Exophytic Firm Papulonodule on the Labia in a Patient With Nonspecific Gastrointestinal Symptoms

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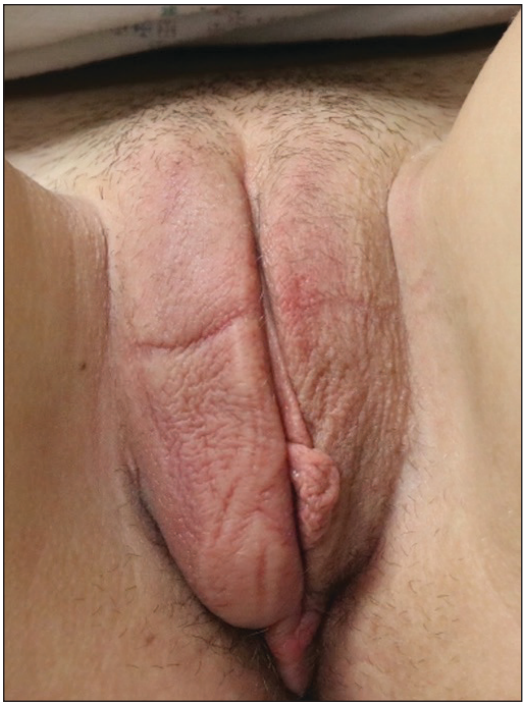

An 18-year-old woman with chronic constipation presented with an enlarging, painful, and edematous "lump" in the perineum of 1 year's duration. The lesion became firmer and more painful with bowel movements. Physical examination revealed an enlarged right labia majora, as well as a pink to flesh-colored, exophytic, firm papulonodule in the perineum posterior to the right labia. The patient concomitantly was following with gastroenterology due to abdominal pain that worsened with eating, as well as constipation, nausea, weight loss, and rectal bleeding of 5 years' duration. The patient denied rash, joint arthralgia, or oral ulcers. A biopsy from the labial lesion was performed.

WHAT'S YOUR **DIAGNOSIS?**

a. condyloma acuminatum
b. cutaneous Crohn disease
c. cutaneous sarcoidosis
d. irritable bowel syndrome with constipation
e. large sentinel skin tag

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THE **DIAGNOSIS:** Cutaneous Crohn Disease

Kinyoun and Grocott-Gomori methenamine-silver staining of the labial biopsy were negative for mycobacteria and fungi, respectively. A complete blood cell count, erythrocyte sedimentation rate, C-reactive protein, celiac disease serologies, stool occult blood, and stool calprotectin laboratory test results were within reference range. Magnetic resonance imaging of the pelvis demonstrated an anal fissure extending from the anal verge at the 6 o'clock position, abnormal T2 bright signal in the skin of the buttocks and perineum extending to the labia, and mild mucosal enhancement of the rectal and anal mucosa. Esophagogastroduodenoscopy and magnetic resonance elastography were unremarkable. Colonoscopy demonstrated scattered superficial erythematous patches and erosions in the rectum. Histologically, there was mild to moderately active colitis in the rectum with no evidence of chronicity. Given our patient's labial edema and exophytic papulonodule (Figure 1) in the setting of nonspecific gastrointestinal symptoms and granulomatous dermatitis seen on pathology (Figure 2), she was diagnosed with cutaneous Crohn disease (CD).

In our patient, labial biopsy was necessary to definitively diagnose CD. Prior to biopsy of the lesion, our patient was diagnosed with irritable bowel syndrome with constipation leading to an anal fissure and skin tag due to lack of laboratory, imaging, and colonoscopy findings commonly associated with CD. Her biopsy results and gastrointestinal symptoms made these diagnoses, as well as condyloma or a large sentinel skin tag, less likely.

Extraintestinal findings of CD, especially cutaneous manifestations, are relatively frequent and may be present in as many as 44% of patients.[1,2] Cutaneous CD often is characterized based on pathogenic mechanisms as either reactive, associated, or CD specific. Reactive cutaneous manifestations include erythema nodosum, pyoderma gangrenosum, and oral aphthae. Associated cutaneous manifestations include vitiligo, palmar erythema, and palmoplantar pustulosis.[2] Crohn disease–specific manifestations, including genital or extragenital metastatic CD (MCD), fistulas, and oral involvement, are granulomatous in nature, similar to intestinal CD. Genital manifestations of MCD include edema, erythema, fissures, and/or ulceration of the vulva, penis, or scrotum. Labial swelling is the most common presenting symptom of MCD in females in both pediatric and adult age groups.[2] Lymphedema, skin tags, and condylomalike growths also can be seen but are relatively less common.[2]

Given the labial edema, exophytic papulonodule, and granulomatous dermatitis seen on histopathology, our patient likely fit into the MCD category.[2] In adults, most

