Recalcitrant vulvar pyoderma gangrenosum successfully treated with mycophenolate mofetil

Dear Editors,

Pyoderma gangrenosum (PG) is an inflammatory neutrophilic dermatosis characterized by recurrent, painful, ulcerations. Treatment typically involves anti-inflammatory agents, including corticosteroids, calcineurin inhibitors, antimetabolites, and TNF-alpha inhibitors. We present a case of vulvar PG, mimicking vulvar cellulitis and refractory to steroids, with successful response to mycophenolate mofetil (MM).

A 47-year-old female was referred to our clinic with a 2-month history of multiple, recurrent, and painful ulcerations on the vulva, perineum, and buttocks and a 1-year history of oral ulcerations, and daily joint pain. She had previously been treated for vulvar cellulitis without improvement and had subsequent development of ulcerations. She denied family history of inflammatory bowel disease, Behcet’s, or other connective tissue disorders.

Examination revealed diffuse painful ulcerations with erythematous rims on the mons pubis, labia, perineum, and gluteal cleft (Fig. 1). Labs demonstrated normal human leukocyte antigen B51, antinuclear antibody mildly positive at 1:320, mildly elevated erythrocyte sedimentation rate, and C-reactive protein. Serum and urine protein electrophoresis were within normal limits. Biopsy revealed epidermal necrosis and ulceration with neutrophilic inflammation (Fig. 2) and negative direct immunofluorescence. Initially, a working diagnosis of Behcet’s versus PG was made, and she was started on prednisone, colchicine, and high potency topical steroids. She acutely worsened on this regimen, prompting additional incisional biopsies, which revealed neutrophilic dermatosis with bacterial cocci, concerning for PG with bacterial superinfection. At this time, a diagnosis of PG was favored, given the absence of features concerning for vasculitis on histology, normal HLA B51 levels, lack of eye or other skin involvement, and negative pathergy test. Tissue cultures were positive for methicillin-susceptible Staphylococcus aureus and enterococcus, and she was started on antibiotics. Esophagogastroduodenoscopy and colonoscopy were unrevealing for features suggestive of inflammatory bowel disease.

Additional therapies included cyclosporine (efficacious; discontinued due to acute kidney injury) and dapsone (discontinued due to intolerance). Patient declined infliximab. She was seen by gynecology at an outside institution and received 2 trials of wound vacuum-assisted closure with mild improvement. Six months after initial presentation, she was started on MM 1,500 mg twice daily and reported complete resolution 8 months later. She has maintained clearance for over 1 year.

The classical course of PG involves progression of a papule/pustule into an ulcer with an erythematous, rolled border, and purulent base. Although most commonly found on the lower extremities and frequently associated with systemic inflammatory conditions, several reports have demonstrated isolated PG affecting mucocutaneous regions, including the vulva. PG is a diagnosis of exclusion, and active infection, malignancy, and underlying disease should be investigated. Our patient’s vulvar ulcerations were initially treated as cellulitis. This case highlights that the differential for vulvar swelling and erythema should also include early PG.

First line therapy includes corticosteroids followed by systemic immunosuppressants. Infliximab is efficacious in treating vulvar PG, but we were unable to initiate infliximab due to patient preference and insurance limitations. In PG, MM reduces ulcer size with a favorable side-effect profile. Thus, MM may be an appropriate monotherapy in cases where biologics are not feasible. Additional beneficial therapies in our case included wound vacuum-assisted closure, which removes pressure surrounding a wound to eliminate excess exudate and microorganisms and increase blood flow. Prior reports demonstrate efficacy of negative-pressure wound therapy in PG.

In summary, PG is an ulcerating condition that can affect the vulva and mimic vulvar cellulitis. In this case of severe vulvar PG, treatment with MM resulted in complete resolution of ulcers.

Conflicts of interest
None.

Patient consent
Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

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Fig. 1. Genital erythema and swelling upon presentation (A), prior to starting mycophenolate mofetil (B, C), and after 8 mo of mycophenolate mofetil (D, E).

Fig. 2. At low power in this punch biopsy specimen, there is loss of the epidermis with superficial ulceration (H&E, 40x, original magnification). Inset: on high power, there is a moderately dense infiltrate of neutrophils at the base of the ulcer. There is some associated necrosis and hemorrhage (H&E, 200x, original magnification).