Crohn Disease Presenting as Vaginal Discharge in a Patient With Systemic Lupus Erythematosus

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Crohn disease (CD), a subtype of inflammatory bowel disease, may be complicated by extraintestinal manifestations. Although cutaneous findings are among the most frequent, metastatic Crohn disease (MCD), defined as mucocutaneous CD at sites noncontiguous with the gastrointestinal tract, is rare. Children with MCD are more likely than adults to develop genital inflammation. Case reports describe the vulva, scrotum, and penis as common locations of involvement in children with genital MCD. We report a case of a pediatric patient with systemic lupus erythematosus who developed vaginal discharge as a presenting symptom of MCD in the absence of an anorectal-vaginal fistula. We describe an unusual combination of clinical features and intend to improve awareness of uncommon extraintestinal manifestations of CD.

INTRODUCTION

Crohn disease (CD) is a chronic inflammatory bowel disease (IBD) associated with extraintestinal manifestations in up to 35% of cases, with mucocutaneous symptoms occurring most frequently.[1,2] Metastatic CD (MCD), a form of mucocutaneous CD at sites noncontiguous with the gastrointestinal tract, is rare, and involvement of genital sites is very rare.[3] With increasing incidence and prevalence of pediatric-onset IBD, it is especially important for pediatricians to recognize the potential presenting symptoms of CD.[4] Furthermore, the coexistence of systemic lupus erythematosus (SLE) and CD is limited to case reports and case series.[5,6] We report a case of a pediatric patient with SLE and with vaginal discharge as a presenting symptom of vaginal MCD.

CASE REPORT

A 15-year-old female patient with well-controlled SLE presented with rash and vaginal discharge. She was diagnosed with SLE at 13 years old after developing fever, vomiting, diarrhea, hypotension, transient arthritis of the small joints of the hands and 1 knee, and pleural effusions. Laboratory testing revealed elevated serum antinuclear antibodies, anti–double-stranded DNA (dsDNA) antibodies, anti-ribonucleoprotein antibodies, positive direct Coombs test, thrombocytopenia, and hypocomplementemia. She was treated with hydroxychloroquine and subsequently experienced resolution of systemic symptoms, pleural effusions, arthritis, hypocomplementemia, and anti-dsDNA antibodies. Approximately 1 year after the diagnosis of SLE was made, she developed a pruritic, painful vulvar rash and profuse vaginal discharge. She was postmenarchal and not sexually active. The copious vaginal discharge was malodorous, thin, clear to white, and soaked through multiple menstrual pads daily. Physical examination revealed Tanner stage 5

abstract

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Ms Ries and Dr Chiu conceptualized and designed the study, collected data, drafted the initial manuscript, and critically reviewed and revised the manuscript. Drs Samalik, Nocton, Francis, Arkin, Seroogy, and Young collected data and critically reviewed and revised the manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

CONFLICT OF INTEREST DISCLOSURES: The authors have no conflicts of interest relevant to this article to disclose.

FUNDING: The authors have no funding relevant to this study to disclose.

Accepted for Publication Date: February 17, 2026

https://doi.org/10.1542/peds.2025-073979

To cite: Ries AG, Samalik J, Nocton JJ, et al. Crohn Disease Presenting as Vaginal Discharge in a Patient With Systemic Lupus Erythematosus. *Pediatrics*. 2026;158(1):e2025073979

genitalia, with a pink, scaly, eroded and fissured plaque over the mons pubis, vulva, and perianal region. A vaginal wet mount revealed yeast; however, fluconazole 150 mg for 3 days and then weekly for 6 months did not improve the vaginal discharge. Cultures of the skin and vaginal discharge revealed methicillin-sensitive *Staphylococcus aureus* (MSSA), and the vaginal discharge repeatedly grew Group B *Streptococcus* (GBS) and multidrug-resistant *Escherichia coli*. Initially, the vaginal discharge and vulvar rash would nearly resolve after treatment with oral cephalexin and reappear shortly after discontinuation. To reduce vaginal discharge through suppression of the hypothalamic-pituitary-ovarian axis, intramuscular medroxyprogesterone was administered; however, there was no improvement.

Two months after presenting with genital symptoms, the patient developed fever, abdominal pain, and a pilonidal abscess. She had continued vaginal discharge and vulvar rash, with new large perianal skin tags. The abscess was incised and drained, growing multidrug-resistant *E coli*, MSSA, and GBS, and resolved with ertapenem. Magnetic resonance imaging (MRI) of the pelvis to evaluate for fistulizing disease showed mild rectal wall thickening and enhancement with surrounding inflammation/edema. Fecal calprotectin (>3000 μg/g, normal ≤49 μg/g), C-reactive protein (CRP) (8.9 mg/dL, normal 0–1.0 mg/dL), and erythrocyte sedimentation rate (>100 mm, normal 0–10 mm) were elevated. An esophagogastroduodenoscopy was grossly normal, while colonoscopy showed a perianal fistula, perianal skin tags, and diffuse areas of mildly congested, friable and inflamed mucosa in the rectum, recto-sigmoid colon, sigmoid colon and descending colon. The pathology showed active esophagitis, gastritis, ileitis, and colitis with a mixed inflammatory infiltrate, largely intact architecture, and no granulomas. Due to the clinical presentation and perianal disease, CD was favored over lupus enteritis, but a CD diagnosis could not be confirmed. To evaluate the ongoing vaginal discharge, a sedated vaginal speculum exam was completed and unremarkable, and pelvic ultrasonography and magnetic resonance enterography with intravenous contrast did not reveal an anorectal vaginal fistula.

