Vegetating Plaques in the Groin: A Manifestation of Metastatic Crohn's Disease

Chetana Jagatgere Math and Anju George

Abstract

Crohn's disease (CD) is a type of inflammatory bowel disease that can affect any part of the gut from mouth to anus. It also may occur at contiguous sites, like, lip, perineal or peristomal regions or may occur at sites separated from the bowel by normal tissue referred to as metastatic CD. The condition is relatively rare and may mimic or coexist with other dermatoses. The presence of noncaseating granuloma on histopathological examination confirms the diagnosis. Here, we report a case of metastatic CD with cutaneous symptoms of long duration and presenting with vegetating plaques in the lower abdomen, groins, and vulva. The gastrointestinal disease was under remission. The diagnosis was confirmed by histopathological examination and patient responded well to topical as well as systemic steroids.

Key Words: *Crohn's disease*, *metastatic Crohn's disease*, *noncaseating granulomas*, *vegetating plaques*

What was known?

- Metastatic Crohn's disease is characterized by specific non-caseating granulomatous cutaneous lesions with the same histopathology as intestinal lesions.
- Genital lesions usually manifest as erythema, edema, fissures or ulcers of the labiae, scrotum, or the penis.
- Atypical genital variants described include lymphedema, knife cut ulcers, and hypertrophic exophytic lesions.

Introduction

Crohn's disease (CD) is a type of inflammatory bowel disease that can affect any part of the gut from mouth to anus.[1,2] The cutaneous lesions in CD may occur at sites separated from the bowel by normal tissues and is referred to as “metastatic CD.” It is characterized by noncaseating granulomas in the superficial and deep dermis.[3] A few cases have been reported in the literature until date and most of them highlight the varied expositions of the condition.[1,4]

Here, we report a case of metastatic CD with the gastrointestinal counterpart under remission, presenting with vegetating plaques in the lower abdomen, groins, and vulva.

Case Report

An elderly lady presented with pruritus and burning sensation over lower abdomen, mons pubis, and both thighs of 1-year duration. Patient had been diagnosed with CD 15 years ago and was subsequently treated with azathioprine, salazopyrin, and other supportives. A sigmoid colostomy was performed 5 years ago for rectovaginal fistula, which was followed by an end colostomy after 6 months. She did not have additional comorbidities. At the time of presentation, the patient was off immunosuppressants and did not have active gastrointestinal involvement. The cutaneous symptoms gradually progressed over 3 weeks, and she developed raised erythematous lesions on the above-mentioned sites accompanied by oozing.

Dermatological examination revealed edema of both labia majora with a mousy odor. Multiple confluent erythematous and hyperpigmented papules and vegetating fissured plaques with oozing, erosions, and adherent crusting were present on the lower abdomen, mons pubis, labia majora, and both groins and thighs [Figure 1]. The perineal and submammary regions were spared. Differential diagnoses of metastatic CD, hidradenitis suppurativa, and pemphigus vegetans were considered. A skin biopsy was performed, and she was initiated on topical steroid-antibiotic combination.

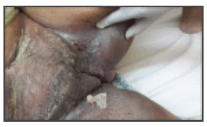

Figure 1
Vegetating plaques involving entire vulva extending to the groins

Histopathological examination revealed mild-to-moderate epidermal hyperplasia [Figure 2]. The dermis showed predominant chronic inflammation and foci of nonsuppurative granulomas with occasional multinucleated giant cells. Deeper dermis showed moderate-to-dense mixed inflammation with ill-defined granulomas [Figures 3 and 4]. Ziehl–Neelsen and PAS staining were negative for AFB and fungus, respectively. The results were consistent with metastatic CD. She was started on oral prednisolone 40 mg, tapered, and stopped over 2 weeks. On follow-up after 2 weeks, she had a significant improvement and the affected areas had almost completely healed [Figures 5 and 6].

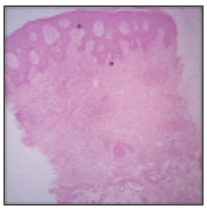

Figure 2
Epidermal hyperplasia with epithelioid cell granulomas in the dermis (H and E ×40)

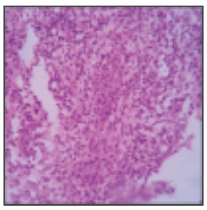

Figure 3
Granuloma composed of epithelioid cells, multinucleated giant cells, and lymphocytes (H and E, ×100)

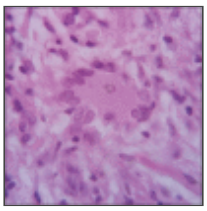

Figure 4
Langhans giant cells, lymphocytes in the granuloma (H and E, ×400)

