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Case Report

Idiopathic genital pyoderma gangrenosum successfully treated with infliximab biosimilar

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Abstract Pyoderma gangrenosum (PG) is an atypical, chronic, sterile pustular rapidly progressing ulcerating condition categorized as neutrophilic dermatosis, whose etiology remains a mystery. Other ulcerating wounds can also imitate PG, and cause a difficulty in diagnosing the condition and starting appropriate management. In our case, we had a 22-years old female with genital PG. PG has no specific clinical, histologic or lab findings, therefore diagnosis is based on exclusion of all other diagnostic possibilities, especially infectious causes. The clinical history, examination findings and histopathology confirmed it as genital pyoderma gangrenosum. Treatment with subcutaneous infliximab biosimilar led to complete healing.

Key words
Pyoderma gangrenosum; Neutrophilic dermatoses; Infliximab biosimilar.

Introduction

Pyoderma gangrenosum (PG) is characterized as a non-infectious inflammatory skin condition.[1] It presents in four primary clinical forms: ulcerative, pustular, bullous, and vegetative. Almost 50% of PG patients have an underlying systemic illness that may precede, follow, or occur concurrently with PG.[2] About 50% of cases are idiopathic. Although genital PG is rare, documented cases emphasize the importance of considering it in the differential diagnosis for persistent genital ulcers, especially when associated with underlying systemic disease. Often, initial treatments are ineffective, and extensive laboratory investigations are necessary to rule out other diseases.[3] We report the case of a young girl who presented with a genital ulcer diagnosed as PG and was successfully treated with an infliximab biosimilar injection.

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Case report

A 22-years old married lady presented with painful genital ulcers for 1 year with disease aggravation for last 6 months. She developed these genital ulcers 3 months after her marriage that started off with a solitary painful pustule on her genitalia, rapidly evolving into a painful ulcer with foul smelling serosanguinous discharge. Later on, the ulcer increased in size as well as number and all the lesions followed the same evolution. There was history of intermittent low-grade fever that was relieved by over-the-counter antipyretics and single episode of oral ulcers and erythema in left eye that lasted for 2 days and resolved spontaneously. She also gave history of joint pains i.e. bilaterally symmetrical involvement of knee and ankle joints, however there was no associated morning stiffness or limitation of movements.

There was no history of vesicles, itching, vaginal discharge, weight loss, lumps and bumps in the body or long-term eye complaints. Also, there was no history of any extramarital relationship.

Figure 1 Multiple phagedenic ulcers with undermined edges

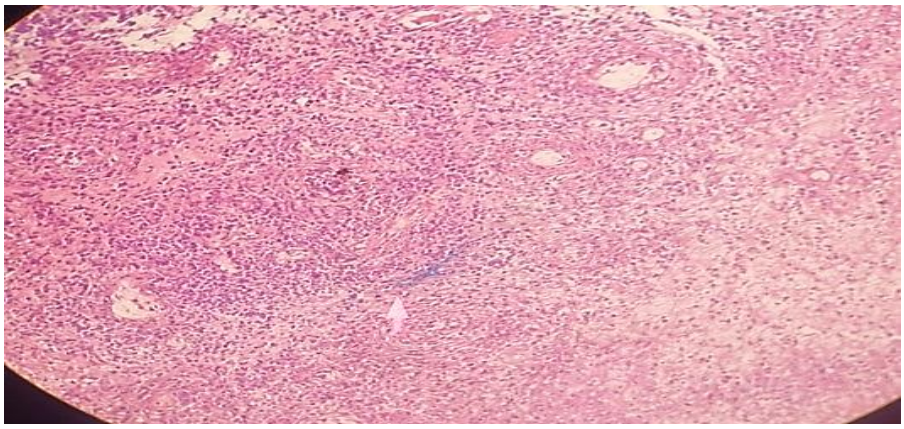

Figure 2 Histopathology: hematoxylin and eosin stained section (40x) showing moderate to dense mixed infiltrate comprising mainly of neutrophils.

Neither did her spouse had any discharge, genital lesion or history of multiple sexual partners. She took multiple medical and non-medical treatments including antibiotics, antivirals and pain killers but her lesions never settled.

