

КЛИНИКА, ДИАГНОСТИКА И ЛЕЧЕНИЕ ДЕРМАТОЗОВ

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Maheshwari K.¹, Yousif A.¹, Burova E.²**Pyoderma gangrenosum of the hand: a missed diagnosis and lessons learned**¹Department of Plastic Surgery of Bedford Hospital, Bedfordshire Hospitals NHS Foundation Trust, MK42 9DJ, UK;²Department of Dermatology Lead, Skin Cancer MDT Lead of Bedford Hospital, Bedfordshire Hospitals NHS Foundation Trust, MK42 9DJ, UK**INTRODUCTION:** *Pyoderma gangrenosum (PG) is a challenging condition to diagnose and treat.***CASE REPORT:** *We discuss the case of a man who presented with a swollen and extremely tender finger, with bullous lesions, erythema, and a clinical picture suggestive of a necrotising fasciitis. He had a history of ulcerative colitis (UC) and had been taking azathioprine. Initial debridement of the affected tissues was carried out as an emergency, with no improvement. Pyoderma gangrenosum had been suspected at that stage and the patient was prescribed oral Prednisolone. He showed a remarkable recovery and his finger healed completely. Due to a persistent pancytopenia he underwent further tests and was subsequently diagnosed with Acute myeloid leukaemia (AML).***DISCUSSION:** *Pyoderma gangrenosum is a diagnosis of exclusion. It can be a sign of internal malignancy and other medical conditions. Therefore, a low threshold must be kept for the diagnosis of this disorder, especially in patients with known risk factors.***CONCLUSION:** *Based on our experience, in similar clinical scenarios, we suggest limb elevation and antibiotics initially, with a close observation and consideration of the diagnosis of pyoderma gangrenosum prior to any surgical intervention. A thorough investigation of the possible reasons for PG is vitally important.***Key words:** *pyoderma gangrenosum; skin infection; hand surgery; paraneoplastic.***For citation:** Maheshwari K, Yousif A, Burova E. Pyoderma gangrenosum of the hand: a missed diagnosis and lessons learned. *Russian Journal of Skin and Venereal Diseases*. 2020;23(4):208-11. DOI: <https://doi.org/10.17816/dv48906>*Acknowledgments.* No financial disclosures.*Conflict of interest.* No conflicts of interest declared.

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Introduction

Pyoderma gangrenosum is a known great mimicker of cutaneous infections. Its appearance and fulminant course often prompt urgent surgical intervention in most clinical scenarios and it is usually in hindsight that a diagnosis of pyoderma gangrenosum is made, due to the lack of clinical improvement and worsening of the condition. Literature is full of case reports of hand complaints, which were eventually labelled as Pyoderma gangrenosum after surgeons were too quick to “put a knife” to them [1–3]. We present a similar case where PG was misdiagnosed initially, but eventually treated medically resulting in complete healing, but also leading to the diagnosis of AML.

Case report

A 63 year old gentleman presented to our accident and emergency department with a swollen finger (**Figure 1**). It was excruciatingly painful, with the swelling starting at the tip of his left middle finger. Over a week it came to involve the whole digit. The patient had already been started on amoxicillin-clavulanic acid by the general practitioner but had to come to the accident and emergency department due to progression of the condition, and plastic surgeons were asked to review him. On examination, the whole finger was swollen down to the palm of the hand, and covered in multiple bullae.

An initial diagnosis of acute infection and possible necrotising fasciitis was made. The fluid from the bullae was sent for culture and sensitivity and blood tests including full blood count. Liver function tests, urea and

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Figure 1. Finger at presentation to the accident and emergency department.

electrolytes and inflammatory markers were requested. Antibiotics and limb elevation were advocated as initial management. The C-reactive protein (CRP) was high, at 240 mg/l, but haemoglobin (Hb), platelet (Plt) and total white blood counts (WBC), were low.

The culture grew *Staphylococcus aureus*, and Teicoplanin was introduced following the results of the culture and sensitivity report. Unfortunately, there was no improvement and 4 days later the finger was covered in dark necrotic crusts (**Figures 2, a, b**). The laboratory risk indicator for necrotising fasciitis (LRINEC) score was moderate, which carried a 50–75% probability of the diagnosis being of necrotising fasciitis. We proceeded with surgical debridement of the wound, and the tissue was sent for histopathological examination.

The condition progressed despite the debridement and the case was discussed with the dermatology team. Based on the histopathology report showing a very dense diffuse neutrophil-rich inflammatory infiltrate extending throughout the thickness of the dermis, a diagnosis of PG was suggested and the patient was commenced on 40 mg of oral prednisolone.

The patient had a remarkable improvement. Over the span of the next 2 to 4 weeks (**Figures 3, 4**) the wound almost completely epithelialised and the patient was started on an early hand physiotherapy to improve hand functions and resolve stiffness. Prednisolone was tapered down gradually by the dermatology team and stopped two months later without any signs of recurrence.

Six years previously the patient had been diagnosed with ulcerative colitis (UC) and had been taking azathioprine, but for two months prior to the development of PG, had been experiencing a flare up. Gastroenterology department was considering the introduction of Adalimumab when the problem with the finger started. After a discussion with dermatology team the patient was started on Infliximab in order to control both UC



Figure 2. Three days later.

a – palmar aspect; *b* – dorsal aspect; at this point the decision was made to proceed with surgical debridement.

and PG. Unfortunately, the patient's low WBC, Hb and Plt, which were originally thought to be due to possible sepsis, continued to drop. He underwent a bone marrow biopsy because of a persistent pancytopenia and had been diagnosed with AML. Sadly, the chemotherapy was not successful, and the patient succumbed to AML seven months after presenting with PG.



