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Management of Pyoderma Gangrenosum in a Patient with Crohn's Disease

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There is no definitive guidelines for the management pyoderma gangrenosum (PG); it is mainly guided by small studies and case reports.

We are reporting a case of a middle aged man who presented with pyoderma gangrenosum who was diagnosed with Crohn's disease (CD). His condition was resistant to the initial systemic steroid therapy but responded well to infliximab.

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Pyoderma gangrenosum (PG) is an uncommon inflammatory dermatosis characterized histo-pathologically by accumulation of neutrophils in the skin and usually presents with an acutely painful, rapidly enlarging deep ulcer. It is neither an infection nor gangrenous, it is probably an inappropriate reaction to an unidentified stimulus[1-3]. Although, rare cases of PG have presented as idiopathic; in most of the cases it has been associated with inflammatory bowel diseases, hematological conditions (leukemias and polycythemia) and collagen vascular diseases[4].

The aim of this presentation is to report a resistant case of pyoderma gangrenosum associated with Crohn's disease.

THE CASE

A thirty-nine-year-old healthy gentleman initially had a painful papule in the medial aspect of the left thigh. The papule was ruptured by the patient and started to ulcerate. This caused him to seek medical advice in a local health centre where dressing was done.

The patient started to have bloody diarrhea, 8-10 times/day. One week later, he presented to A/E department with diarrhea and was treated as infective diarrhea. After two days, he presented again to the A/E department complaining of painful left thigh ulcer and was admitted in the surgical ward with tentative diagnosis of unspecified ulcer; swabs and biopsies were taken and debridement was done.

However, the ulcer continued to increase in size and his bowel symptoms were persistent; therefore, internal medicine was consulted. Flexible sigmoidoscopy was performed and multiple biopsies were taken with an impression of inflammatory bowel disease. Systemic steroid therapy has started and dermatology consultation was recommended.

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The dermatology consultation revealed a clinical diagnosis of pyoderma gangrenosum and advised to start immunosuppressive therapy in the form of cyclosporine because the patient was not showing any regression in his ulcer despite of being on systemic steroid for 7 days, see figure 1.

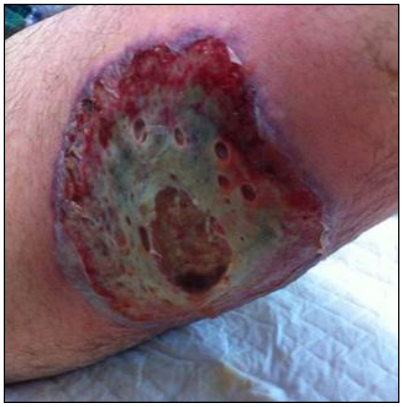

Figure 1: Pyoderma Gangrenosum on the Left Thigh before Starting Infliximab, Typical Appearance of Ulcerative Skin with Violet Border

The sigmoid biopsies revealed Crohn's disease and the patient was transferred to the medical ward. He continued to receive steroid therapy with antibiotics and wound dressing was done with silver sulfadiazine.

The patient did not receive immunosuppressive therapy due to positive wound culture of *ESBL E.coli*. However, due to the absence of systemic and local signs of infection and two negative wound swabs, the condition was considered as contaminated PG ulcer and the patient eventually had started infliximab.

The patient had improved dramatically after the first dose of infliximab for both GI and skin symptoms. His GI symptoms had subsided on the fourth day along with good healing of PG, see figure 2.

The patient had an attack of bloody diarrhea for two weeks last year, which resolved spontaneously. He had a negative family history of gastrointestinal-dermatological diseases and he was not known to have any medical illnesses.

Endoscopy: The initial flexible sigmoidoscopy revealed active colitis with crypt abscesses and cobblestone appearance. A follow-up of complete colonoscopy was performed after four doses of infliximab; it showed mild patchy inflammation of the sigmoid and the distal descending colon with multiple small pseudopolyps. The rest of bowel mucosa appeared normal.

A follow-up of the patient showed complete healing of PG with residual scarring and has no GI symptoms, see figure 3.

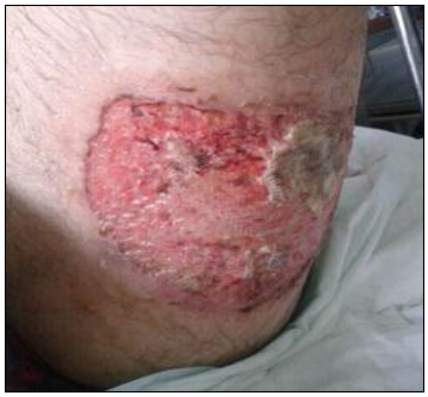

Figure 2: Pyoderma Gangrenosum Four Days after the First Dose of Infliximab

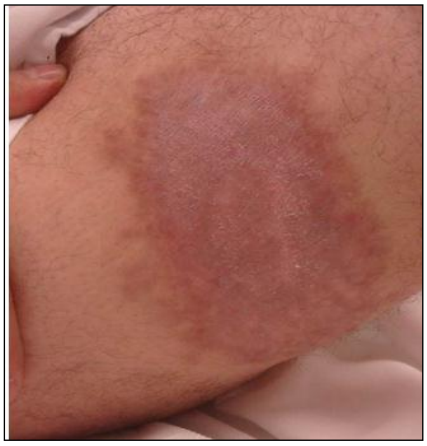

Figure 3: The Healed Pyoderma Gangrenosum with Scarring after Nine Months

DISCUSSION

Reviewing the literature, three lines of management have been suggested in the treatment of PG. The first line therapy would include local care, topical and systemic therapy.

Local care with gentle cleanser and sterile saline along with dressing is advised to promote a moist wound environment. Intralesional corticosteroids injections have been reported to be effective[1]. Beside steroids, topical calcineurin inhibitors in the form of tacrolimus have been also reported to be helpful[3].

The decision to administer systemic treatment is usually made after failure of the local treatment, the presence of deep ulcers and the association of systemic disease. Oral prednisolone or intervenous pulse steroids are usually used together with steroid sparing agent, such as Mycophenolate mofetil and Azathioprine. Systemic Cyclosporine could be used as an alternative for steroids[3].

The second line and adjunctive therapies would include dapsone and minocycline as anti-tumor necrosis factors. The most used anti TNF agent is infliximab; a chimeric antibody against tumor necrosis factor alpha[1].

In refractory disease, intervenous immunoglobulin and alkylating agents have been suggested as a third line of therapy[1,3].

In our case, the patient had presented with bloody diarrhea (CD) and skin manifestation (PG), steroid therapy was used with no improvement, infliximab was started with significant improvement from the first dose in both GI and skin symptoms. There have been multiple cases reported with a similar scenario[5-6].

In a prospective clinical trial, 30 patients with CD were enrolled. They were treated with infliximab. The trial showed an overall short-term efficacy of 100% in the treatment of all extraintestinal manifestations[7].

CONCLUSION

The use of anti-tumor necrosis factors alpha is to be considered as a good armament in the early stages of treating pyoderma gangrenosum with Crohn's disease.

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