Orogenital Ulcers of Pyoderma Gangrenosum Resembling Sexually Transmitted Disease

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ABSTRACT
Pyoderma gangrenosum is a non-infectious neutrophilic dermatosis that may be either idiopathic or associated with some underlying diseases such as inflammatory bowel diseases, SLE, sarcoidosis, vasculitis etc. It can occur in any part of the body but pyoderma gangrenosum involving oral cavity and genital regions presenting as orogenital ulcers would be clinically quite difficult to be distinguished from Sexually Transmitted Diseases (STDs). We present such a case that was initially managed on the lines of sexually transmitted diseases, which later on came out to be pyoderma gangrenosum, after excluding all other diseases having almost same clinical presentations, on the basis of laboratory results and histopathology.

Key Words: Pyoderma gangrenosum. Orogenital ulcers. Sexually transmitted diseases.

INTRODUCTION
Pyoderma gangrenosum (PG) is a sterile inflammatory neutrophilic dermatosis.¹ It has an idiopathic form as well as the one associated with some underlying diseases such as inflammatory bowel disease, rheumatoid arthritis, ankylosing spondylitis, leukemias, myelodysplasia, sarcoidosis, human immunodeficiency syndromes and solid tumors.²

Pyoderma gangrenosum usually presents as eruptions that starts as an inflamed nodule. It then breaks down centrally and progresses rapidly to an ulcer with an indurated and undermined purplish edges. These lesions usually have predilection for pretibial areas, however, PG can affect any part of the body including oral and genital mucosa.¹ On the other hand, the one occurring in orogenital region will be quite difficult to be distinguished from sexually transmitted diseases.³ Therefore, proper history taking, clinical examination and histopathological examination are required for correct and early diagnosis.

CASE REPORT
An 18 years old boy with no known previous co-morbid condition, history of alcohol intake and unprotected sexual intercourse, presented with the complaints of high grade fever with chills along with painful oral and genital ulcers for the last one week. Ulcers were painful to the extent that he could not swallow anything nor could walk.

Examination of the oral cavity revealed multiple small sized ulcers over the soft palate, tongue and pharynx along with white coat (Figure 1). Genital examination showed, he had one large shallow ulcer over the scrotum and 2 over the shaft of penis along with pustular lesions on the genitalia without inguinal lymphadenopathy (Figure 2). Rest of the examination was unremarkable.

Based upon the history and clinical examinations, he was investigated for sexually transmitted diseases. Workup for STDs like TPHA, RPR, HIV antibodies and swab for gram stain and culture and for autoimmune diseases like ANA and ANCA were sent to laboratory. His initial laboratory reports showed lymphocytosis and CRP of 56 mg/dL and ESR of 20 mm after 1st hour. Based on these results, he was started empirically on oral acyclovir, benzathine penicillin and ceftriaxone considering that he has some STDs. Despite this treatment, his condition did not improve even after 3 days of treatment, and he remained febrile. All the workup for STDs and autoimmune diseases came out to be negative. Therefore, further workup in the form of

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scrotal biopsy was sent for histopathology and Behcet’s syndrome at this time was ruled out by ophthalmologist as there was no evidence of uveitis. It was further clarified by biopsy report which showed skin ulceration and necrotizing suppurative inflammation including sheets of neutrophils at the margin of lesion with luminal fibrin deposition. Findings were suggestive of pyoderma gangrenosum. Therefore, all the antibiotics were stopped and started on with oral prednisolone 40 mg once a day. There was a poor response to steroids earlier on. Cyclosporine was then added at a dose of 6 mg/kg. He responded to the treatment and became afebrile and ulcers started to heal. He was discharged after 10 days of treatment. At that time, his general condition was much improved.

He was later on put on oral prednisolone in tapering doses. During follow-up visits, cyclosporine was discontinued due to derangement of liver functions. He was also advised to apply local antibiotic ointment and do antiseptic dressing to prevent secondary bacterial infection of the genital ulcers.

DISCUSSION

Pyoderma gangrenosum is a non-infectious neutrophilic dermatosis and was first described in 1930. Pyoderma gangrenosum can affect all age groups and its incidence rate is 3 - 10 per millions per year. The cause of pyoderma gangrenosum is unknown and is believed that the dysregulation of immune system may be involved; it mostly occurs with systemic diseases but can occur independently as well. PG can involve any part of body including skin, oro-genital regions and organ systems like liver, heart, CNS, spleen, bones, lymph nodes and eyes. It has different forms like ulcerative, pustular, bullous and vegetative and each form can develop into mixed dermal infiltrate. The treatment of choice is ulceration, superficial dermal edema and dense abscess which later evolved into epidermal necrosis, ulceration, superficial dermal edema and dense mixed dermal infiltrate. The treatment of choice is corticosteroids but in corticosteroids resistant cases, or in cases of adverse effects, cyclosporin A is the treatment of choice. Other alternative therapeutic options are mycophenolate mofetil, tacrolimus, infliximab, plasmapheresis and Intravenous Immunoglobulin (IVIG).

Pyoderma gangrenosum of oral cavity and genital region must be differentiated from sexually transmitted diseases like syphilis, genital herpes, HIV, chancroid. It is quite difficult to diagnose earlier on due to the fact that there are no specific immunofluorescence markers or laboratory tests that can pinpoint pyoderma gangrenosum. Syphilis is a sexually transmitted disease and its primary stage can present as painless genital ulcer. It appears as dull red macule and later becomes papule and then erodes to form chancre. It can be diagnosed with RPR and TPHA which was negative in this case. Genital herpes presents as irritative vesicles that ruptures to form small tender ulcers on external genitalia. It can be diagnosed by analyzing vesicular fluid for DNA by PCR, which was not sent to laboratory in this case due to lack of facilities. Chancroid is another sexually transmitted disease that causes single or multiple ulcers with ragged undermined edges. It can be diagnosed with microscopy and culture of scrapings from ulcer, which was negative in this case. HIV can present as multiple genital and oral ulcers. It can be diagnosed with antibodies against HIV which was negative in this case.

REFERENCES