

Painful Nonhealing Vulvar and Perianal Erosions

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A 38-year-old woman with a history of Crohn disease presented with painful nonhealing vulvar and perianal erosions of 6 months' duration. The erosions developed 4 months after discontinuing adalimumab for a planned surgery. During this time, the patient also had an exacerbation of Crohn colitis and developed an anal fistula. Prior to this break in adalimumab, the patient's Crohn disease was well controlled on adalimumab 40 mg every 2 weeks, azathioprine 100 mg daily, and mesalamine 4.8 g daily. Despite restarting adalimumab and therapy with multiple antibiotics (ie, metronidazole, ciprofloxacin), the erosions persisted. On physical examination erythematous plaques and nodules were present at the vulvar (top) and perianal (bottom) skin. In addition, well-demarcated erosions measuring 20 mm and 80 mm were present on the vulvar and perianal skin, respectively. Human immunodeficiency virus screening and rapid plasma reagin were negative.

WHAT'S THE DIAGNOSIS?

- Behçet disease
- cutaneous Crohn disease
- genital herpes infection
- Jacquet dermatitis
- sarcoidosis

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The authors report no conflict of interest.

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THE DIAGNOSIS: Cutaneous Crohn Disease

A punch biopsy of the vulvar skin revealed epidermal hyperplasia with moderate spongiosis and exocytosis of lymphocytes and neutrophils in the epidermis. A brisk mixed inflammatory infiltrate of epithelioid histiocytes, multinucleate foreign body–type giant cells, lymphocytes, plasma cells, neutrophils, and eosinophils in a granulomatous pattern also were present in the dermis (Figure). Periodic acid–Schiff and acid-fast bacillus stains were negative. Given the history of Crohn disease (CD) and the characteristic dermal noncaseating granulomas on histology, the patient was diagnosed with cutaneous CD.

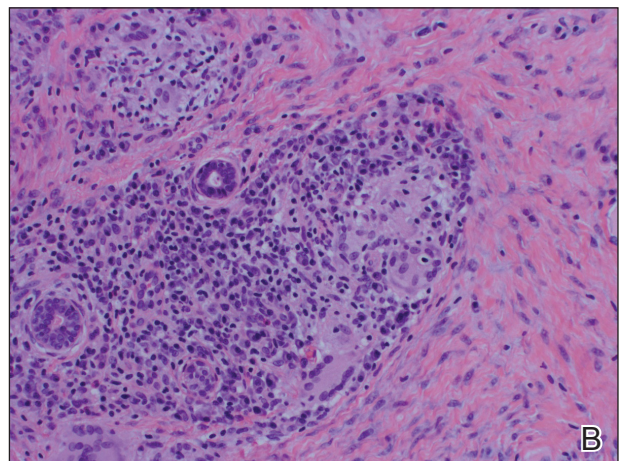
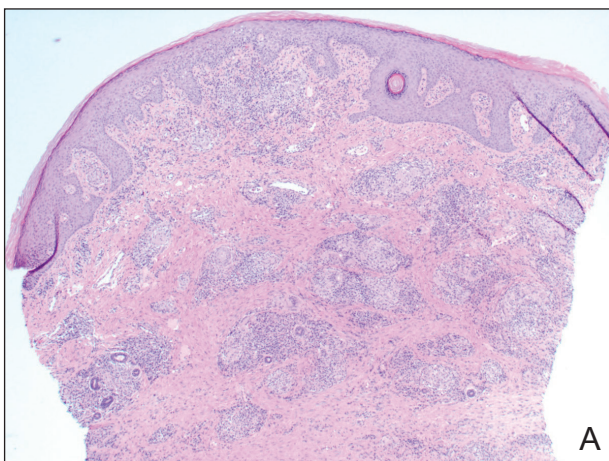
Although the patient was offered a topical corticosteroid, she deferred topical therapy. Given the lack of response to adalimumab, the gastroenterology department switched the patient to a treatment of infliximab 5 mg/kg every 8 weeks. Azathioprine was discontinued and the patient was switched to intramuscular methotrexate 25 mg/mL weekly. Slow reepithelialization of the vulvar and perianal erosions occurred on this regimen.

Although CD has numerous cutaneous features, cutaneous CD, also known as metastatic CD, is the rarest cutaneous manifestation of CD.¹ This disease process is characterized by noncaseating granulomatous cutaneous lesions that are not contiguous with the affected gastrointestinal tract.² The pathogenesis of cutaneous CD is unknown. Young adults tend to be more predisposed to developing cutaneous CD, likely due to the age distribution of CD.³

Cutaneous CD commonly presents in patients with a well-established history of gastrointestinal CD but occasionally can be the presenting sign of CD.¹ The most common sites of involvement are the legs, vulva, penis, trunk, face, and intertriginous areas. Cutaneous CD findings can be divided into 2 subgroups: genital and nongenital lesions. Genital findings involve ulceration, erythema, edema, and fissuring of the vulva, labia, clitoris, scrotum, penis, and perineum. Nongenital cutaneous manifestations include ulcers; erythematous papules, plaques, and nodules; abscesslike lesions; and lichenoid papules.^{4,5} The severity of cutaneous lesions does not correlate to the severity of gastrointestinal disease; however, colon involvement is more common in patients with cutaneous CD.⁶

Histologically, cutaneous CD presents as noncaseating granulomatous inflammation in the papillary and reticular dermis. These granulomas consist of epithelioid histiocytes and multinucleated giant cells with a lymphocytic infiltrate.⁵

Given the rarity of cutaneous CD, treatment approach is based on anecdotal evidence from case reports and case series. For a single lesion or localized disease, topical superpotent or intralesional steroids are recommended for initial therapy.³ Oral metronidazole also is an effective treatment and can be combined with topical or intralesional steroids.⁷ For disseminated disease, systemic corticosteroids have shown efficacy.³ Other reported treatment options include oral corticosteroids, sulfasalazine, azathioprine, 6-mercaptopurine, infliximab, and



Epidermal hyperplasia with exocytosis of lymphocytes and neutrophils (A)(H&E, original magnification $\times 4$) and mixed inflammatory granulomas (B)(H&E, original magnification $\times 40$).

adalimumab. If monotherapy fails, combination therapy may be needed. Surgical debridement may be attempted if medical therapy fails but is complicated by wound dehiscence and disease recurrence.³

Although genital ulcers can be a presentation of Behçet disease and genital herpes infection, genital nodules and plaques are not typical for these 2 diseases. Also, the patient did not have oral ulcers, which is a common feature of Behçet disease. Genital sarcoidosis is extremely rare, and cutaneous CD was more likely given the patient's medical history. Finally, Jacquet dermatitis is more common in children, and patients with this condition typically have history of fecal and urinary incontinence.

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