Bilateral pyoderma gangrenosum associated with ulcerative colitis and restorative proctocolectomy☆

Pioderma gangrenoso bilateral asociado a colitis ulcerosa y proctocolectomía restauradora

Inflammatory bowel disease (IBD) is associated with a broad spectrum of extraintestinal manifestations. The second most common type of extraintestinal manifestation is a group of dermatological disorders, which include erythema nodosum (EN), aphthous stomatitis and pyoderma gangrenosum (PG).

PG is a neutrophilic dermatosis which occurs in approximately 1% of patients with IBD, being more common in females than males.[1] Recent studies point to a higher incidence of PG in patients with ulcerative colitis (UC) than in Crohn's disease (CD), and in almost all published cases there is or has been colonic involvement.[1,2] PG can occur before, after or simultaneously with the diagnosis of IBD, and it seems to be unrelated to its course and severity.[2] It is characterised by the appearance of one or more pustules on the skin which quickly evolve into a very painful ulcer, sometimes large in size, with an exudative base and raised perilesional violaceous growth around the edges. Pathology study is non-specific, but allows other potential causes of ulceration to be excluded, such as infection or chronic ischaemia. Treatment includes local measures, such as debridement of necrotic tissue and topical antibiotics and, in most cases, systemic treatment with corticosteroids is necessary (prednisone 1–1.5 mg/kg/day or equivalent). In refractory cases (up to 50% in some series), immunosuppressants such as calcineurin inhibitors or anti-TNF alpha are used.[3,4]

We present the case of a 48-year-old male who had a severe extensive flare-up of UC in 2007, refractory to corticosteroids and infliximab, which required surgery (proctocolectomy with an ileo-anal pouch). The patient made a good recovery, with isolated episodes of pouchitis, but which were mild and easy to control. In 2013, he had anterior uveitis, and in the last year he had been referred to rheumatology for peripheral arthritis. He developed both conditions without intestinal symptoms.