After a negative QuantiFERON tuberculosis test, the patient was treated with an infliximab biosimilar intravenously for suspected CD, with rapid resolution of fever and abdominal pain, and radiographic improvement of the perianal inflammation on repeat MRI. However, the vaginal discharge and vulvar dermatosis continued unabated. The vulvar rash developed into red plaques with overlying knife-like erosions and extensive labial and gluteal edema. Biopsy of the mons pubis revealed nonspecific spongiotic dermatitis, with an additional biopsy of the left labia majora 3 months later showing similar findings. There was minimal improvement with empirical treatment for desquamative inflammatory vaginitis using 25 mg hydrocortisone intravaginal suppository and clindamycin 2% vaginal cream for 3 months. The patient developed a generalized psoriasiform dermatitis that improved with 100 mg cyclosporine twice daily without associated improvement in the vaginal discharge. Eight months after initial presentation, a vaginal biopsy revealed a mixed infiltrate involving the mucosa and subepithelial stroma, with notable eosinophils and a solitary epithelioid granuloma (Figure 1), confirming the diagnosis of CD with vaginal involvement. At this time, the fecal calprotectin was still elevated at 1403 μg/mg (normal <80 μg/mg), with normal CRP. Given the psoriasis attributed to the infliximab biosimilar and the incomplete efficacy for the vaginal, vulvar, and luminal CD, the biosimilar was discontinued, and upadacitinib was initiated to treat both CD and psoriasis.[7] The psoriasis, vaginal discharge, and vulvar inflammation gradually resolved over several weeks, and the cyclosporine was discontinued. Further evaluation for an underlying inborn error of immunity (IEI) was prompted by the patient's comorbidity of both SLE and CD. Extensive immune testing was notable for elevated IgG (1710 mg/dL; normal: 508–1080 mg/dL) and IgA

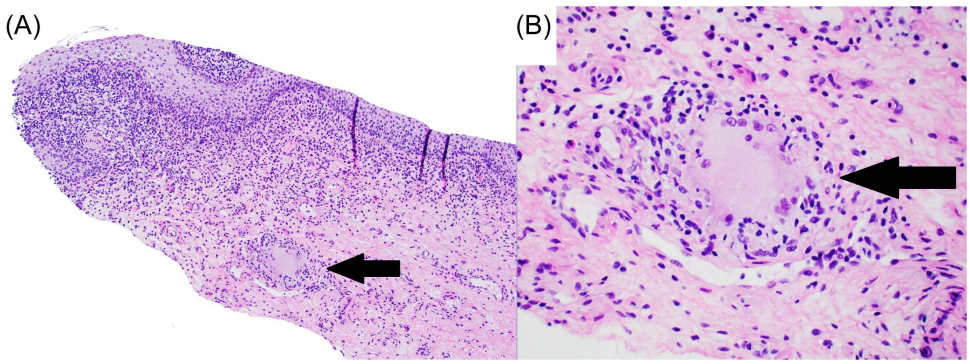

FIGURE 1.
Right vaginal biopsy consistent with MCD. (A) H&E staining on 20 × power with acanthosis of the epithelium with rare neutrophils on the surface, moderate spongiosis, exocytosis of lymphocytes, and superficial subepithelial infiltrate composed of lymphocytes, plasma cells, histiocytes and eosinophils with a focal noncaseating granuloma (arrow). (B) H&E staining on 100 × with a focal noncaseating granuloma (arrow).
Abbreviation: H&E, hematoxylin & eosin.

(429 mg/dL; normal: 52–232 mg/dL) and increased activated T cells. The following tests were notably normal: neutrophil oxidative burst, plasma C-X-C motif chemokine ligand 9 level, and frequency of double-negative alpha-beta T cells. A clinically available gene panel for IEI (474 genes, Invitae), as well as whole-exome sequencing (Prevention Genetics) did not reveal pathogenic variants.

