Hand Surgery and Rehabilitation 38 (2019) 332–334

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Case report

A case of pyoderma gangrenosum in hand surgery

Un cas de pyoderma gangrenosum en chirurgie de la main

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ARTICLE INFO

Article history:
Received 28 February 2019
Received in revised form 22 May 2019
Accepted 29 July 2019
Available online 6 August 2019

Keywords:
Pyoderma gangrenosum
Chemical burn
Hand surgery

Mots clés :
Pyoderma gangrenosum
Brûlure chimique
Chirurgie de la main

ABSTRACT

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis that can take many clinical forms. Its diagnosis is difficult and often delayed. We report the case of a 36-year-old man with PG complicating a chemical burn of the wrist. This mode of atypical onset delayed the diagnosis and the initiation of appropriate treatment. This was not sufficient since the patient's hand was amputated at his request because of unbearable pain, and failed treatment. Surgeons should be aware of this pathology in cases of rapidly progressing ulcers, especially if aggravated by repeated debridement.

RÉSUMÉ

Le pyoderma gangrenosum (PG) est une dermatose neutrophilique rare, qui peut prendre de multiples formes cliniques. Son diagnostic est difficile et souvent retardé. Nous rapportons le cas d'un homme de 36 ans ayant présenté un PG compliquant une brûlure chimique du poignet. Ce mode de survenue atypique a retardé le diagnostic et l'introduction d'un traitement adapté. Cela n'a pas été suffisant car le patient a été amputé à sa demande en raison de douleurs insupportables, et en échec de traitement. Cette pathologie doit être connue de tous les chirurgiens et être évoquée devant une ulcération d'évolution anormale, et d'autant plus si elle est aggravée par des parages itératifs.

1. Introduction

Pyoderma gangrenosum (PG) is a neutrophilic dermatosis, i.e. disease characterized by cutaneous infiltration of neutrophils, without infectious cause. The incidence of PG is estimated at two cases per year per million people. The peak onset of the disease occurs between the ages of 20 and 50, most often in women [1].

Initial skin lesions are papules, pustules or nodules. They progress quickly to very painful ulcers and centrifugal expansion. The edges are circular, inflammatory and hypertrophic. They are the seat of small purulent openings. An inflammatory peripheral halo is often found. Preferential locations are the lower limbs (75 to 80% of cases) and the trunk [2]. Postoperative PG is a variant that illustrates the phenomenon of pathergy — an inappropriate inflammatory response to surgical skin aggression.

2. Case report

A 36-year-old male patient, without a profession, right-handed, consulted us with a chemical burn on the anterior surface of his left wrist. He was a non-smoker, with no personal or family history of autoimmune disease. The case history was very dubious, since he claimed to have plunged his bare hand into a bucket that he had filled with water himself. According to the patient, its content was replaced with a corrosive substance by a neighbor with malicious intent with whom he had disputes. However, he had no injuries to his fingers and hands, which should have been the case in such circumstances. We were never able to determine the true course of events or the nature of the corrosive substance (acidic or basic). After the diagnosis of third-degree burns and initial debridement at a burn center, he was referred to our hand surgery unit when deep lesions were discovered.

The first debridement we carried out 5 days after the accident identified necrosis of the flexor digitorum superficialis, flexor carpi

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https://doi.org/10.1016/j.hansur.2019.07.012
2468-1229/

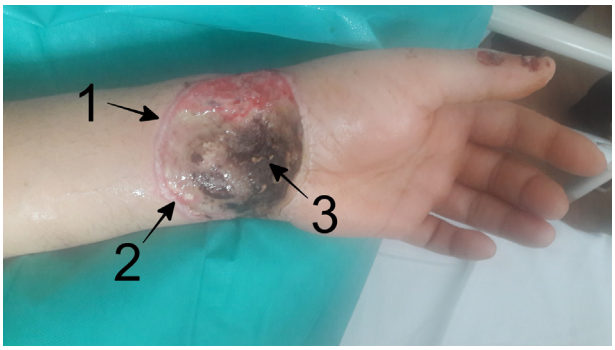

Fig. 1. Pyoderma gangrenosum of the palmar surface of the left wrist of 2 weeks duration. 1: peripheral inflammatory halo; 2: hypertrophic circular border; 3: central ulceration.

radialis and flexor carpi ulnaris tendons. The median nerve was also damaged. It was completely deficient clinically, both in terms of sensitivity and motor activity. However, it was not tested by intraoperative neurostimulation. Initially, the radial and ulnar arteries were permeable Fig. 1.

