A case of parastomal pyoderma gangrenosum

Zexi Allan, Alex Wong

ABSTRACT

Parastomal pyoderma gangrenosum is a rare condition where classical skin lesions are found around or near abdominal stomas. Literature reports a 2–4.3% prevalence of parastomal pyoderma gangrenosum in patients with inflammatory bowel disease who undergo stoma surgery. This debilitating skin condition is often missed or wrongly treated due to lack of awareness and understanding. We present a 51-year-old female with parastomal pyoderma gangrenosum on a background of Crohn’s disease. Current literature of diagnosis and management is also discussed.
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Parastomal pyoderma gangrenosum is a rare condition where classical skin lesions are found around or near abdominal stomas. Literature reports a 2–4.3% prevalence of parastomal pyoderma gangrenosum in patients with inflammatory bowel disease who undergo stoma surgery. This debilitating skin condition is often missed or wrongly treated due to lack of awareness and understanding. We present a 51-year-old female with parastomal pyoderma gangrenosum on a background of Crohn’s disease. Current literature of diagnosis and management is also discussed.

Keywords: Colorectal surgery, Inflammatory bowel disease, Pyoderma gangrenosum, Stoma

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INTRODUCTION

Pyoderma gangrenosum is a type of neutrophilic dermatosis which causes progressive painful cutaneous ulcerations. Half of the cases are found to be associated with underlying systemic diseases, such as inflammatory bowel disease (IBD) [1]. Pyoderma gangrenosum can affect any age, with an average onset between 40–60 years, and females are more common affected than males [2]. Parastomal pyoderma gangrenosum is a rare subset where classical skin lesions are found around or near abdominal stomas. Literature reports a 2–4.3% prevalence of parastomal pyoderma gangrenosum in IBD patients who undergo stoma surgery [3]. This is a debilitating skin condition which is often missed or wrongly treated. Timely recognition and appropriate treatment can significantly improve patients’ quality of life.

CASE REPORT

A 51-year-old female with Crohn’s disease presented electively for reversal of loop ileostomy. She previously underwent subtotal colectomy with ileorectal anastomosis and loop ileostomy six months ago for Crohn’s colitis.

The patient had ongoing issues with her stoma since surgery. She was admitted two weeks postoperatively with suspected parastomal wound infection with severe pain and purulent discharge. Abdominal computed tomography scan showed no collection. She was discharged home with oral antibiotics. Since then, she had constant stoma leakage and pain around stoma site. However, parastomal skin remained intact. Outpatient review five months postoperatively indicated that the parastomal skin started to show signs of disease with circumferential ragged edges, induration and severe
tenderness to touch. Patient also reported occasional fluid discharge from pinhole breaks of the surrounding skin. Crohn’s associated pyoderma gangrenosum was suspected by stomal therapist, however patient was not referred for medical assessment and was managed with stoma care only.

Pain and local skin changes continued to the time of her reversal procedure. At the time of surgery, her stoma was retracted, with parastomal skin erythema, induration, ulceration and breakdown (Figure 1) and it was very tender to touch around the stoma site. Reversal surgery was performed and skin biopsies were only taken post parastomal skin debridement and curettage. Subsequent histopathology showed chronic inflammation along with reactive epidermal hypertrophy and some fibrosis; features of pyoderma gangrenosum were not seen. Despite the biopsy results, we highly suspect parastomal pyoderma gangrenosum given the clinical appearance and severe pain.

**DISCUSSION**

Pyoderma gangrenosum typically presents as cutaneous ulceration with a well-defined, undermined violaceous border [4]. This can be mistaken for other causes such as infection, stitch abscess, contact dermatitis and irritation from fecal material [1]. Missed diagnosis occur also due to lack of knowledge and familiarity of clinicians to parastomal pyoderma gangrenosum [5]. It is often a diagnosis of exclusion and is particularly difficult to manage owing to its constant pain resulting in poor application of stoma appliance. Correct recognition and appropriate management is essential to improve patients’ quality of life.

There are no absolute diagnostic criteria for pyoderma gangrenosum. While histology from skin biopsy typically show edema and massive neutrophil infiltration confined to the dermis, diagnosis of parastomal pyoderma gangrenosum is mainly via its clinical appearance, a high degree of pain and its rapid progression. The ulcers are very destructive and can expand by 1 to 2 cm from hours to a day [1, 5]. Biopsies are done often to rule out other causes of ulceration rather than to diagnose pyoderma gangrenosum histologically [3]. One study from the Mayo clinic [6] put together a diagnostic guideline; as given in Table 1. The diagnosis is strongly indicated when both major criteria and at least two minor criteria are met.

Once diagnosis is suspected or other causes have been excluded, appropriate management can significantly improve stoma care and patient’s quality of life. A multidisciplinary approach is encouraged involving gastroenterologist, dermatologist, colorectal surgeons and stoma therapist [3]. Medical rather than surgical management is preferred as debridement is not recommended and relocation of stoma is contraindicated unless it is done for other reasons [3]. Given the nature of pathergy, debridement will only exacerbate the problem and relocation often result in recurrence of the disease at the new stoma site [1, 5]. Treatment regimen depends on the severity of the disease and whether there is active systemic disease (i.e., IBD). For mild cases, wound management with moisture-retentive dressings plus topical steroids or intralesional injection of triamcinolone or cyclosporine has been shown to be effective [3]. Systemic therapy is indicated when there is lack of response to topical therapy, if the disease is severe and rapid or if there is active underlying disease.

**CONCLUSION**

Parastomal pyoderma gangrenosum is a rare but debilitating condition and prompt recognition, diagnosis

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**Table 1: Proposed diagnostic criteria of classic, ulcerative pyoderma gangrenosum**

<table>
<thead>
<tr>
<th><strong>Major criteria</strong></th>
<th><strong>Minor criteria</strong></th>
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<tbody>
<tr>
<td>• Rapid progression of a painful, necrolytic cutaneous ulcer with an irregular, violaceous, and undermined border</td>
<td>• History suggestive of pathergy or clinical finding of cribriform scarring</td>
</tr>
<tr>
<td>• Other causes of cutaneous ulceration have been excluded</td>
<td>• Systemic diseases associated with pyoderma gangrenosum</td>
</tr>
<tr>
<td>• Histopathologic findings (sterile dermal neutrophilic, +/- mixed inflammation, +/- lymphocytic vasculitis)</td>
<td>• Histopathologic findings (sterile dermal neutrophilic, +/- mixed inflammation, +/- lymphocytic vasculitis)</td>
</tr>
<tr>
<td>• Treatment response (rapid response to systemic steroid treatment)</td>
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and management is required to improve stoma care and patients’ quality of life.

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Author Contributions
Zexi Allan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Alex Wong – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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