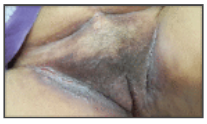

Figure 5
Lesions improving with topical steroid-antibiotic combination

Figure 6
Significant resolution with oral steroids

Discussion

CD is a type of inflammatory bowel disease that can affect any part of the gut from mouth to anus as noncontinuous or skip lesions and involve the entire thickness of the bowel. Extraintestinal manifestations are more common in CD when compared to ulcerative colitis, with an incidence of 22%–44%.[1,2] Skin is one of the most common organs affected. Broadly, the cutaneous involvement may be classified as contiguous, metastatic, and nonspecific lesions. Either the lesions may occur at contiguous sites such as the lip, peristomal and perineal regions or may occur at sites separated from the bowel by normal tissues referred to as “metastatic Crohn's disease.” The nonspecific lesions include erythema nodosum, pyoderma gangrenosum, neutrophilic dermatosis, Stevens-Johnson syndrome, erythema multiforme, acrodermatitis enteropathica, and epidermolysis bullosa acquisita.[3,4,5]

The term “metastatic Crohn's disease” was coined in 1970 by Mountain[6] and relatively few cases had been reported in literature. Metastatic CD is defined as the occurrence of specific granulomatous cutaneous lesions with the same histopathology (noncaseating granulomas with multinucleated giant cells in the dermis surrounded by lymphocytes, plasma cells, and eosinophils) as the intestinal lesions. There is no significant correlation between the gastrointestinal activity and onset of skin lesions or treatment response. It can be divided into two main clinical forms, the genital (56%) and the nongenital form (44%).[1] Genital lesions manifest usually as erythema, edema, fissures or ulcers of the labiae, scrotum, or the penis. The nongenital variant affects lower extremities and soles (38%), trunk and abdomen (24%), upper extremities and palms (15%), face and lips (11%), and intertriginous areas (8%). Here, the morphology varies consisting of papules, nodules, plaques with or without ulceration, abscesses, draining sinuses, hidradenitis suppurativa, lobular panniculitis, and scars.[1,7] Atypical genital lesions may mimic a variety of dermatoses. These include labial edema mimicking lymphedema,[8] knife cut ulcers, and hypertrophic exophytic variants.[9]

A slight female preponderance had been observed in a few studies. In the vast majority of cases, skin lesions developed in an established intestinal disease. However, in 20% cases, the cutaneous involvement might precede gastrointestinal involvement. The age of onset ranged from 29 to 39 years in adults, whereas children between 10 and 14 years were usually affected.[2,8]

Pathogenesis It is a well-known fact that Th1 pathway mediates CD with elevated levels of interferon gamma and interleukin (IL-12). However, current evidence suggests that the Th17 pathway plays a more important role by inducing IL-23 production which is known to produce IL-17 and IL-22, vital for inflammation. Immune dysregulation, resulting in a lymphocyte-mediated destructive process, is thought to be the basic underlying mechanism behind extraintestinal manifestations of CD. An abnormal T-cell response to microbial antigens disrupts the intestinal homeostasis which results in the formation of immune complexes. These immune complexes traverse the circulation and are deposited as cutaneous granulomas.[1,2,10]

Management The diagnosis can be confirmed by a histopathological examination which characteristically shows sterile, noncaseating granulomas composed of Langerhans giant cells, epithelioid histiocytes, lymphocytes, and occasional plasma cells, localized to the superficial papillary and deep reticular dermis. Perivascular granulomatous inflammation, necrobiosis, and presence of eosinophils are other rare findings.[2,5]

There are no definite treatment guidelines. A good response has been obtained with topical steroids, systemic steroids, and antibiotics such as metronidazole and ciprofloxacin. The other drugs which have been tried include sulfasalazine, azathioprine, cyclosporine, intravenous infliximab, adalimumab, mycophenolate mofetil, oral zinc sulfate, and hyperbaric oxygen therapy.[2,4,8,9]

Conclusion

Metastatic CD is a relatively rare condition with varying presentations. We report this case to highlight the long duration of cutaneous symptoms before onset of skin lesions and the presence of vegetating plaques and erosions with a mousy odor.

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Conflicts of interest There are no conflicts of interest.

What is new?

- Metastatic CD can simulate an immunobullous disorder with vegetating lesions and a mousy odor.
- We would also like to highlight the long duration of cutaneous symptoms before the onset of skin lesions.

Article information

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Chetana Jagatgere Math and Anju George

From the Department of Dermatology, Bangalore Baptist Hospital, Bengaluru, Karnataka, India
Address for correspondence: *Dr. Anju George, Bangalore Baptist Hospital, Bellary Road, Hebbal - 560 024, Bengaluru, Karnataka, India. E-mail: dranjugeo@gmail.com*

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