DISCUSSION

Extraintestinal manifestations are present in 15% to 28% of children with CD, and cutaneous manifestations, specifically, are present in 7.5% of patients.[8–10] Mucocutaneous manifestations of CD include reactive cutaneous manifestations such as erythema nodosum, pyoderma gangrenosum, or Sweet syndrome; associated conditions such as aphthous stomatitis; or treatment-related conditions such as psoriasiform eruptions.[11,12] Mucocutaneous CD describes an inflammatory process similar to the luminal disease with shared histopathologic findings. It is classified as contiguous extension into adjacent sites, including perioral or perianal, or noncontiguous involvement, termed MCD. MCD in general is scarcely reported.[13,14] In a case series and review by McKay et al, 89 pediatric patients of MCD were reported, with genital involvement observed in 75% (67/89) of patients.[13] Notably, mucocutaneous CD has been reported to precede the clinical manifestations of intestinal CD in 15% of pediatric patients.[12,15,16]

Genital MCD is difficult to diagnose, particularly in children, as it can be mistaken for sexual abuse or posttraumatic lymphedema.[13] The most common vulvar presentation is labial swelling and chronic ulcerations.[1] Complications of CD involving the genitals and reproductive tract by penetrating disease can cause significant morbidity. Vaginal involvement of CD typically arises from direct extension of anorectal fistulas, and imaging is critical in identifying this complication, if not detected on physical exam.[17] Case reports have described vaginal discharge and desquamative inflammatory vaginitis in patients with CD,[18–21] but none had granulomas on biopsy without anorectal-vaginal fistula.

Diagnostic criteria have recently been proposed but not yet validated for MCD.[12,14] Genital and cutaneous MCD require the presence of 1 major and 1 minor, 2 major, or 3 minor criteria. Major criteria include genital swelling; knifelike ulcers of the genital, inguinal, or intertriginous skin; or lymphedematous changes of the genital skin. Minor criteria include perianal skin tags, genital fissuring, nonhealing genital ulcerations, a cutaneous biopsy suggestive of MCD, or a diagnosis of CD. Our patient met proposed criteria with 2 major criteria (genital swelling and knife-cut erosions) and 2 minor criteria (perianal skin tags and CD diagnosis); her diagnosis of CD was not confirmed until the granuloma was seen on vaginal biopsy, as her gastrointestinal findings were atypical.[14]

This case highlights the difficulty when diagnosing MCD due to the variable clinical presentation and presenting symptoms. Granulomas are not universally present in CD biopsy samples, with a literature review reporting prevalence rates ranging from 12.76% to 66.67%.[22] While the proposed diagnostic criteria do not require histopathologic diagnosis of MCD due to the potential for false-negative testing, the histopathologic findings from the vaginal biopsy of noncaseating granulomas were ultimately integral to providing a diagnosis and treatment direction for the vaginal discharge. Two biopsies of the vulva were unrevealing, and it is unclear if the vulvar dermatosis was MCD as well or a reactive cutaneous manifestation, but would meet MCD diagnostic criteria with the newly proposed criteria.[12] Although desquamative inflammatory vaginitis has been reported in women with CD and a therapeutic trial was attempted in our patient, it was ultimately felt not to be the cause of her vaginal discharge.[23] The case presented here of vaginal and vulvar MCD preceding the diagnosis of CD and in the absence of an anorectal-vaginal fistula raises awareness of this atypical presentation of CD.[3] MCD should be considered in the differential diagnosis in patients with a refractory, persistent vulvar rash and vaginal discharge, even if histopathologic evidence of granulomas are not found.

The coexistence of SLE and CD is uncommon, and a 2020 literature review found only 9 reported cases of SLE preceding CD.[5,6] In patients with SLE, CD can be difficult to differentiate from lupus enteritis. Pathologic examination is key to distinguishing the 2 diseases, with vasculitis, thrombosis, and a transmural inflammatory infiltrate with lymphocytes, plasma cells, and neutrophils seen in lupus enteritis and colitis with transmural lymphoid aggregates, fissuring ulcers, and sometimes granulomas seen in CD; however, histopathologic features can be nonspecific or overlap between the 2 entities.[5] Extensive testing in our patient did not identify a unifying immune disorder to explain the unusual co-occurrence of SLE and CD.

This complex case underscores the importance for pediatricians and other subspecialists to recognize the rare manifestations of the common autoimmune disorders of SLE and CD. Multidisciplinary collaboration and repeat investigative studies were necessary to ultimately make the correct diagnosis and direct treatment.

CONSENT

Written permission was obtained from the patient's guardian(s) to publish this case and the corresponding photos or imaging studies.

ACKNOWLEDGMENTS

We acknowledge the contribution of Drs Rebecca Czaja and Allison Miller, whose clinical expertise and compassionate care were integral to the treatment of this patient. While they could not be included as authors, their involvement

was essential to the multidisciplinary care and overall outcome for this patient.

ABBREVIATIONS

CD: Crohn disease
CRP: C-reactive protein
dsDNA: double-stranded DNA
GBS: Group B *Streptococcus*
IBD: inflammatory bowel disease
IEI: inborn error of immunity
MCD: metastatic Crohn disease
MRI: magnetic resonance imaging
MSSA: methicillin-sensitive *Staphylococcus aureus*
SLE: systemic lupus erythematosus

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