Funding

No funding was received for this work.

Conflicts of interest

The authors declare that they have no conflicts of interest.

References

1. Wu H, Meng YH, Lu P, Ning HY, Hong L, Kang XL, Duan MG. Epithelioid inflammatory myofibroblastic sarcoma in abdominal cavity: a case report and review of literature. Int J Clin Exp Pathol. 2015;8:4213–9.
2. Bai Y, Jiang M, Liang W, Chen F. Incomplete intestinal obstruction caused by a rare epithelioid inflammatory myofibroblastic sarcoma of the colon: a case report. Medicine (Baltimore). 2015;94:e2342, http://dx.doi.org/10.1097/MD.0000000000002342.
3. Spunt SL, Francotte N, De Salvo GL, Chi YY, Zanetti I, Hayes-Jordan A, et al. Clinical features and outcomes of young patients with epithelioid sarcoma: an analysis from the Children's Oncology Group and the European paediatric soft tissue Sarcoma Study Group prospective clinical trials. Eur J Cancer. 2019;112:98–106, http://dx.doi.org/10.1016/j.ejca.2019.02.001.
4. Thway K, Jones RL, Noujaim J, Fisher C. Epithelioid sarcoma: diagnostic features and genetics. Adv Anat Pathol. 2016;23(1):41–9, http://dx.doi.org/10.1097/PAP.0000000000000102.
5. Needs T, Fillman EP. Cancer, Epithelioid Sarcoma. In: StatPearls. Treasure Island (FL). StatPearls Publishing; 2020.

Mario Serradilla-Martín[a,*], Ana Palomares-Cano[b], Ana María Carrillo-Colmenero[c], César Luis Ramírez-Tortosa[d]

[a] *Instituto de Investigación Sanitaria Aragón, Servicio de Cirugía General, Hospital Universitario Miguel Servet, Zaragoza, Spain*
[b] *Servicio de Cirugía General, Hospital Universitario Miguel Servet, Zaragoza, Spain*
[c] *Servicio de Radiodiagnóstico, Complejo Hospitalario de Jaén, Jaén, Spain*
[d] *Unidad de Gestión Clínica de Anatomía Patológica, Hospital Clínico San Cecilio, Granada, Spain*

* Corresponding author.
E-mail address: marioserradilla@hotmail.com
(M. Serradilla-Martín).

Pyoderma gangrenosum solved by ustekinumab therapy☆

Pioderma gangrenoso resuelto mediante terapia con ustekinumab

Pyoderma gangrenosum is a neutrophilic ulcerative-necrotic dermatosis which manifests as a sterile nodule or pustule that rapidly progresses to a painful purulent ulcer with irregular, serpiginous, oedematous, violaceous and undermined borders. They can occur in any location, but the most striking and common forms occur on the legs.

It is the most serious skin manifestation associated with inflammatory bowel disease (IBD), although it can be associated with other systemic conditions such as myelodysplastic syndromes, monoclonal gammopathy, leukaemia and rheumatoid arthritis. In general, it is more common in ulcerative colitis (UC) than in Crohn's disease (CD).[1] Although it usually coincides with the reactivation of underlying IBD, that is not always the case.

We present the case of a patient with CD and pyoderma gangrenosum that responded after treatment with ustekinumab.

The patient was a 29-year-old woman with complex fistulising CD, with total colectomy and terminal ileostomy performed eight years earlier, and a history of pyoderma gangrenosum which had previously responded to ciclosporin and local measures. She was referred with a two-month history of very painful pretibial skin lesions on both legs, accompanied by fever. At that time, the patient was being treated with adalimumab (with an intensified regimen of

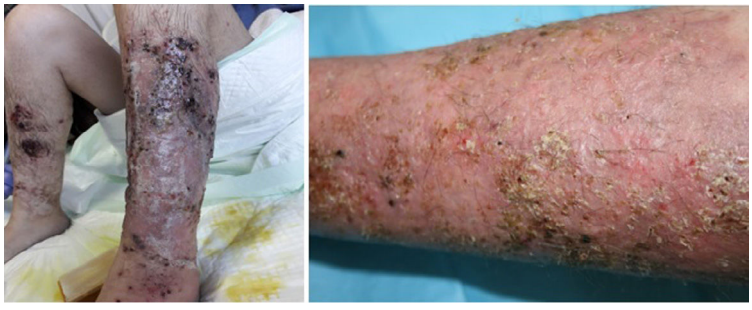

Figure 1

☆ Please cite this article as: López González J, Lázaro Sáez M, Moreno Moraleda I, Hernández Martínez Á. Pioderma gangrenoso resuelto mediante terapia con ustekinumab. Gastroenterol Hepatol. 2021;44:299–300.

80 mg/2 weeks) with good control of her gastrointestinal symptoms.

