Pyoderma gangrenosum masquerading as Donovanosis

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Abstract

Pyoderma gangrenosum (PG) is a rare inflammatory disorder of unknown etiology characterized by neutrophilic infiltration of the dermis and destruction of tissue. PG is diagnosed after excluding more commonly occurring condition presenting with similar manifestation. Though PG has been reported to occur over the genitalia, it rarely presents with concurrent involvement of the groin. Herein, we present a case of PG masquerading as Donovanosis.

Key words: Donovanosis, genitalia, pyoderma gangrenosum

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare inflammatory disorder of unknown etiology characterized by neutrophilic infiltration of the dermis and destruction of tissue. It is more common in females between 40 and 60 years with a prevalence of 3 cases per million per year.¹² PG is diagnosed after excluding more commonly occurring condition presenting with similar manifestation. Herein, we present a case of PG masquerading as Donovanosis.

CASE REPORT

A 70-year-old male patient reported to our outpatient department with bilateral erosive lesions over the groin since 5 months. The lesions initially started as a small erosion which rapidly progressed over 1 month to involve the both sides of the groin and thereby remain as such. Ulcers were associated with intense pain and foul smelling discharge. There was no history of vesiculation, joint pain, or eye complaints. The patient denied any history of sexual exposure, drug use, or alcohol consumption, diabetes or hypertension. He reported many previous consultations due to his current disease, but the lesions failed to heal despite treatment.

On cutaneous examination linear ulcers with well-defined edges were present on either side of the groin extending on to the natal cleft. The borders were irregular with granulomatous base and serosanguineous discharge. Lesion was tender on palpation. Inguinal lymphadenopathy was absent. A solitary ulcer with well-circumscribed border was present over the glans of the penis [Figures 1 and 2]. The floor of the ulcer was covered with slough and pus discharge was noted. Provisional diagnosis of Donovanosis was made.

Complete hemogram was normal. Biochemical investigations such as random blood sugar liver function tests, and renal function tests were within normal limits. Chest X-ray, venereal disease research laboratory, HBsAg and HIV tests were normal. Dark ground microscopy, ZN stain, Wrights stain of the discharge, and tissue smear was negative. Swab for fungal and bacterial culture did not reveal any growth.
Stool for occult blood was negative. Antinuclear antibody, anti-ds DNA, cytoplasmic anti-neutrophil cytoplasmic antibodies, perinuclear anti-neutrophil cytoplasmic antibodies, rheumatoid arthritis factor were negative. Ultrasound abdomen and pelvis did not reveal any abnormalities. Histopathology from the ulcer margin showed irregularly acanthotic and spongiotic epidermis with marked neutrophil exocytosis. Papillary and reticular dermis showed mixed inflammatory infiltrate consisting neutrophils, lymphocytes, and plasma cells. Deeper parts of dermis showed liquefactive necrosis surrounded by neutrophils. Other parts showed dense lymphohistiocytic infiltrate, epithelioid cell granuloma with giant cells and fibrinous exudates. One vessel wall showed neutrophilic infiltrates without fibrinoid necrosis. These features were suggestive of PG. Colonoscopy was also normal.

Patient was empirically started on parenteral antibiotics, oral Fluconazole, and Dapsone. Once biopsy confirmed the diagnosis of PG, injection Dexamethasone 8 mg was given on a weekly tapering dose. Topically Metronidazole gel was given for local application. Healing at the margins and decrease in size of the lesion was noted after 1 month of follow-up. Later, he was lost to follow-up.

**DISCUSSION**

Pyoderma gangrenosum was first described by Brocq in 1916, the diagnosis of which is based on classical clinical presentation and exclusion of other cutaneous ulcerative diseases.[3]

Though the exact etiology is unknown, it is postulated that it is probably related to a hyperergic reaction causing lymphocytic antigenic stimulation that results in clonal proliferation in the regional lymph node with cytokine release and neutrophilic recruitment. Also suggested is a defect in neutrophilic chemotaxis and reactivity as the basic pathology in PG.[4,5]
Pyoderma gangrenosum is protean in its clinical manifestation.\[1\] The clinical presentations in PG can be differentiated into classic and atypical forms.\[6\] Classical type is the most common and best recognized variant of PG, presenting with small, tender, red–blue papules, plaques, or pustules that evolve into painful ulcers with characteristic violaceous undermined edges, the lesions of which heal with atrophic cibriiform scar. The atypical forms may be bullous, pustular, or superficial variant over the face and the arm.\[3\] Pathergy, the development of cutaneous lesions at sites of trauma is seen in 25% of patients with classical type of PG. More than 70% of PG have an underlying systemic disease, most commonly inflammatory bowel disease, myeloproliferative disorders, and internal malignancies.\[3\] Bullous PG is commonly associated with hematologic disease and most often appears on the upper limbs.\[7\] Pustular PG is almost always associated with inflammatory bowel disease, with rapid onset of pustules over the trunk face and limbs. Vegetative PG represents a milder and localized form of lesion which presents as a furuncle, abscess or ulcer typically over the trunk.\[1\] Site specific PG is also present over the lips, face, orbital adnexae, peristomal region, trunk, breast, or the genitals.\[3\]

Though penile pyoderma gangrenosum has been reported in normal and immunocompromised individuals, they mainly presented as isolated penile lesions, which differed from our case with involvement of the groin.\[8,9\] Donovanosis was the most probable differential diagnosis because it presents with ulceration over genitalia with rare features of granulation in groin and flexures of varying duration. But the absence of Donovan bodies and histopathological features of PG ruled out Donovanosis. Similar clinical features have been reported in the case of metastatic Crohn's disease, but since our patient did not have other features of Crohn's disease, it was ruled out.\[10\] The other differential diagnosis were Sweets syndrome, Wegeners granulomatosis, Behcet's disease, squamous cell carcinoma, and systemic lupus erythematosus. PG is often a diagnosis of exclusion as laboratory and histopathological findings are variable and nonspecific. A detailed history, physical examination, hematology, culture, and sensitivity and histopathology are important to exclude other possible causes from the site with skin.\[3\] The histopathological picture is nonspecific. Biopsies taken from the center of established PG lesions show marked neutrophilic infiltration with abscess formation, whereas those taken from edge show a mixed neutrophilic and lymphocytic inflammatory infiltrate and biopsies from the marginal erythematous zone show a predominantly lymphocytic infiltrate.\[11\] In our case, the marginal biopsy revealed both neutrophilic and lymphocytic infiltration.

Choice of treatment generally depends on disease severity as well as on the presence of associated disease. For early or mild lesions, topical therapy such as wet compresses, hydrophilic occlusive dressings, antimicrobial agents, and topical corticosteroids may be sufficient.\[1,2\] For more severe disease, or for PG resistant to topical therapy, oral corticosteroids have been the mainstay of therapy. The other systemically used drugs are Dapsone, Cyclophosphamide, Minocycline, Cyclosporine, Thalidomide, Methotrexate, or other immunomodulators.\[1,6\]

This case is reported in order to emphasize upon considering PG as one of the differential diagnosis in those genital ulcers which do not respond to the prescribed line of management.

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