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Case Report: Pyoderma Gangrenosum in IBD Treated with Tofacitinib

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Abstract

Pyoderma gangrenosum is a rare ulcerating skin disease often presenting as an extra intestinal manifestation of IBD and is difficult to manage. Treatment options for pyoderma gangrenosum include steroids, calcineurin inhibitors and anti TNF agents. Here in, we report a case of recurrent Pyoderma gangrenosum with Ulcerative Colitis that was successfully managed with Tofacintib, a JAK inhibitor.

Keywords: Pyoderma Gangrenosum; Ulcerative Colitis; Tofacitinib

Introduction:

Pyoderma gangrenosum (PG) is a reactive non-infectious inflammatory dermatosis falling under the spectrum of the neutrophilic dermatoses. 30% of Pyoderma gangrenosum cases have associated IBD [1]. Clinically it presents as a sterile papule or pustule which rapidly progresses to a purulent ulcer with irregular, serpiginous, edematous, violaceous and undercut edges [2]. Immunosuppression is the mainstay of treatment for pyoderma gangrenosum. Treatment options include corticosteroids, calcineurin inhibitors and anti TNF agent's [3]. Here in, we report a case of ulcerative colitis with refractory PG which improved on Tofacitinib therapy which is an oral JAK inhibitor.

Case Report:

A 31 year old man who is a known case of IBD – UC diagnosed 3 years prior, stopped treatment 2 years ago. He presented with complaints of painful, non-healing erythematous ulcer over dorsum left foot (Figure 1).

Figure 1: Ulcerated PG over dorsum of foot

The skin lesion started innocuously as a vesicle, allegedly after dropping hot water on his left foot and developed into a pustule within 3 days and eventually became a large ulcer over 2 weeks. The ulcer measured about 8 x 6cms, over the dorsum of left foot with active pus discharge. Active toe movements were present. X-ray showed soft tissue irregularity in dorsal aspect of mid foot with normal density of bones. He approached a plastic surgeon without revealing the history of IBD. He underwent debridement, incision and drainage along with splitskin grafting. He was discharged with oral antibiotics.

Three months later he presented with recurrence of the foot ulcer with purulent discharge. He was admitted for the same and was being managed with negative pressure wound therapy and ivy antibiotics. During the admission he developed 2-3 episodes of rectal bleed. In view of the lower GI bleed he was taken up for sigmoidoscopy which showed severe proctosigmoiditis (Figure 2) with UCEIS- 7/8, Mayo Score - 3, biopsy revealed chronic, severe active inflammation (Figure 3).

