CASE REPORT

When an ulcer is not 'just an ulcer': pyoderma gangrenosum

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SUMMARY

A 94-year-old man presented with painful leg ulcers that were affecting his mobility. No clear history of these ulcers was obtained on admission or during his initial 7 days of admission and there was no examination made of the ulcer by medical staff during this time. On day 8, a clear history was obtained. It was established that this was a rapidly deteriorating and painful ulcer developing within a 2-week period in previously normal skin. On examination, he had an ulcer clinically consistent with pyoderma gangrenosum which had further enlarged since admission with evidence of newly developing lesions on the legs. He was started on oral and topical therapies and the ulcer improved symptomatically and aesthetically. He was discharged a few weeks later and the patient was followed up by the Dermatology department to ensure continued improvement and eventual resolution.

BACKGROUND

Leg ulcers are a common problem among the elderly population which can result in direct admission to an acute hospital bed. Leg ulcers can arise as a result of multiple pathologies and it is imperative that medical staff perform an adequate history collection and examination to identify those which may have a more sinister aetiology.

We report a case where the diagnosis and initiation of appropriate treatment was potentially delayed due to inadequate assessment in a patient presenting with leg ulcers.

CASE PRESENTATION

A 94-year-old man with a medical history of ischaemic heart disease, prostate cancer and Sjogren's disease and a history of 'leg ulcers' presented to the acute admissions unit. This had initially been managed in the community, but they were increasingly painful to an extent where he was unable to mobilise safely at home. It was unclear in his clerking documentation how long these ulcers had been present and no examination was made of the legs as they had been dressed that morning. Tissue viability reviewed the ulcer 48 h after admission and documented its size (4×3 cm), commenting that the lesion was sloughy with granulation tissue and noted a purple edge. Advice with regard to dressings was given, with no plans for further follow-up.

The patient was reviewed on a near daily basis by junior and senior medical staff for the initial 7 days of his admission. During this time, there was no detailed documentation of the history and progression of the ulcer. No medical staff examined

his legs due to the legs being dressed at the time of ward rounds. Nursing staff were re-dressing his leg every second day, but the appearance of the ulcer at these times was not documented. Throughout this time, the patient continued to report severe and worsening pain in his legs, despite opiate analgesia.

A full history was obtained after 1 week of admission. His daughter stated that the ulcer had developed from entirely normal skin over the 2 weeks preceding admission. She described a small 'black' area that developed initially and rapidly broke down resulting in a painful ulcer. The dressings were removed and the appearance had deteriorated since the last documented examination by tissue viability 5 days ago. The legs were warm and well perfused, with palpable peripheral pulses. The primary ulcer was evident on the left ankle inferior to the lateral malleolus. This was 6×4 cm in size with a maximal depth of approximately 5 mm and a shiny purple edge with deep inflammation (figures 1–3). There were also several new black areas with purple borders elsewhere on the legs (figure 4), with evidence of superficial ulceration around the right medial malleolus.

INVESTIGATIONS

Routine blood analyses demonstrated a raised white cell count and C reactive protein, but were otherwise unremarkable and a vasculitis screen was negative. A wound swab was positive for mixed anaerobes and treated accordingly. A skin biopsy demonstrated parakeratosis and epidermal spongiosis with active inflammatory infiltrate with interstitial haemorrhage, haemosiderin-laden macrophages and fibrin. There was no evidence of vasculitis, squamous dysplasia or invasive malignancy. The features were in keeping with a frictional aetiology; however, this was not

Figure 1 Ulcer to the left lower leg (taken after the start of treatment).

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Figure 2 Ulcer, closeup.

consistent with the clinical picture and given the positive response to treatment this result was disregarded.

DIFFERENTIAL DIAGNOSIS

The findings in this patient were clinically consistent with a pyoderma gangrenosum (PG); however, venous ulceration, arterial ulceration, secondary infection, cutaneous vasculitis and malignancy were considered and excluded.

TREATMENT

This patient had been managed in the community with regular dressings by the district nurses. On admission to hospital, he was initially managed with flomazine and the legs were dressed daily. Flucloxacillin and benzylpenicillin were also started, before being changed to metronidazole in response to a positive microbiology. He was started on a reducing dose of oral steroid (initially 20 mg daily, reduced by 5 mg every 2 weeks) and his topical therapy changed to a dermivate/flomazine mix under Duoderm occlusion when there was a clinical suspicion of pyoderma gangrenosum.