Figure 3. Two weeks after starting oral steroids.

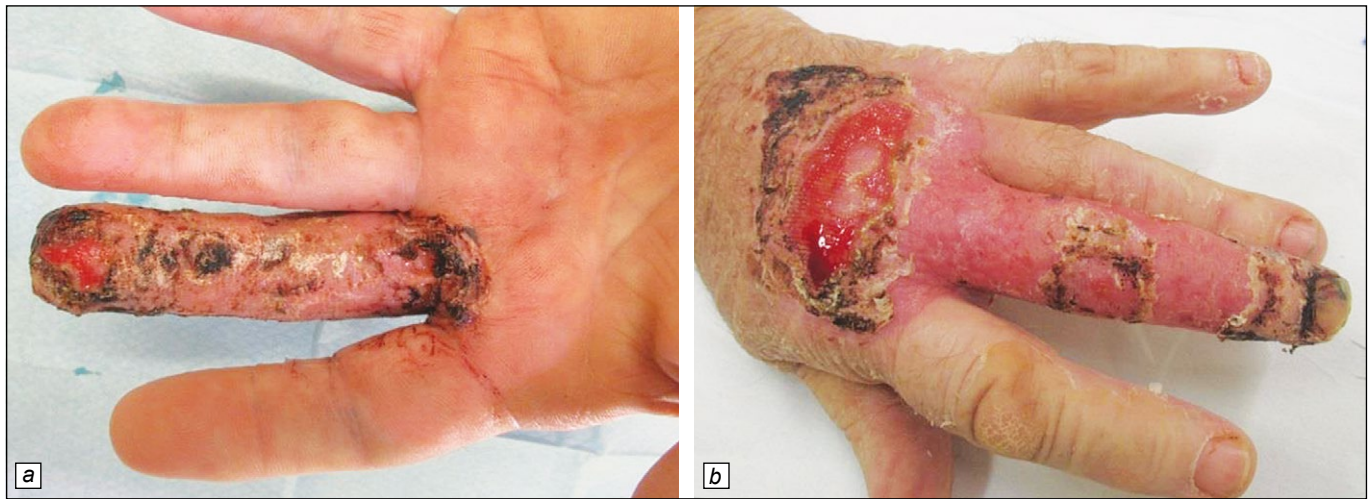


Figure 4. Four weeks after starting steroids (a); b – four weeks after commencing steroids, dorsal aspect. The patient showed remarkable re-epithelialisation without the need for any surgical intervention.

Discussion

PG is a diagnosis of exclusion [4]. It was described by Louis Brocq in 1916 and the term “Pyoderma gangrenosum” was first introduced in 1930 by Brusting et al, who thought it was a cutaneous gangrene caused by a disseminated streptococcal infection [5]. It is now known to be of an auto-immune/auto-inflammatory origin rather than an infective gangrene [6].

Various subtypes of PG are known. The most common of these are the ulcerative type (85%). Other subtypes include bullous, vegetative, pustular, peristomal, and superficial granulomatous variants [7]. It usually presents as an extremely painful lesion, rapidly progressing to a blister or a necrotic ulcer. The ulcers often have a ragged undermined edge with an erythematous border [8]. This was the case in our patient who developed it on the finger, an unusual site [2], with lower limbs being the most common site for PG [7].

Pathogenesis of PG is still poorly understood, though it is recognised that neutrophils play a key role [9]. It belongs to the group of neutrophilic dermatoses [10], and skin biopsy can play a supportive role in diagnosis [7], as happened in our case.

The problem with PG is that there are no widely accepted diagnostic criteria [7]. It is still a diagnosis of exclusion, with histopathology and other tests playing only an adjunctive role [4, 7]. The key to diagnosis is a multi-disciplinary approach. The joint approach by the plastic hand surgery and dermatology teams helped to make the correct diagnosis, and the patient healed well.

The treatment of PG is largely anecdotal with no national or international consensus guidelines, which makes it a challenging condition to treat [11]. Apart from

local wound care and analgesia, systemic treatment with oral corticosteroids (0.5–1 mg/kg/day) is the mainstay of treatment. Steroid-sparing oral medications such as cyclosporin, colchicine, sulphasalazine, dapsone, minocycline, apremilast and thalidomide can be used [7], as well as methotrexate, mycophenolate mofetil, cyclophosphamide, azathioprine and high dose intravenous immunoglobulin [12]. Biologic therapy, targeting a number of cytokines, is the latest in the armamentarium of treatments of PG, with infliximab being the most widely used [13]. There have been reports demonstrating good response to adalimumab, etanercept, ustekinumab, anakinra, and canakinumab [7].

Surgery carries out a high risk of exacerbating PG. This should be kept in mind prior to any aggressive debridement. Taking good history, skin biopsy and appropriate blood tests may be all that is needed initially in order to make the correct diagnosis. Complete healing is achieved in a majority of patients managed conservatively with limb elevation, initial course of antibiotics and treatment with oral steroids and/or steroid-sparing agents, without the need for reconstruction of the wounds [4].

Conclusion

PG is a challenging condition and is best managed by a multi-disciplinary team. It can be a sign of a variety of medical conditions, including malignancies. Surgical intervention should be delayed if there is a high index of suspicion of PG diagnosis. Most of the PG wounds resolve with appropriate systemic therapy, without the need for surgical intervention and reconstruction. A thorough investigation of the possible reasons for PG is vitally important.

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