Repeated debridements were carried out in the operating room due to the rapid growth of the lesions, in depth and circumference. In particular, the flexor digitorum profundus tendons were now affected. Two more surgical sessions were necessary due to spontaneous and significant bleeding from the radial artery. Ulcers appeared, probably due to lesions in the arterial wall. Suturing of the ulcer was performed the first time, but ligation of the artery was necessary upon recurrence, because the lesion was not repairable. An artery bypass operation was discussed but did not seem appropriate due to the ever-changing nature of the lesions.

At the same time, ulnar artery thrombosis occurred, but it did not cause ischemia of the hand, due to collateral circulation, probably through the posterior interosseous artery.

Given the persistence of the extensive lesion for 30 days, a diagnosis of PG was suspected, and confirmed by a skin biopsy, revealing significant dermal and hypodermal neutrophil deposits, associated with necrotic areas. The differential diagnosis of leishmaniasis was ruled out by PCR, since this disease is highly prevalent in the Mediterranean region, including Marseille.

At no time did systemic clinical signs appear, especially hyperthermia. However, the presence of a strong biological inflammatory response (leukocytes at 20 G/L and CRP 250 mg/L) made us suspect a local superinfection.

Pain management was difficult, with nerve damage to the median nerve and PG-related pain combined with burn-related pain. On a visual analog scale, the pain exceeded 8/10, prevented the patient from sleeping and was resistant to morphine. This led to the patient making repeated requests for amputation. He refused any non-drug treatment, such as peripheral nerve catheters, which could have temporarily relieved the pain, and disrupted the painful process underway.

As soon as the possibility of PG was brought up, and even before the histological results, systemic corticosteroid therapy at a dose of 1 mg/kg/day was initiated. The result was good, since the lesions stopped progressing, and the inflammatory syndrome decreased after a few days.

We offered the patient reconstruction solutions: tendon, nerve and cover flap grafts, but the patient categorically refused them. He was concerned about the need for multiple surgeries, the length of recovery, the persistence of pain, and the possible occurrence of PG at a biopsy site. We also suspected he would get a secondary benefit from radical treatment, especially since the circumstances of the accident and the psychological profile of the patient were intriguing.

Forty-five days after the accident, we performed the amputation, proximal to the lesion, at the distal third of the forearm. The pain quickly disappeared, and the stump healed without problems. After healing, he was lost to follow-up.

3. Discussion

PG is a rare disease thus diagnosis is difficult and often delayed. Its natural history involves successive relapses, usually with complete remission in an average of 1 year [3]. The patient's overall condition varies, as PG can be associated with high fever and intense pain. The clinical presentation may be misleading and suggestive of infectious, malignant or vascular disease. Some cases of suspected necrotizing fasciitis have even been reported [4].

The diagnosis is histological. It is performed on a spindle biopsy, removing the edges and bed of the lesion. Neutrophilic infiltration is massive in the dermis and hypodermis, associated with major tissue destruction, and hemorrhagic and necrotic suffusions. Microbiology cultures must also rule out an ongoing infectious process.

Inflammatory diseases (rheumatoid arthritis, chronic inflammatory bowel diseases in 14 to 36% of cases) and blood disorders (myeloid hemopathies, monoclonal gamma diseases) are regularly associated with PG. Neutrophilic disease can be associated, i.e. the generalization of neutrophilic deposits to extra-cutaneous organs [3]. Its treatment is similar to that of PG. Lower limb involvement is the most common. Microbiological samples are negative, unless there is contamination of surface tissues. Skin biopsies of the lesion confirm the diagnosis. However, false negatives are possible [1].

