and Regenerative Medicine service.

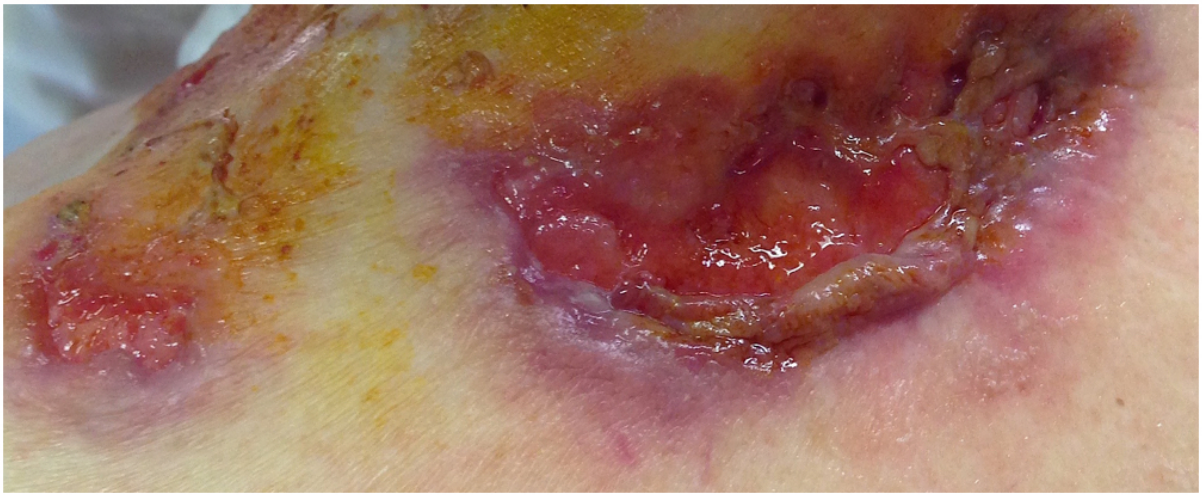

Figure 1: Clinical aspect of the laterocervical ulcers at the first visit in our center. Those ulcers started where a surgical intervention (namely patch substitution after left carotid trombendarterectomy) was performed 2 years before. The macroscopic features are those typical of pyoderma gangrenosum (PG): exudating ulcers, with infiltrated and actively inflamed violaceous borders.

In the next few months we observed a series of relapsing and remitting phases, with the appearance of confluent ulcers of the vertex, with infiltrating erythemato-violaceous borders (Figures 2a and 2b). Scalp involvement wasn't contiguous with cervical area previously involved. The scalp didn't have pustules, so the hypothesis of a concomitant disorder, such as an erosive pustolosis of the scalp was excluded.

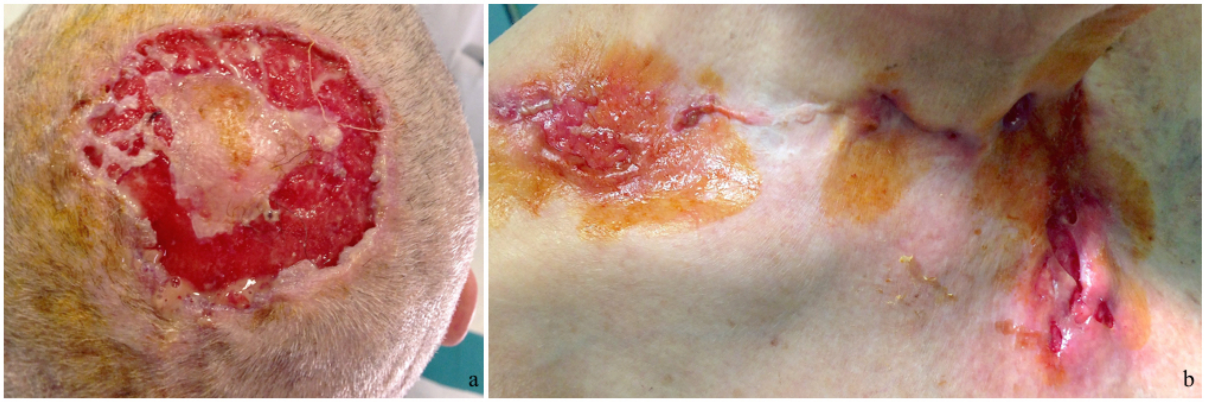

Figure 2: Spreading and enlargement of PG ulcers. Because of a delay in diagnosis, ulcers were left untreated for several months and therefore spread from the laterocervical region up to the vertex (Figure 2a) and to the neckline and the upper part of the chest (Figure 2b). According to the patient, those areas were intensely itchy and the ulcers started from scratching lesions.

