Cutaneous Crohn’s disease of the vulva after a total colectomy and without gastrointestinal manifestations: A case report

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Abstract
Crohn’s disease has many cutaneous manifestations. We report a case of a 54-year-old woman, who had non-contiguous vulvar Crohn’s disease despite a total colectomy and the absence of gastrointestinal Crohn’s disease on ileoscopy.

Keywords
Cutaneous Crohn’s disease, vulvar disease

Introduction
The cutaneous manifestations of Crohn’s disease (CD) are varied and occur in up to 53% of patients.¹ Cutaneous lesions due to non-caseating granulomas are quite rare, especially when they are non-contiguous with the bowel. We report a patient, who was thought to have quiescent CD after a total colectomy. However, she developed vulvar erythema and swelling, which was consistent with cutaneous CD on histology.

Case report
A 54-year-old, post-menopausal woman presented with a 6-month history of vulvar swelling and burning. She had a 36-year history of CD that was surgically treated with a total colectomy and permanent ileostomy. Her intestinal CD was quiescent on her last ileoscopy. She had a history of uveitis that was thought to be associated with CD. The uveitis lasted for 3 years and resolved 2 years before presentation.

The patient was not receiving any treatment besides hormone replacement therapy and antidepressants. Review of systems was negative, specifically she had no oral or joint symptoms.

On examination, there was erythema of the vulva and suprapubic area as well as vulvar edema (Figure 1). Clitoral hood architecture was lost, but there was no purulent material, bleeding, vesicles, fistulas, or blisters. The rest of the perineum was unremarkable. Pelvic ultrasound revealed bilateral avascular thickening of the superficial labial tissues without collections or fistulas. A punch biopsy of the skin was performed from the mons pubis and it showed superficial and deep lymphohistiocytic perivascular and interstitial inflammation with numerous plasma cells as well as scattered interstitial giant cells on a background dermal fibrosis (Figure 2). Based on the non-caseating aggregations of lymphocytes and histiocytes, a diagnosis of metastatic or non-contiguous cutaneous CD was favored.

Discussion
CD is a relapsing-remitting inflammatory disease that can affect any part of the gastrointestinal tract. It can also affect other organs as well, such as the eyes, joints, and skin. The cutaneous manifestations of CD vary in pathogenesis and can be crudely classified using the SMART memory aid: specific (e.g. contiguous cutaneous CD), malabsorption-induced (e.g. acrodermatitis enteropathica), associated inflammatory (e.g. psoriasis), reactive (e.g. erythema nodosum), and treatment-induced (e.g. steroid-induced acne).¹ The specific lesions of cutaneous CD are characterized by non-caseating granulomas found within the skin that mimic

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the histopathological features of CD in the gastrointestinal tract. These granulomatous lesions are usually divided into two categories: contiguous and metastatic. Contiguous cutaneous CD connects to the intestinal tract through direct spread, fissures or fistulas, while metastatic cutaneous CD is separated from the intestine by healthy tissue. The word “metastatic” is misleading because CD is not a neoplasm and patients may have cutaneous lesions before they develop gastrointestinal disease and, in fact, some patients may never develop gastrointestinal disease. Therefore, the term non-contiguous cutaneous CD is more accurate.

Although rarely reported, the vulva is increasingly recognized as a site of both contiguous and non-contiguous CD. The most common presentation of vulvar non-contiguous CD (VNCD) is symmetric or asymmetric vulvar swelling with or without associated tenderness, as in our patient.\textsuperscript{2} This swelling could represent lymphedema from the granulomas impinging on lymphatic vessels. Other potential findings are ulceration and knife-like fissuring. Less commonly, VNCD presents with hypertrophic lesions or with chronic suppuration. Around 25% of patients with VNCD have no gastrointestinal symptoms at the time of diagnosis.\textsuperscript{2} Our patient had a history of CD that was in remission. Although she had ocular symptoms ascribed to CD that had resolved 2 years before presentation, her most recent ileoscopy did not reveal any CD activity. In fact, her disease had been well-controlled without any systemic medications since her total colectomy.

The differential diagnosis for vulvar swelling includes granuloma inguinale, lymphogranuloma venereum, syphilis, vulvar sarcoid, contact dermatitis, traumatic vulvitis, and metastatic carcinoma. A skin biopsy is necessary to make the diagnosis of VNCD, especially in patients who lack intestinal manifestations. On histologic examination, VNCD most commonly demonstrates non-caseating granulomas and a chronic, mixed inflammatory infiltrate. Blood work is less helpful, but it can help rule out infectious disorders. Radiographic imaging aids in excluding any fistulous connections to the intestines.

Consensus guidelines do not yet exist for treating VNCD. However, treatment algorithms have been proposed by multiple authors.\textsuperscript{2-5} Behavioral interventions involve compression underwear or shorts to aid with lymphatic drainage. First-line medical therapies include potent topical corticosteroids or topical calcineurin inhibitors. For resistant localized areas, intralesional triamcinolone acetonide can be tried. Patients with more extensive disease require systemic agents such as metronidazole, tumor necrosis factor (TNF)-alpha inhibitors, 5-aminosalicylates, or azathioprine. In refractory cases, surgical management is an option of last resort because it can be mutilating and necessitate subsequent reconstructive surgeries.

More work needs to be done to understand the pathogenesis and best treatment strategies for this rare condition. The reason why CD targets specific areas is unknown. Although Prezyna and Kalyanaraman\textsuperscript{6} reported a patient who developed vulvar CD after a subtotal colectomy, we could not find any other reports of a patient developing VNCD after a total colectomy. This case emphasizes that active intestinal CD is not necessary for vulvar CD.

Author contributions
C.F.R. contributed to patient recruitment and consent. M.B., C.F.R., and D.G. contributed to manuscript preparation. C.F.R. and D.G. contributed to figure preparation.

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