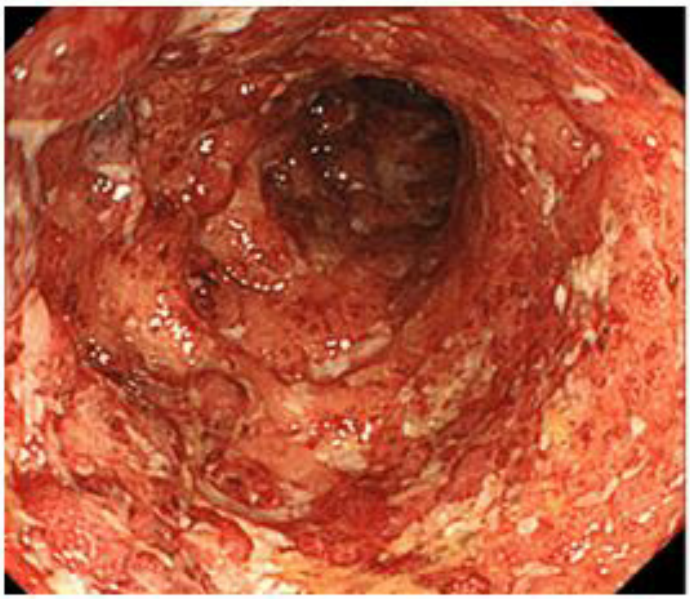

Figure 2: Severe colitis on Colonoscopy

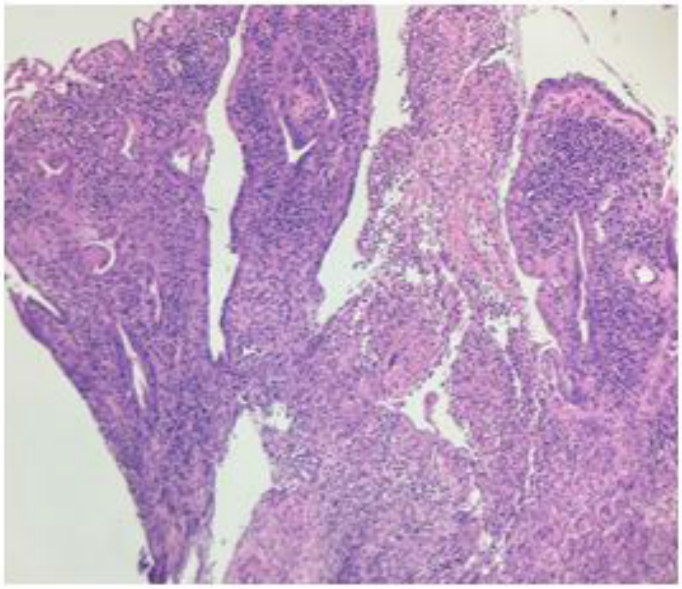

Figure 3: HPE showing severe colitis.

After interviewing the patient, he confirmed that he was diagnosed with IBD-UC in 2018 and had been treated with Mesalamine and corticosteroids until June 2022, later patient stopped medication due to symptom resolution.

Biopsy of foot ulcer revealed granulation tissue and dense neutrophilic infiltrate (Figure 4), Tissue culture from wound site was sterile and gene Xpert was negative. Considering the background history of IBD –UC, clinical presentation and histopathology of foot ulcer a diagnosis of pyoderma gangrenosum was considered. Patient was started on TOFACITINIB 10mg BD and Mesalamine 1.2 g BD for severe colitis and pyoderma gangrenosum. He then underwent a repeat split skin grafting and was discharged with oral antibiotics.

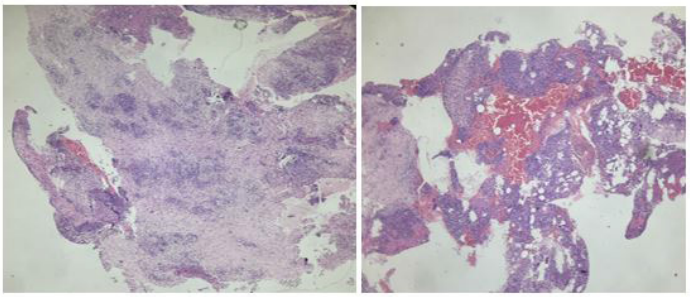

Figure 4: HPE showing Neutrophil inflammation in dermis.

After 2 months follow up, foot ulcer healed and the skin graft remained healthy. He also achieved clinical and endoscopic remission of ulcerative colitis.

Discussion:

21–54% of IBD patients have at least one "extra-intestinal manifestation" (EIM). Of these patients, about one-third are diagnosed with a cutaneous manifestation. The most common cutaneous manifestations are erythema nodosum and pyoderma gangrenosum (PG). The most common cutaneous manifestations are erythema nodosum and pyoderma gangrenosum (PG) Overall prevalence of pyoderma gangrenosum in IBD is 0.4% - 2.6 % [4].

PG has been reported in 1–10% of UC patients and 0.5–20% of CD patients (Figure 5).