OUTCOME AND FOLLOW-UP

Prior to starting oral prednisolone, this patient's skin continued to deteriorate despite regular topical therapies and dressings. The primary ulcer had enlarged in size and new black areas were developed elsewhere on the legs. Within a few days of initiating steroid treatment, the patient reported his legs to be less painful and the inflammation in the legs was noted to be settling. When reviewed after 3 months by Dermatology department, the lesions had filled in and the edges were pink and healthy in appearance. He remains under review to ensure continued resolution.

Figure 3 Ulcer, demonstrating depth.

Figure 4 New deteriorating areas to lower leg.

DISCUSSION

There is little in the literature examining physicians' attitudes towards and understanding of the assessment and management of leg ulcers. A Canadian study by Graham *et al*¹ examined this in a group of general practitioners. It was found that only 16% felt confident in their ability to manage leg ulcers and 61% felt that they did not have enough knowledge of topical products. Lack of evidence-based guidelines was also a perceived barrier to effective management. A short survey of medical staff in the author's workplace discovered that there was a feeling of lack of knowledge of how to manage ulcers and a perception that it was inconvenient to nursing staff to ask for ulcers to be examined during ward rounds.

The Scottish Intercollegiate Guidelines Network (SIGN) has issued guidelines on the management of leg ulcers, providing some guidance for medical staff. Guideline 120 specifically relates to chronic venous ulceration, but also includes some general ulcer guidance.² Serial surface area measurement is recommended, including description of the ulcer edge. There is also recommendation that referral for specialist assessment is made in the case of non-healing or atypical ulcers.

This patient presented with a history and clinical findings consistent with PG. This diagnosis was delayed in part due to inadequate history and examination in the initial days of admission which was exacerbated by a lack of clear documentation of the changing appearance of the ulcer. This case highlights the importance of history and examination when assessing leg ulcers. Medical staff must be aware of common ulcer aetiology and be able to identify concerning features in either the history or the examination to enable appropriate referral and timely diagnosis and management. In addition, regular review and documentation of the appearance of the ulcer must be completed to ensure that any deterioration is identified and addressed in an appropriate manner.

PG is a rare neutrophilic dermatosis that presents as an ulcerating skin disease.³ It is typically seen as a rapidly enlarging and painful ulcer with an undermined and violaceous border, although it can exist in many variants and this can delay diagnosis and treatment. PG is largely a clinical diagnosis and there are no specific diagnostic features on biopsy,⁴ although a neutrophilic inflammatory infiltrate may be seen. Biopsy is, however, useful to exclude other pathology such as cutaneous vasculitis and malignancy.

PG is idiopathic in nature in approximately half of cases; however, it has also been shown to be associated with underlying systemic disease such as inflammatory bowel disease, leukaemia and other haematological conditions and arthritis.⁴ The patient presented in this case is known to have Sjogren's disease. Sjogren's disease is rarely associated with PG, but there are a few case reports indicating a relationship such as Ravic-Nikolic

Learning points

- ▶ A good history taking and examination is vital when assessing an ulcer, as is regular assessment of wound healing. "If you do not unveil the patient's dressings, you will not unveil the patient's diagnosis".
- ▶ Always reconsider a diagnosis if there is no improvement with treatment.
- ▶ Consider pyoderma gangrenosum in a rapidly enlarging and painful ulcer. Typically, a violaceous and undermined edge is also seen.

*et al.*⁵ They describe a 61-year-old man with known Sjogren's disease who presented with PG over the left thigh. He was treated with prednisolone and dapsone. His ulcer healed completely with treatment after 2 months, leaving an atrophic scar.

The main principle of treatment in PG is immunosuppression,⁴ most commonly with corticosteroids as in this case. However, in some cases, topical potent steroids alone may also induce recovery. The prognosis is unpredictable³ and often long-term steroid

sparing immunosuppressive therapies are required, where most commonly ciclosporin is used. There is also evidence that infliximab, an antitumour necrosis factor, can be effective.⁴ Without adequate and prompt treatment, there is risk of significant deterioration in the ulcer(s) and ultimately scarring.⁴

Contributors MLA researched and wrote the case report. GM provided assistance with background research. Both cared for the patient as inpatient.

Competing interests None.

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