A Rare Clinical Presentation of Metastatic Crohn’s Disease

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Abstract

A 31-year-old female with a history of systemic lupus erythematosus, IgA nephropathy, and psoriasis presented with a one-month history of a painful, pruritic rash under the bilateral breasts and in the genital region. Cutaneous examination revealed a large, tender ulcer under the left breast with a shiny erythematous base and peripheral hypertrophic changes. Small ulcers were present on the bilateral inguinal folds, and the labia majora were edematous with multiple erythematous papules. Histological examination of the left breast revealed ulceration with granulomatous dermatitis, consistent with a diagnosis of metastatic Crohn’s disease. Metastatic Crohn’s disease is a rare cutaneous manifestation of Crohn’s disease characterized by non-caseating granulomas in regions non-contiguous with the gastrointestinal tract. At the time of diagnosis, our patient reported no gastrointestinal symptoms aside from occasional blood-streaked stools from hemorrhoids. This case demonstrates the importance of considering the disease when a patient presents with intertriginous or genital lesions, even in the absence of systemic manifestations.

Categories: Dermatology

Keywords: metastatic crohn’s disease, cutaneous crohn’s disease, cutaneous manifestations of systemic disease, crohn’s disease

Introduction

Crohn’s disease is a subtype of inflammatory bowel disease characterized by segmental, granulomatous lesions of the intestinal tract [1]. Cutaneous manifestations are common and typically occur in regions contiguous with the gastrointestinal tract, such as the perianal and oral region. Metastatic Crohn’s disease (MCD), in contrast, is a rare cutaneous manifestation of Crohn’s disease characterized histologically by non-caseating granulomas in regions non-contiguous with the gastrointestinal tract [1-3]. While most patients with MCD carry a previous diagnosis of Crohn’s disease, some patients present without classic gastrointestinal manifestations [4]. The clinical characteristics of MCD also vary, suggesting that the disease may be underrecognized due to misdiagnosis [5]. Herein, we report a rare presentation of MCD with involvement of the breast and genital regions in the absence of active intestinal manifestations.

Case Presentation

A 31-year-old female with a history of systemic lupus erythematosus, IgA nephropathy, and psoriasis presented with a painful, pruritic rash involving the inframammary and genital skin. The rash appeared approximately one month prior and progressively worsened since its onset. The patient stated that she applied topical hydrocortisone under her breasts without relief. She denied any diarrhea, hematochezia, or abdominal pain but admitted to occasional blood-streaked stools from hemorrhoids. Colonoscopy revealed rectal ulceration and exam-limiting stricture.

Cutaneous examination revealed a large, tender ulcer under the left breast with a shiny erythematous base and peripheral hypertrophic changes (Figure 1).
An erythematous patch without ulceration was present under the right breast. There were small ulcers on the bilateral inguinal folds and multiple verrucous, erythematous, and skin-colored papules on the labia majora with edema (Figure 2).

Multiple papules with interspersed fissures were found between the intergluteal folds (Figure 3). Tangential biopsy of the left breast showed ulceration with granulomatous dermatitis, consistent with a diagnosis of MCD.

Discussion
Crohn’s disease is an inflammatory disease characterized by segmental, granulomatous lesions of the intestinal tract. Cutaneous manifestations occur in approximately 44% of patients and are confirmed by the
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Human subjects:

Disclosures

Additional Information

Histologically, MCD presents similarly to the gastrointestinal lesions of Crohn’s disease, with non-caseating granulomas in the superficial papillary and deep reticular dermis and occasional extension into the subcutaneous fat [3,4]. These granulomas consist of epithelioid and multinucleated histiocytes with a lymphocytic infiltrate, occasionally surrounding blood vessels in a phenomenon called granulomatous perivasculitis. While plasma cells and eosinophils may be present, neutrophils and focal necrobiosis are generally absent [5].

The differential diagnosis of MCD is broad due to the variable clinical presentation. When involving intertriginous regions, as in our patient, MCD can resemble hidradenitis suppurativa, seborrheic dermatitis, or intertrigo. On the limbs, other diagnoses to consider include cellulitis, allergic contact dermatitis, pyoderma gangrenosum, and frequently misdiagnosed [5].

Due to the rarity of MCD, there is currently no established treatment. Although spontaneous resolution has been reported, most cases persist without treatment [7]. Previously employed therapies with mixed results include topical steroids, topical calcineurin inhibitors, oral antibiotics (metronidazole) for mild and limited disease, oral steroids, azathioprine, sulfasalazine, 6-mercaptopurine, and infliximab for severe cases. Surgical excision with debridement has also shown success in cases refractory to other therapies [4]. With any treatment modality, recurrence may occur [7].

Conclusions

Although rare, this case demonstrates the importance of considering MCD when a patient presents with intertriginous or genital lesions, even in the absence of active intestinal disease.

Additional Information

Disclosures

Human subjects: Consent was obtained by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References