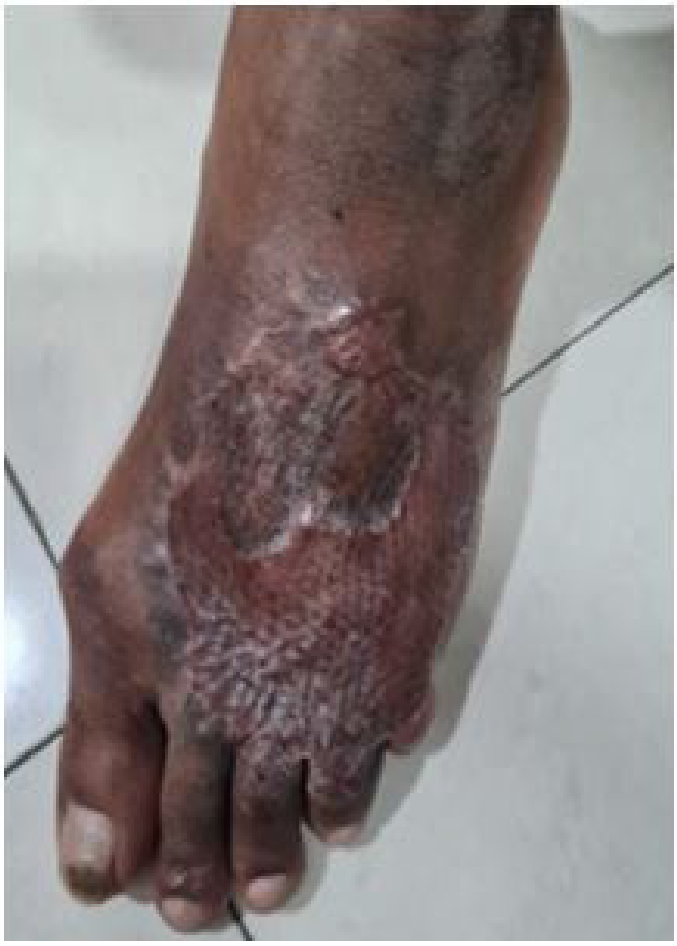

Figure 5: Healed ulcer after skin grafting.

Clinically Pyoderma gangrenosum (PG) frequently begins as an erythematous pustule or nodule rapidly developing into deep ulcers, irregular violaceous edges and purulent material in the ulcer ground which is usually sterile on culture. PG is found mainly on the legs but may also occur in 4–8% on the head and neck and in 4–5% on the trunk. The ulcers can be solitary or multiple, unilateral, or bilateral, and can range in size from several centimeters to an entire limb [6].

Treatment options for pyoderma gangrenosum include oral steroids, prednisolone (40–60 mg/day and tapering, cyclosporine (initial target blood levels of 150–300 ng/ml), tacrolimus (initial target blood levels between 10 and 15 ng/ml) or anti-TNF antibodies (infliximab and adalimumab). Topical tacrolimus is successful in the treatment of early lesion. (0.1% ointment 2 times daily) [6].

In our case, patient was a known case of IBD – Ulcerative colitis who had stopped treatment and now presented with a foot ulcer. On evaluation he was found to have severe colitis on colonoscopy. Diagnosis of pyoderma gangrenosum was confirmed based on criteris proposed by the Delphi consensus of international experts [7]. Our patient had a few constraints and couldn't be considered for biologicals. Hence we considered tofacitinib (JAK inhibitor) as a possible therapy for his colitis and pyoderma gangrenosum. Response to tofacitinib 10 mg BD after 2 months was adequate.

Tofacitinib is an oral JAK-1 and JAK-3 inhibitor that has been approved for the treatment of rheumatoid arthritis and ulcerative colitis and currently is being evaluated for plaque psoriasis and inflammatory arthritis. Upregulation of the JAK-STAT pathway has been demonstrated in immunohistochemical analyses of skin biopsies from patients with PG and EN [8]. Evidence for use of tofacitinib in pyoderma gangrenosum has also been reported from 2 case series with satisfactory results [9,10].

Conclusion:

PG is a rapidly progressing, debilitating disease that is difficult to manage. Current treatment options are not adequate for treatment of pyoderma gangrenosum either due to lack of efficacy, requirement of high dosage, adverse effects or due to high financial costs. Hence tofacitib could be considered as a possible treatment option for pyoderma gangrenosum in view of the oral route of administration, affordability and limited adverse events. Randomised controlled trials and long term studies will however be needed before tofacitinib can be confirmed as a first line option for IBD eith pyoderma gangrenosum and other extra intestinal manifestations.

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