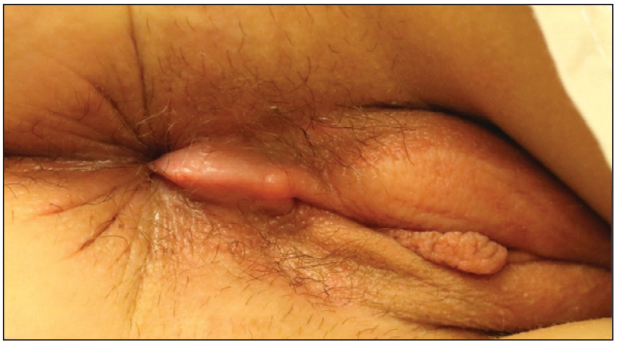

FIGURE 1. Enlarged view of the patient's right labia on initial presentation to dermatology, characterized by a flesh-colored, exophytic, firm papulonodule.

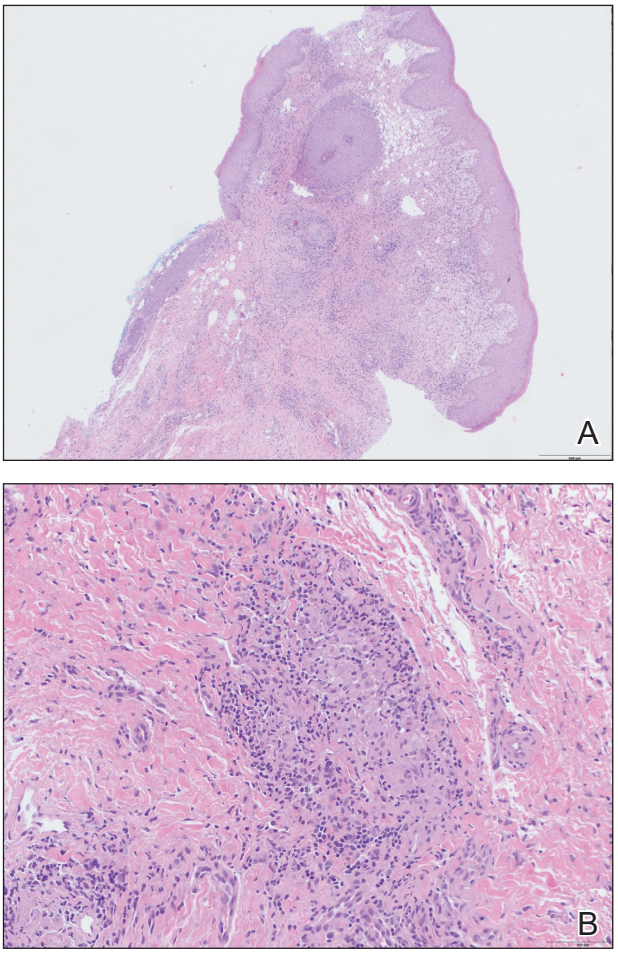

FIGURE 2. A and B, Histopathology of a biopsy from the right labia showed granulomatous dermatitis (H&E, original magnifications ×4 and ×20). Reference bars indicate 500 μm.

instances of MCD arise in the setting of well-established intestinal CD disease,[3] whereas in children 86% of cases occur in patients without concurrent intestinal CD.[2]

Given the nonspecific and variable presentation of MCD, the differential diagnosis is broad. The differential diagnosis could include infectious etiologies such as condyloma acuminatum (human papillomavirus); syphilitic chancre; or mycobacterial, bacterial, fungal, or parasitic vulvovaginitis. Sexual abuse, sarcoidosis, Behçet disease, or hidradenitis suppurativa, among other diagnoses, also should be considered. Diagnostic workup should include biopsy of the lesion with special stains, polarizing microscopy, and tissue cultures.[4] A thorough evaluation for gastrointestinal CD should be completed after diagnosis.[3]

The clinical course of vulvar CD can be unpredictable, with some cases healing spontaneously but most persisting despite treatment and sometimes prompting surgical removal.[2,4] Early recognition is crucial, as long-standing MCD lesions can be therapy resistant.[5] Due to the rarity of the condition and lack of data, there is a lack of treatment consensus for MCD. In 2014, the American Academy of Dermatology published treatment guidelines recommending superpotent topical steroids or topical tacrolimus as first-line therapy. Next-line therapy includes oral metronidazole, followed by prednisolone if still symptomatic.[3] Treatment-resistant disease can warrant treatment with immunomodulators or tumor necrosis factor α inhibitors. Our patient was started on adalimumab; after just 2 months of therapy, the labial swelling decreased and the exophytic nodule was less firm and smaller.

Metastatic CD is a rare manifestation of cutaneous CD and can be present in the absence of gastrointestinal disease.[3] This case demonstrates the importance of recognizing the cutaneous signs of CD and the necessity of lesional biopsy for the diagnosis of MCD, as our patient presented with nonspecific gastrointestinal symptoms and a diagnostic workup, including endoscopies, that proved inconclusive for the diagnosis of CD.

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