The pathophysiology of PG is currently unknown; it can occur spontaneously or following trauma or surgery. Any type of surgery can contribute to PG. Breast surgery appears to be the most at risk, but there are several reports of PG in hip or knee replacements and peristomal PG after intestinal surgery in Crohn's disease [5]. Lesions appear on average 7 days after surgery and intraoperative corticosteroids appear to reduce the risk of PG [6].

Systematic histological sampling in the event of a potentially infectious lesion would make it possible to diagnose this type of rare pathology and guide therapeutic management. This is all the more so as the main differential diagnoses can be atypical infections (fungal, mycobacterial), tumors or autoimmune diseases with vasculitis.

The reference treatment for PG is systemic corticosteroid therapy initiated at a dose of 1 mg/kg/day, continued for several months [7,8]. Ciclosporin and tumor necrosis factor inhibitors appear to be effective for PGs associated with rheumatic disease [9], or corticosteroid-resistant forms [10]. Local treatment (topical steroids, tacrolimus) is an option, with some authors [3,7] reporting good results. Surgical management of PG lesions, through repeated debridement, contributes to the very phenomenon of pathergy. Therefore, the diagnosis must be made as early as to avoid treatments that increase morbidity and mortality. Once the diagnosis has been made and appropriate treatment has been implemented, skin grafts can be performed.

The diagnostic challenge in our case was due to the two intertwined causes responsible for the ulcerative lesion: chemical burn and PG. This delayed the diagnosis and the implementation of appropriate treatment.

4. Conclusion

Postoperative PG is a rare condition that hand surgeons do not typically encounter. Its diagnosis must be considered when faced with any postoperative recurrent ulcer, especially since it seems to worsen with each additional surgical procedure, and because it

occurs in a patient with a history of inflammatory disease or blood disorder. This diagnosis is all the more difficult because it can occur on an already ulcerative and unstable lesion, as was the case in our patient.

Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Binus AM, Qureshi AA, Li VW, Winterfield LS. Pyoderma gangrenosum: a retrospective review of patient characteristics, comorbidities and therapy in 103 patients: PG: patient characteristics, comorbidities and therapy. Br J Dermatol 2011;165:1244–50.
[2] Hadi A, Lebwohl M. Clinical features of pyoderma gangrenosum and current diagnostic trends. J Am Acad Dermatol 2011;64:950–4 [e2].
[3] Ebrad S, Severyns M, Benzakour A, Roze B, Derancourt C, Odri G-A, et al. Pyoderma gangrenosum after orthopaedic or traumatologic surgery: a systematic revue of the literature. Int Orthop 2018;42:950–4.
[4] Vaysse-Vic M, Mathieu PA, Charissoux A, Charissoux JL, Marcheix PS. Pyoderma gangrenosum or necrotising fasciitis? Diagnostic and therapeutic wanderings. Orthop Traumatol Surg Res 2017;103:615–7.
[5] Wu XR, Shen B. Diagnosis and management of parastomal pyoderma gangrenosum. Gastroenterol Rep 2013;1:273–83.
[6] Zuo KJ, Fung E, Tredget EE, Lin AN. A systematic review of post-surgical pyoderma gangrenosum: Identification of risk factors and proposed management strategy. J Plast Reconstr Aesthet Surg 2015;68:295–303.
[7] Cohen P, Neutrophilic dermatoses. A review of current treatment options. Am J Clin Dermatol 2009;10:301–12.
[8] Reichrath J, Bens G, Bonowitz A, Tilgen W. Treatment recommendations for pyoderma gangrenosum: An evidence-based review of the literature based on more than 350 patients. J Am Acad Dermatol 2005;53:273–83.
[9] Brooklyn TN, Dunnill MG, Shetty A, Bowden JJ, Williams JD, et al. Infliximab for the treatment of pyoderma gangrenosum: a randomised, double blind, placebo controlled trial. Gut 2006;55:505–9.
[10] Lê Huu S, Spertini F, Roggero P, Egloff DV, Raffoul W. Le pyoderma gangrenosum : pathologie rare ou diagnostic omis ? Ann Chir Plast Esthét 2009;54:82–7.