Genital pyoderma gangrenosum

Sir,

Pyoderma gangrenosum (PG) is defined as an inflammatory, reactive, noninfective, nonneoplastic skin disease.[1] The four main clinical types of PG are ulcerative, pustular, bullous, and vegetative. A few atypical variants have also been described such as peristomal PG, genital PG, an oral variant that is pyostomatitis vegetans, and an extracutaneous neutrophilic disease.[1,2] The diagnosis of PG is one of the exclusion. Classic PG usually occurs on the lower extremities, but may occur anywhere. A very few cases of vulvar PG are reported in the literature. We describe a young girl who presented with an ulcer over the genitalia who was diagnosed as a case of PG and treated successfully with systemic steroids.

A 21-year-old unmarried girl presented with the history of genital ulcer of 2 weeks duration that had developed from a small pus filled lesion. There was history of intermittent fever and painful oral aphthae. The ulcer was asymptomatic initially, later associated with severe pain and foul smelling discharge. Patient denied history of any genital trauma and had never been sexually active. There was no history of vaginal discharge, bleeding on touch, joint pain, or eye complaints. Her menstrual cycles were regular. Review of systems was noncontributory.

On local genital examination, there was a solitary well-defined ulcer with foul smelling discharge and slough over the lower aspect of the left labia majora extending to labia minora and the adjacent groin [Figure 1]. It was tender and nonindurated. The ulcer did not bleed on touch. There was no regional lymphadenopathy. Pathergy test was negative. Evaluation for sexually transmitted infections was negative. Blood investigations revealed anemia and elevated erythrocyte sedimentation rate. Liver and renal function tests, urinalysis, and stool examinations were within the normal limits.

The skin biopsy specimen for histopathological examination revealed epidermis with a canthosis and granulation tissue along with diffuse infiltration of the dermis with neutrophils, occasionally lymphocytes and macrophages [Figure 2]. These findings were consistent with PG. Patient was then treated with

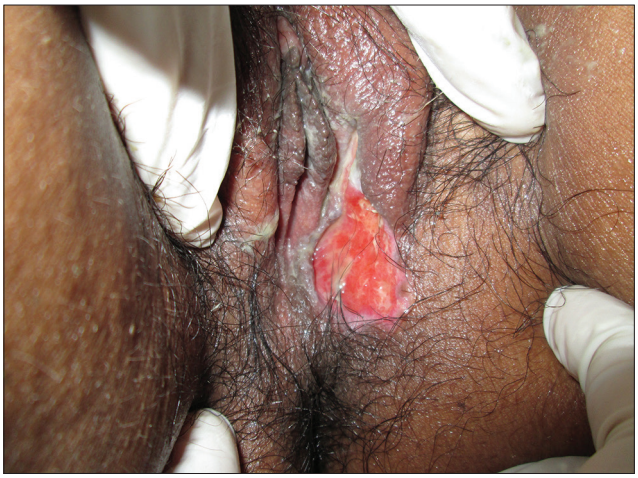

Figure 1: Well-defined ulcer with slough over the lower aspect of the left labia majora extending to labia minora

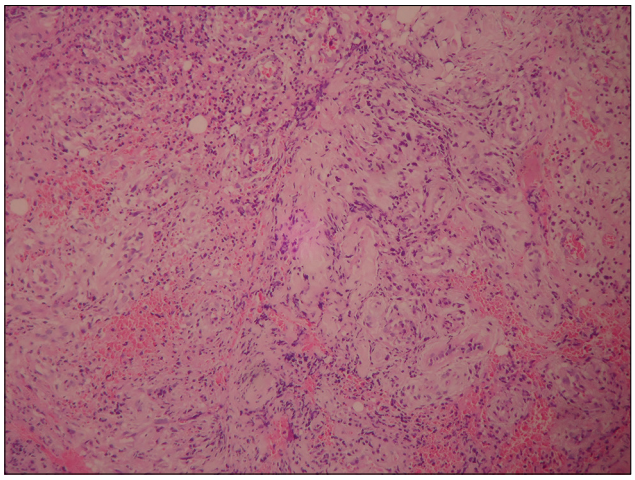

Figure 2: Massive neutrophilic infiltration forming a "sea of neutrophils" (H and E, magnification ×10)

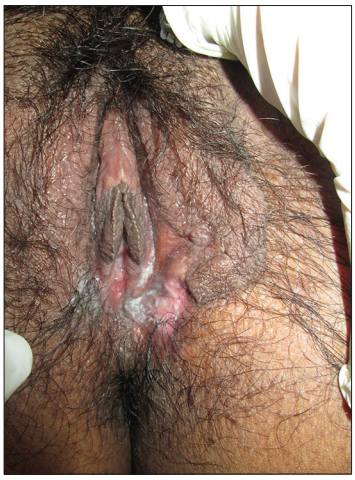

Figure 3: Ulcer healed with post-inflammatory hypopigmentation

injection betamethasone 4 mg 12th hourly, which was then gradually tapered. The lesion responded and healed well [Figure 3].

Approximately, 50% of patients with PG have an associated systemic disease. These diseases may precede, follow, or occur simultaneously.[2] Our patient did not have any evidence of systemic involvement. Involvement of the perineum is considered pathognomonic of patients with human immunodeficiency virus (HIV).[3] HIV was ruled out in our patient.

Even though biopsy can sometimes lead to extension of the ulceration (via pathergy) it should be done in most cases. The histopathological features of PG, though not specific, are useful in ruling out other causes of ulceration.[4] Massive neutrophilic infiltration("sea of neutrophils"), in the absence of vasculitis and granuloma formation, is seen in PG.[2] The histopathological examination in our patient confirmed the diagnosis of PG.

In our patient, we emphasized on local hygiene and sterile dressings. Systemic corticosteroids are the drug of choice for the treatment of PG and are particularly effective in treating the acute, rapidly progressive form of this disease.[5] Our patient was successfully treated with systemic corticosteroids.

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Quick Response Code:

Website: www.ijstd.org

DOI: 10.4103/0253-7184.142426