The dramatic worsening and enlargement of the skin lesions - despite optimal wound care treatment-suggested us the hypothesis of a pyoderma gangrenosum, with relapsing phases associated with local traumas (such as surgical interventions and scratching). A systemic corticosteroid therapy was then prescribed (prednisone 1 mg/kg/die), with quick improvement and nearly complete clinical remission (Figures 3a and 3b).

Results

PG is a diagnosis of exclusion and recognizing PG can be very challenging [6,7]. There are no definitive tests, and excluding other causes almost always requires a skin biopsy and tissue culture. PG is usually glucocorticoid responsive; failure to respond to therapy should prompt evaluation for an alternative diagnosis. A small number (only 20% to 30%) of patients with PG will exhibit pathergy. It consists in an exaggerated skin injury occurring after minor traumas (bruise, needle stick injury). The induction of more severe lesions in response to trauma, such as surgery, is often described. Pathergy is seen in many neutrophilic dermatoses (PG, Sweet syndrome, and Behçet syndrome in particular).

Pathergy can be demonstrated clinically with pathergy test: a small sterile needle is introduced into the skin of the forearm of the patient

and the test is considered positive when a small pustule and/or an erythematous papule are observed at site of injection after 24-48 h.

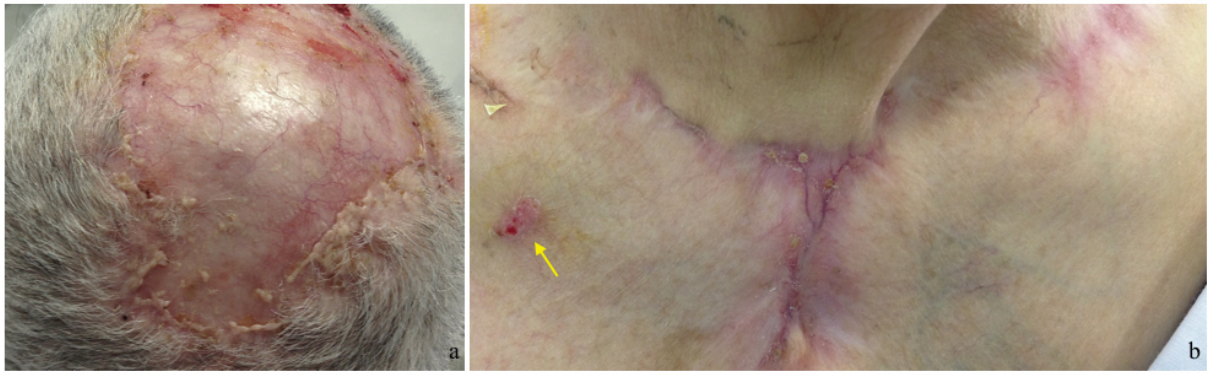

Figure 3: Clinical remission of head (Figure 3a) and cervical (Figure 3b) lesions. As expected, PG ulcers quickly responded to systemic glucocorticoids. The only new lesions (Fig. 3b, arrows) arose from the patch previously applied on the dressing, thus confirming the presence of pathergy in this case of PG.

Management is threefold: wound care, evaluation of underlying causes, and treatment of PG as a chronic inflammatory disease. Treating PG can be very tricky, and if there is an associated underlying disease, therapy should be directed at controlling that process. PG is often responsive to topical or intralesional glucocorticoids. Cyclosporine and infliximab may also be effective, with infliximab having the highest quality evidence, although most clinicians use systemic glucocorticoids as first-line therapy [8-10].

Acknowledgements

We conduct our studies in accordance with recognized international standards, including the principles of the Declaration of Helsinki.

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