Case Report

Cutaneous Crohn Disease without Intestinal Manifestations

Sadegh Vahabi-Amlashi[1,2], Sara Molkara[1], Yahya Shahrokhi[1]

Departments of [1]Dermatology, Faculty of Medicine, Mashhad University of Medical Sciences, [2]Cutaneous Leishmaniasis Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

Abstract

Extraintestinal manifestations (EIMs) are common in patients with Crohn's disease (CD). Various reactive cutaneous conditions, including erythema nodosum and pyoderma gangrenosum frequently occur as a part of EIMs. However, cutaneous metastasis of CD is rarely encountered in CD patients. Here, we report a 28-year-old female patient presenting with discharging deep fissures on genital and intergluteal regions. The result of a skin biopsy showed noncaseating granulomas. After rule out all the other differential diagnoses for granulomatous skin lesions, we believe this patient may be a case of CD, presenting with skin metastasis and GI tract involvement has not been occurred during 1-year follow-up. We suggest including cutaneous (metastatic) CD in the list of dermatologic differential diagnoses for cutaneous lesions of these sites. These lesions can occasionally precede gastrointestinal (GI) involvement by months and years, therefore, an appropriate follow-up needs to be done to detect GI lesions as soon as they appear.

Keywords: *Metastatic Crohn disease, noncaseating granulomas, vulvoperineal fissures*

Introduction

Crohn's disease (CD) is a form of inflammatory bowel disease (IBD) with key features which differentiate it from ulcerative colitis (UC). One of these differentiating features is the frequent presence of extraintestinal manifestations (EIMs) in CD. EIMs affect approximately 25%–40% of IBD patients and are known to be more frequent in CD than UC cases.[1] These signs and symptoms can occur during or before the onset of gastrointestinal (GI) symptoms.[2,3] In terms of skin lesions, they affect about 15% of IBD cases.[3,4] A broad range of cutaneous conditions can occur in CD, of which erythema nodosum and pyoderma gangrenosum (PG) is considered the most common.[1] One of the rarest entities in this category is metastatic CD (MCD) with only about 100 cases reported from 1965 to date.[5] There are several previous reports of CD manifesting as cutaneous lesions in external genitalia, abdominal, and inguinal regions.[6,7] A high level of suspicion needs to be maintained to diagnose this rare condition since early diagnosis of CD is of prognostic importance. However, since GI symptoms of CD may not be present at the time of presentation for cutaneous lesions, this can prove to be a hard task. GI involvement usually occurs before the skin lesions, however, occasionally cutaneous manifestations can precede these symptoms and interestingly, it has been suggested that these can precede GI symptoms by months and years.[7]

Here, we report the case of a female patient presenting with cutaneous lesions, who based on our investigations may be a case of MCD.

Case Report

We describe a previously healthy 28-year-old female patient complaining of skin lesions involving genital, inguinal, perineal, and gluteal regions. She recalls the onset of these lesions to be approximately 50 days ago. The initial erythematous lesions appeared on the suprapubic area then gradually spread to other regions. In our first assessment of this patient, the external genital was edematous and infiltrative; and discharging erosions and deep fissures were seen on labia major, groin, perineal and intergluteal regions [Figure 1]. There were erythematous, eroded plaques in both sides of her axilla. She denies having constitutional symptoms during this time period. She did not have a history of being bitten. She recalls having similar suppurative though less intense skin lesions

How to cite this article: Vahabi-Amlashi S, Molkara S, Shahrokhi Y. Cutaneous Crohn disease without intestinal manifestations. Adv Biomed Res 2021;10:39.

Address for correspondence:
Dr. Sadegh Vahabi-Amlashi, Cutaneous Leishmaniasis Research Center, Mashhad University of Medical Sciences, Mashhad, Iran.
E-mail: vahabis@mums.ac.ir

Received: 26 August 2019
Revised: 25 August 2020
Accepted: 19 September 2020
Published: 26 November 2021

Access this article online

Website: www.advbiores.net

DOI: 10.4103/abr.abr_180_19

Quick Response Code:

in axillary and pubic regions about 2 years ago which resolved after a course of topical corticosteroid treatment. She denied having any other symptoms, including GI, musculoskeletal, and neurological.

During the course of her recent condition, first, she had been consulted by a gynecologist who put her on valacyclovir 1 g twice a day based on an assumed diagnosis of Herpes simplex virus infection with no improvement. After few days of getting progressively worse, she was treated with cloxacillin 500 mg four times daily and rifampin 400 mg two times daily by another dermatologist based on an assumed diagnosis of Hidradenitis suppurativa. This course of treatment did not result in the improvement of her condition either.