Her general physical examination was normal except for pallor. Her genital examination revealed multiple, tender, round to oval ulcers with a gangrenous slough, undermined violaceous edges and non-adherence to underlying skin involving labia majora, minora and perianal skin of both sides. Few atrophic cribriform scars were also noted along with perilesional hyperpigmentation **(Figure 1)**. However, there was no regional lymphadenopathy. Pathergy test was negative.

Laboratory investigations objectified anemia with raised ESR, CRP and platelets. ANA profile was negative. She was screened for HIV, Hepatitis B and C and sexually transmitted diseases, and they were all negative. Smear for Donovan's bodies was also negative. Urine analysis and ultrasound abdomen and pelvis was unremarkable. Tissue microbial culture did not detect any bacteria, fungi or atypical mycobacteria.

However, skin biopsy revealed a mixed dermal inflammatory infiltrate comprising predominantly of neutrophils **(Figure 2)** that favor neutrophilic dermatosis as seen in pyoderma gangrenosum and didn't have features of any other disease considered as differential diagnosis.

The histopathological features of PG, though not particular, are useful in ruling out other causes of ulceration.[4]

The patient was treated with oral prednisolone 0.75mg/kg/day to which she responded well and after 2 weeks of treatment her lesions started to heal **(Figure 3)**. However, she was not willing to take steroids because of long term side effects. Therefore, she was treated with subcutaneous injection infliximab biosimilar-dyyb, 120mg weekly for 5 weeks followed by another 5 doses fortnightly. Within 2 months of treatment, her lesion resolved completely **(Figure 4)** and she was put on maintenance dose of injection infliximab 120mg once monthly. Patient was kept in follow up for one year and there was no relapse. Patient's photographs are being shared here with her consent.

Discussion

Pustular lesions can be the first cutaneous manifestation in pyoderma gangrenosum and skin hyperreactivity and pathergy is often associated. In PG, genital ulcers appear similar

Figure 3 Improvement after 2 weeks of treatment with oral steroid.

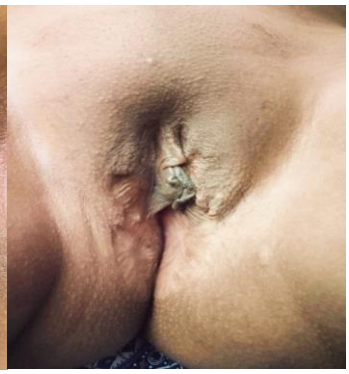

Figure 4 Improvement after 2 months of treatment with injection infliximab

to those of the skin that basically are rapidly evolving tender ulcers with undermined violaceous borders that respond swiftly to systemic steroids and heal with atrophic cribriform scarring.[5]

It has also been estimated that approximately 50% cases of PG are associated with inflammatory bowel disease, inflammatory arthritis, solid organ malignancies and hematological malignancies.[6] However most cases are idiopathic.[7] In our case, based upon proposed diagnostic criteria of classic ulcerative pyoderma gangrenosum, our patient fulfilled the following features: rapid progression of a painful ulcer with irregular violaceous undermined edges, clinical finding of cribriform scarring, histopathologic evidence and rapid response to systemic steroids. For severe disease, the mainstay of therapy is systemic steroids however, there is gaining evidence that anti-TNF biologic therapy should be considered as first line in affording patients. As our patient was not willing for the use of systemic steroids, we managed her with injection infliximab. In the recent advent, tumor necrosis alpha blockers and other biologics have been found particularly effective in treating PG associated with inflammatory bowel disease[8] but we noticed an excellent response to treatment with injection infliximab biosimilar in idiopathic genital pyoderma gangrenosum without a relapse.

Conclusion

This case emphasizes importance of early recognition of PG to provide a timely diagnosis, appropriate management and an excellent response to injection infliximab biosimilar-dyyb in halting the disease process.

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Declaration of patient consent The authors certify that they have obtained all appropriate patient consent.

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Conflict of interest Authors declared no conflict of interest.

Authors' contribution

ANC,JTB,SB,AA,FA: Diagnosis and management of the case, critical review of the manuscript, has given final approval of the version of the manuscript to be published.

HT,UA: Identification and management of the case, manuscript writing, has given final approval of the version of the manuscript to be published.

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