CASE REPORT

Pyoderma gangrenosum as a first presentation of inflammatory bowel disease

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SUMMARY

Up to 40% of patients with inflammatory bowel disease (IBD) develop an extraintestinal manifestation of the disease with the skin being the most commonly involved organ. Pyoderma gangrenosum (PG), an autoinflammatory non-infectious neutrophilic dermatosis, occurs in 1–2% of patients with IBD. PG can follow a course independent to that of the bowel disease, however, most reported cases describe PG occurring in patients with an established diagnosis of IBD. We present a case of a young patient who presented with axillary skin ulceration, which was subsequently diagnosed as PG. On further investigation for a possible underlying cause, she was found to have Crohn's disease. She had not developed any preceding change in her bowels and did not have abdominal pain; the IBD was diagnosed on endoscopic findings. This case is also unusual for the distribution of the PG lesions that typically occur in the lower limbs.

BACKGROUND

Up to 40% of patients with inflammatory bowel disease (IBD) develop extraintestinal manifestations of the disease. These can involve almost any body system ranging from the eyes to the skeletal system and have a huge impact on patients' symptom burden and quality of life. The extraintestinal manifestations can follow a course similar to that of the bowel disease and respond to treatment for bowel inflammation or can, conversely, follow a course completely independent to the IBD.^{1–3}

Our case is unusual in that the extraintestinal symptoms heralded the presence of an underlying IBD. We also discuss the management of pyoderma gangrenosum (PG), which can overlap with that of the IBD.

CASE PRESENTATION

A 22-year-old female patient presented to hospital with large painful ulcerating lesions under her right axilla and on her back. She had first noticed a few boils in her axilla, which her general practitioner had initially treated with antibiotics and later lanced when they had not responded to the systemic antibiotics. These boils grew bigger, coalesced and formed large sloughy ulcers.

On further questioning, the patient reported lethargy, poor appetite and weight loss. She denied any diarrhoea, constipation or abdominal pain. She was not on any regular medication except an oral contraceptive. She was currently unemployed and had returned from a backpacking holiday in the USA, Mexico and Jamaica.

On examination, there were multiple pustular lesions with a white surface across her back (figure 1) and several deeper ulcers with purple undermined edges and yellow discharge in her axilla, which developed a cribriform appearance on healing (figure 2). Her abdominal examination was unremarkable.

INVESTIGATIONS

Initial blood tests revealed a microcytic anaemia (haemoglobin 9.5 g/dL), hypoalbuminaemia (21 g/L) and a C reactive protein 166 mg/L. Her renal and liver function tests were normal. Wound swabs did not grow any pathogen and a skin biopsy revealed a deep suppurative folliculitis with a dense neutrophilic infiltrate.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis initially was infective ulceration such as *Staphylococcus* or leishmaniasis, or an inflammatory skin condition. The negative wound swabs and findings from histology supported a diagnosis of PG.

TREATMENT

The patient was initially started on intravenous broad spectrum antibiotics, however, with the

Figure 1 Phagedenic ulcers on patient's back with violaceous borders.

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Figure 2 Large ulcer in axilla with cribriform pattern of healing.

diagnosis of PG she was subsequently treated with oral and topical steroids as well as regular moist wound dressings.

OUTCOME AND FOLLOW-UP

Following the diagnosis of PG, there was further investigation to determine the presence of an underlying disorder predisposing to it. Given the history of weight loss and low albumin she went on to have a flexible sigmoidoscopy, which revealed patchy areas of deep ulceration and cobblestone mucosa (figure 3). Histology specimens confirmed a diagnosis of Crohn's disease.

The patient was started on steroids for the PG, which were continued on a reducing dose regimen for Crohn's. She also received daily wound care and her skin lesions showed a good response. She subsequently had thiopurine methyltransferase levels checked and was started on azathioprine for Crohn's disease.

DISCUSSION

The skin is one of the most commonly affected extraintestinal organs in IBD; this can reflect a pathological process identical to the IBD as seen on histology, demonstrate a reactive change, reflect chronic inflammation or occur as a side effect of IBD treatment.³ The underlying process determines whether the skin changes reflect IBD activity or occur independently to periods of IBD flare. The most common skin manifestations are erythema nodosum or PG. While erythema nodosum tends to manifest at periods of IBD flare, PG can also manifest during periods of

Figure 3 Deep ulceration on flexible sigmoidoscopy in keeping with Crohn's disease.

bowel disease quiescence.¹ PG has an annual incidence of up to 10 per million and tends to affect women more commonly than men. There is usually a history of an underlying associated disorder such as IBD, rheumatological disease or lymphoproliferative disorders. It presents as painful pustules or nodules that progress to well-demarcated ulcers with undermined violaceous edges. As the ulcers heal they have a cribriform appearance. Lesions have a predisposition for the lower limbs but can occur at other sites and rarely can become widespread. Associated systemic upset with fever, malaise and arthralgia can also occur.³⁻⁵

Diagnosis of PG is clinical and histology can be supportive. It is important to exclude other causes that can present with similar lesions such as vasculitis and infections.⁴ PG is an inflammatory neutrophilic dermatosis that comes under the larger umbrella of an autoinflammatory disease. Autoinflammatory conditions present with recurrent attacks of non-infectious inflammation at target sites (skin in this instance) without associated high levels of circulating autoantibodies or autoreactive T cells. PG immunohistochemistry studies have shown neutrophil recruitment at the lesion bed and T-cell-mediated and macrophage-mediated ulcer formation at wound edges.^{3 6} PG can demonstrate pathergy (lesions develop at site of trauma). It is not uncommon for the diagnosis of PG to be delayed or missed, which can lead to inappropriate therapy such as surgical debridement.⁵ Literature describing PG in IBD usually presents data for patients where the IBD precedes the skin lesions. Our case highlights the importance of exploring an underlying disorder in a patient with PG.

Treatment for PG can involve a combination of wound care, topical therapy and systemic therapy. Wound care is essential for all lesions and is tailored to the type of lesion and extent of associated exudate. A moist environment is typically recommended to promote wound healing. It is also important to monitor for signs of superimposed infection.

Topical treatment as monotherapy can be applied for localised and mild disease but can also be used in combination with systemic treatment for more severe lesions. Topical therapies that have been tried with some success include corticosteroids, tacrolimus, ciclosporin, 5-aminosalicylic acid and dapsone.⁵

More extensive disease is usually initiated with systemic steroids. Cytotoxic agents, most commonly ciclosporin, and others such as azathioprine can also be used, especially in IBD, where their use is well established for the intestinal disease. Cyclophosphamide, mycophenolate and tacrolimus have been used as well. Systemic dapsone has also been used in PG with good results. Recently biological therapy, specifically infliximab, for PG in the context of IBD, has shown very good results.^{4 5}

Despite use of steroids and cytotoxic agents, lesions can be very resistant to treatment and have a massive impact on patients' quality of life.

Learning points

- ▶ Extraintestinal manifestations of inflammatory bowel disease (IBD) are common and can be the presenting feature of the disease.
- ▶ Pyoderma gangrenosum presents as large painful ulcers; the diagnosis is mainly clinical but histology is supportive.
- ▶ In patients presenting with pyoderma gangrenosum it is important to look for an underlying, predisposing disorder.
- ▶ Treatment involves wound care and immunosuppressive therapy similar to that for the IBD, however, the course of pyoderma gangrenosum can be different to that of the bowel disease.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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