Upon presentation to our dermatology clinic, she was afebrile and had stable vital signs. Her physical examination was unremarkable except for cutaneous lesions. She did not have oral mucosal involvement, organomegaly, or lymphadenopathy.

Initial laboratory's results of the patient were within the normal limits upon presentation. Angiotensin-converting enzyme (ACE), cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA), perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), the venereal disease research laboratory (VDRL) test, antinuclear antibody, anti-HIV antibody, hepatitis B surface antigen, and hepatitis C virus antibody were all either negative or within the normal limits. Procalcitonin levels were <0.2 and within the normal limits. In addition, her urine and blood cultures were negative: and the culture of cutaneous discharges was positive for *Staphylococcus aureus*.

Several differential diagnoses were suggested based on the clinical impression; including eczema with superimposed infection, herpetic skin lesions, hidradenitis suppurativa, hailey-hailey disease, pemphigus vegetans, and PG. Therefore, an incisional biopsy was done from cutaneous lesions on labia majora and pathologic evaluation showed ortho-and parakeratosis, acanthosis, spongiosis, and exocytosis of lymphomononuclear cells in the epidermis, increased fibroblasts, congested vasculatures, and infiltration of lymphohystiocyte and plasmocyte in the dermis. Noncaseating granulomas in the superficial dermis were noted without Leishman bodies or fungal elements [Figure 2].

Based on the histopathology report, appropriate work-up for infectious and noninfectious etiologies of granuloma was done. Tuberculoid skin lesions were ruled out since a direct smear using Ziehl-Neelsen staining was negative for acidfast bacilli (AFB) and the patient had a negative purified protein derivative skin test and normal chest radiograph. The polymerase chain reaction of the biopsy specimen was also negative for AFB. Cutaneous lesions caused by nontuberculous mycobacteria (NTM) was another possibility, but culture for NTM was negative. Fungal

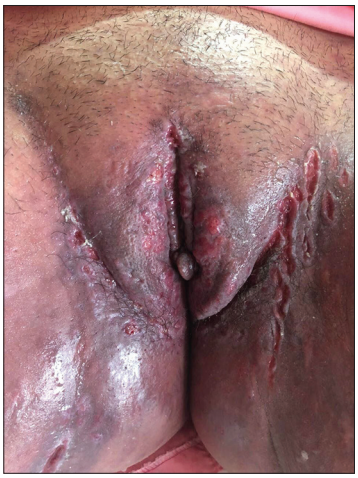

Figure 1: Infiltrative and edematous external genital with discharging erosions and deep fissures on labia major and groin

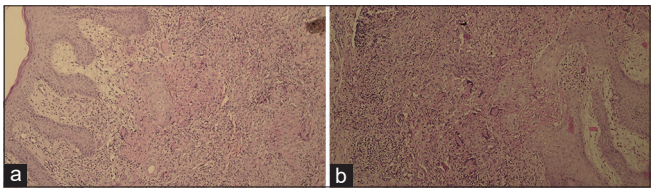

Figure 2: Section showing the ortho-and parakeratosis, acanthosis, spongiosis, and exocytosis of lymphomononuclear cells in the epidermis, increased fibroblasts, congested vasculatures and infiltration of lymphohystiocyte and plasmocyte in the dermis (a, H and E, ×40) and noncaseating granulomas in the superficial dermis (b, H and E, ×100)

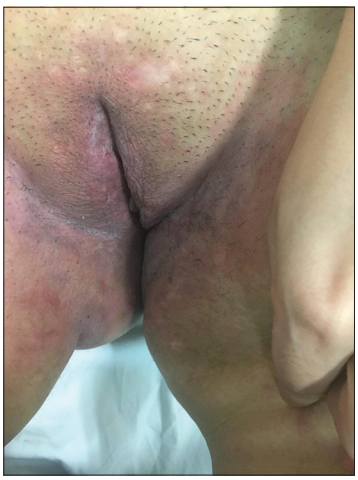

Figure 3: Improvement of the lesions after 3 months of treatment with prednisolone

infections were ruled out since a direct smear for mycotic elements and culture for deep fungal infections were both negative. As the patient had a negative VDRL test, tertiary syphilis was not the case. Granuloma inguinale was also suggested as a possible etiology and was ruled out since no Donovan bodies were noted on histopathologic examination.