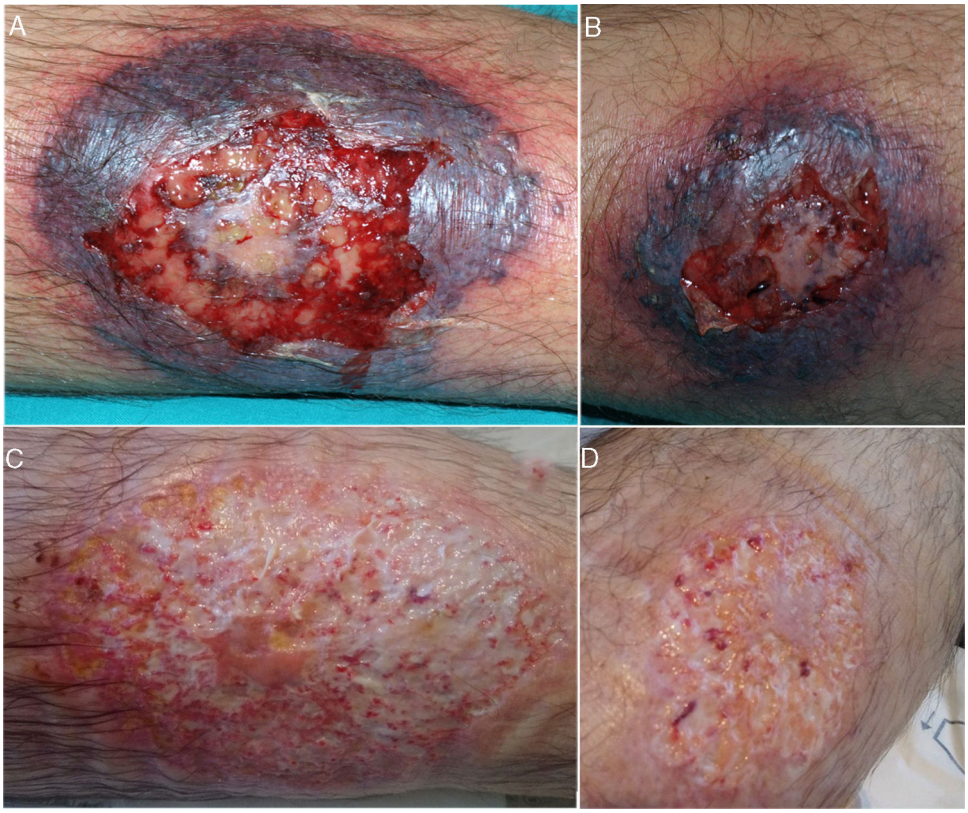

Figure 1 Pyoderma gangrenosum: macroscopic appearance after 1 week (A: right leg and B: left leg) and 3 weeks (C: right leg and D: left leg) of systemic corticosteroid therapy.

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The current episode began in January 2018 with the appearance over the space of a week of millimetre-sized pustular lesions on both lower limbs. The lesions initially improved with oral prednisone, but after that was stopped, they reappeared, this time developing more rapidly and becoming more severe, the reason for which he was admitted to our hospital. Examination revealed very painful ulcers in the right pretibial region and lateral aspect of the left leg, the largest in diameter being of 8 and 4 cm, respectively, with violaceous erythematous borders and a necrotic perilesional growth rim (Fig. 1A and B). The patient was assessed by dermatology. Microbiological study was negative. The biopsy of one of the lesions showed a hypodermis with acute inflammatory infiltrate and local abscess, with septal granulation tissue, all compatible with the diagnosis of PG. Treatment was started with intravenous methylprednisolone at a dose of 1 mg/kg/day and empirical antibiotic therapy, in addition to local wound care. The patient made a slow but favourable recovery (Fig. 1C).

Several cases of postoperative PG have been published in patients with IBD, but our case would be the second to be described of PG with onset over ten years after a restorative proctocolectomy due to UC.[5] A significant association has been found between the presence of a permanent stoma and PG, with this combination being found in up to 28% of cases. In this scenario, lesions usually appear in the early postoperative period, probably due to the pathergy phenomenon in response to surgery. In fact, some authors define this finding as a specific variety of PG: peristomal PG.[4] However, in our patient, the PG took place ten years after the surgery, after a surgical technique without stoma, in a location not related to the site of the surgery. It is often accepted that the development of PG is independent of IBD, and it can occur after long periods of remission of the disease, but for it to occur in a patient who has undergone this type of intervention, and so many years later, is extremely unusual.

Inflammatory involvement of the colon, either in the form of UC or CD, seems to be a key condition for the disorder to occur. A significant association has also been found between PG and a history of extraintestinal eye disease such as uveitis, as occurred in our patient.[2]

This, added to the fact that the patient suffered an autoimmune manifestation such as uveitis, years after surgery, suggest that UC and PG could share a common autoinflammatory and immunological predisposition. The pathophysiological mechanisms behind the development of PG are not known. The most accepted theories include various alterations of the physiological inflammatory cascade, which would culminate in dysregulation of neutrophil chemotaxis, with these alterations also having been found in IBD.[1,2] Some authors suggest a cross-immunogenicity between colon and skin antigens, a theory that would explain the relationship between PG and IBD with colonic involvement.[1]

The relevance of this case lies in the risk of patients with UC having significant extraintestinal autoimmune manifestations such as PG even years after undergoing a curative intervention for their intestinal disease.

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Mónica Llorente Barrio[a,*], Santiago García López[a], Yolanda Gilaberte Calzada[b], Rosario María Monzón Báez[a], Diego Casas Deza[a], Luis Javier Lamuela Calvo[a]

[a] *Servicio de Aparato Digestivo, Hospital Universitario Miguel Servet, Zaragoza, Spain*
[b] *Servicio de Dermatología, Hospital Universitario Miguel Servet, Zaragoza, Spain*

* Corresponding author.
E-mail address: mllorentebarrio@gmail.com
(M. Llorente Barrio).
2444-3824/