On admission, a biopsy was taken of the lesion and the exudate cultured. The culture showed a carbapenemase-producing *Pseudomonas aeruginosa*, sensitive to piperacillin-tazobactam and ciprofloxacin. A histological study showed a necrotic epidermis with abscess formation and granulation tissue with fibrin and blood deposits, acute chronic infiltrate with a predominance of neutrophils, without the presence of granulomas, in addition to mild spongiosis and mixed perivascular inflammatory infiltrate in the adjacent epidermis. All this is compatible with the diagnosis of pyoderma gangrenosum. The patient was started on targeted antibiotic therapy, corticosteroid therapy and local measures (permanganate, clobetasol and mupirocin), and wound dressing sessions were booked in the dermatology operating theatre.

The patient remained in hospital for 15 days, when she was discharged with partial improvement of the lesions. However, at her one-month outpatient follow-up, the skin lesions were found to have worsened, although she remained asymptomatic from a gastrointestinal point of view. With the aim of controlling her dermatological symptoms, it was decided to discontinue adalimumab and start ustekinumab, with a first dose of 260 mg IV, after which she improved significantly, with no observed recurrence of her gastrointestinal symptoms (Fig. 1). The patient is currently on maintenance therapy with ustekinumab (90 mg/8 weeks), and free of both gastrointestinal and dermatological symptoms.

The diagnosis of pyoderma gangrenosum is based on compatible, but not specific, clinical and histological findings, as well as the ruling out of other possible diagnoses. The differential diagnosis varies depending on the form of presentation: ulcerative, pustular, bullous or superficial. Of all the disorders we have to consider, the main one is Sweet's syndrome which, like pyoderma, is a neutrophilic dermatosis that also occurs frequently in IBD. Other disorders to include in the differential diagnosis are vasculitis, bacterial, fungal (sporotrichosis) and viral infections, neoplastic diseases (such as cutaneous squamous cell carcinoma and skin metastases), systemic lupus erythematosus, Behçet's disease, cutaneous Crohn's disease and ulcerated necrobiosis lipoidica.[2]

Traditionally, immunosuppressants such as ciclosporin, azathioprine or mycophenolate mofetil, and antibiotics such as doxycycline have been used to treat pyoderma gangrenosum. More recently, biological drugs have revolutionised the treatment of this disease, the main ones used being infliximab, etanercept, adalimumab and certolizumab.[3]

Ustekinumab, the latest biologic drug approved for use in CD, is emerging as a valid alternative. However, only isolated cases of pyoderma treated with this drug have been described since Guenova et al. did it for the first time in 2011, observing that IL-23 was overexpressed in the biopsy of the lesion compared to biopsies of healthy skin.[4]

In the case of our patient, ustekinumab achieved a significant improvement in her pyoderma gangrenosum, previously refractory to anti-TNFs, in addition to maintaining remission from a gastrointestinal point of view, all of which is extremely interesting in terms of the potential future development of the drug in this disease.

References

1. Ahn C, Negus D, Huang W. Pyoderma gangrenosum: a review of pathogenesis and treatment. Exp Rev Clin Inmunol. 2018;14:225–33.
2. Plumptre I, Knabel D, Tomecki K. Pyoderma Gangrenosum: a review for the gastroenterologist. Inflamm Bowel Dis. 2018;24:2510–7.
3. Soto F, Vera-Kellet C. Pioderma gangrenoso: terapias clasicas y emergentes. Med Clin. 2017;149:256–60.
4. Guenova E, Teske A, Fehrenbacher B, Hoerber S, Adamczyk A, Schaller M, et al. Interleukin 23 expression in pyoderma gangrenosum and targeted therapy with ustekinumab. Arch Dermatol. 2011;147:1203–5.

José López González*, Marta Lázaro Sáez, Isabel Moreno Moraleda, Álvaro Hernández Martínez

Unidad de Gestión Clínica del Aparato Digestivo, Hospital Universitario Torrecárdenas, Almería, Spain

* Corresponding author.
E-mail address: pepe_1993_17@hotmail.com (J. López González).

Experience with tofacitinib in patients with refractory ulcerative colitis☆

Experiencia local con tofacinitib en pacientes con colitis ulcerosa refractaria

Tofacitinib is a Janus kinase (JAK) inhibitor, particularly of JAK 1 and JAK 3, that works at an intracellular level blocking the activity of multiple cytokines. Unlike other biological drugs, which act by blocking a specific cytokine, tofacitinib is also administered orally, making it an easier and possibly preferred route of administration for patients. It has other advantages, such as its short half-life, speed of action and low molecular weight which means that it does not induce immunogenicity.

We carried out a descriptive retrospective study at Hospital Torrecárdenas, analysing the seven patients with ulcerative colitis treated to date with tofacitinib. The main characteristics of these patients are shown in Table 1.

Of the seven patients treated, five were male (75%) and two female (25%). They were aged from 26 to 67 (mean age 50.4, median 56). Four patients had pancolitis and three had left-sided colitis. None of them presented extraintestinal manifestations.

All the patients had previously been treated with at least three biologic drugs (2 anti-TNFs and vedolizumab). All patients had severe endoscopic activity (Mayo endoscopic subscore of 3) and histological activity (Rutter grade 4).

☆ Please cite this article as: López González J, Hernández Martínez Á, Lázaro Sáez M, Vega Sáenz JL. Experiencia local con tofacinitib en pacientes con colitis ulcerosa refractaria. Gastroenterol Hepatol. 2021;44:300–301.