In terms of noninfectious etiologies of granulomatous skin lesions, vasculitis was a possibility but no pathologic

changes compatible with it were noted on pathologic evaluation. Sarcoidosis was also ruled out based on within normal limits ACE, a normal chest radiograph, and the absence of sarcoid-type granulomas on biopsy. Due to the infiltrative character of the skin lesions on clinical examination, neoplasms, and metastases were also suggested but neoplastic changes were not seen on biopsy. The histopathologic evaluation did not detect any changes compatible with a diagnosis of PG.

Cutaneous lesions due to IBD or more specifically, cutaneous metastasis from an underlying CD was the remaining possible diagnosis and was also suggested on a gastroenterology consult. Therefore, we performed a total colonoscopy which was normal except for an anal fissure. In addition, increased wall thickness was noted on the left side of the rectum on the abdominopelvic computed tomography scan. Since we have eliminated all the other differential diagnoses for granulomatous skin lesions, we suspect this patient may be a case of CD presenting with skin metastasis and it is possible that the GI lesions are also present in another site throughout the GI tract. However, the patients' Anti-saccharomyces cerevisiae antibodies were within the normal limits and her stool calprotectin was negative.

Based on our suspicion for MCD, we put the patient on oral corticosteroids (50 mg of prednisolone daily) for 3 months, after which it was tapered and stopped during the next 2 months. Her cutaneous lesions resolved almost completely following this course of treatment [Figure 3] and is currently asymptomatic. In addition, she has not developed GI symptoms or a relapse of cutaneous lesions after 1 year follow-up to date.

Discussion

We argue that the cutaneous lesions of this case can be explained by MCD since other differential diagnoses have been ruled out. There are reports of patients presenting with cutaneous lesions in their abdominal, inguinal and genital regions as the first presentation of CD.[6,7] Interestingly, a previous case report describes the case of a 47-year-old with vulvoperineal lesions without GI involvement (based on endoscopic studies of GI tract) who was diagnosed with vulvoperineal CD and was successfully managed with metronidazole.[7] It is possible for CD to merely involve regions other than colon and this might explain the absence of lesions on total colonoscopy in our patient. For instance, a previous study found that 12 out of 17 patients who were highly suspected to have CD, but had a normal colonoscopy and small bowel X-ray findings, had lesions consistent with the condition on the wireless capsule endoscopy (WCE).[8] Furthermore, the improvement following a course of oral corticosteroid treatment also points toward MCD as a possible diagnosis. The presence of anal fissure, though common in the general population, in this clinical context increases the possibility of this diagnosis. In addition, the wax and waving character of the lesions could also be due to an autoimmune underlying condition such as CD. However, additional work-up, such as WCE, is needed to either confirm or rule out this diagnosis, especially considering the fact that our patient has not developed any GI symptoms to date, which seems quite unusual for a CD case.

Conclusion

We suggest to include MCD in the list of dermatologic differential diagnoses for cutaneous lesions of the mentioned sites when other possible conditions have been excluded through extensive work-up. Furthermore, since these lesions might precede the involvement of the GI tract by months or years, an appropriate follow-up needs to be done to detect GI lesions compatible with CD as soon as they appear.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Levine JS, Burakoff R. Extraintestinal manifestations of inflammatory bowel disease. Gastroenterol Hepatol (N Y) 2011;7:235-41.
2. Ardizzone S, Puttini PS, Cassinotti A, Porro GB. Extraintestinal manifestations of inflammatory bowel disease. Dig Liver Dis 2008;40 Suppl 2:S253-9.
3. Vavricka SR, Brun L, Ballabeni P, Pittet V, Prinz Vavricka BM, Zeitz J, *et al*. Frequency and risk factors for extraintestinal manifestations in the Swiss inflammatory bowel disease cohort. Am J Gastroenterol 2011;106:110-9.
4. Ferreira S, Oliveira B, Morsoletto A. Extraintestinal manifestations of inflammatory bowel disease: Clinical aspects and pathogenesis. J Gastroenterol Dig Dis 2018;3:4-11.
5. Teixeira M, Machado S, Lago P, Sanches M, Selores M. Cutaneous Crohn's disease. Int J Dermatol 2006;45:1074-6.
6. Math CJ, George A. Vegetating plaques in the groin: A manifestation of metastatic Crohn's disease. Indian J Dermatol 2018;63:338-41.
7. Rosmaninho A, Sanches M, Salgado M, Alves R, Selores M. Vulvoperineal Crohn's disease responsive to metronidazole. An Bras Dermatol 2013;88:71-4.
8. Legnani P, Kornbluth A. Video capsule endoscopy in inflammatory bowel disease 2005. Curr Opin Gastroenterol 2005;21:438-42.