Metastatic Genital Crohn’s Disease: Visceral devil in the details- A Case Report

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Abstract

Crohn’s disease (CD) is a chronic inflammatory bowel disease (IBD) commonly associated with extraintestinal manifestations such as arthritis, uveitis, deep venous thrombosis and erythema nodosum. Asymmetrical labial swelling and edema are the most common presentations of genital CD in females. Vulval CD can appear before or after gastrointestinal (GI) CD or it may occur simultaneously with an independent course of disease and response to treatment. Here we discuss an atypical case of metastatic vulvar CD presenting as perineal and perianal ulcers and erosions successfully treated with oral doxycycline and topical corticosteroids hence making it an exciting new treatment avenue to further explore.

Key words: Crohn’s disease, extraintestinal manifestations, metastatic vulvar CD, doxycycline

Introduction:

Crohn’s disease is a chronic relapsing-remitting IBD that can affect any part of the gastrointestinal tract usually presenting with abdominal cramps, abdominal pain, alteration of bowel habits, and weight loss.¹ Involvement of the vulva is an uncommon extraintestinal manifestation of CD (seen in 2 per cent patients)² which can be attributed to its characteristic transmural inflammatory process and may occur as a direct extension of the perineal region pathology or due to metastatic disease. Metastatic CD basically refers to granulomatous lesions of CD in a region that is noncontiguous to the gastrointestinal tract and it may precede the diagnosis of GI CD in 25 per cent of cases.¹,³,⁴ We report here a case of metastatic CD presenting as atypical vulvar lesions treated successfully with doxycycline and topical corticosteroids.

Case Report

A 35 year-old married female presented to the Dermatology and STIs OPD with chief complaints of painful perineal and perianal ulcers and erosions for the last six months. Erythema and edema of the genital region preceded the gradual development of multiple small superficial ulcers over bilateral inguinal folds extending onto the intergluteal folds and mons pubis. She complained of a burning sensation and mild itching over the erosions. There were no lesions involving the labia majora, labia minora, clitoris or periurethral areas. There was no history of rash or premenstrual flare-up of lesions.

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She did not give any history of fever, malaise, persistent lip swelling, abdominal pain or cramps. She gave a history of diarrheal episodes alternating with constipation for the past six months for which she had taken over the counter medications. There was no history of any vaginal discharge or dysuria or dyspareunia. There was no history of any high-risk sexual behaviour or no significant drug history either. She did not provide any previous history of hospitalization or treatment. She is an accountant by profession and has two children. She had nil comorbidities and no significant family history. General examination was unremarkable. The patient had mild pallor and was moderately built and nourished. There was no lymphadenopathy or lymphedema. Systemic examination - no organomegaly. Dermatological examination of the genital region revealed multiple shallow ulcers and erosions ranging from 1 x 1 cm to 2 x 1 cm present over bilateral inguinal folds, perianal region and mons pubis. The ulcers were oval, superficial with irregular margins, sloping edges and a slough with minimal granulation tissue was present on the floor. (Figure 1,2) Localized edema and erythema were present around the erosions. The labia majora, labia minora, clitoris, and periurethral region were normal. There was no associated discharge per vagina. Per speculum and per rectal examination did not reveal any abnormalities. Cutaneous examination of the rest of the body including oral mucosa, scalp, hair and nails were normal. Routine laboratory tests of blood, urine and stool were normal apart from a haemoglobin level of 9 gram% and a total leukocyte count of 11000 cells/cubic mm. Stool examination was negative for ova or cysts and there was no occult blood loss. Serological testing for LGV, Syphilis (VDRL), HIV and other STIs were negative and immunofluorescence staining was negative for spirochaetes, HSV, CMV and VZV. Filariasis was ruled out as no microfilariae could be detected in the blood smear. Mantoux test was negative and ACE levels for sarcoidosis were within a normal range. Gram stain and Tzanck smear did not reveal any microorganisms or multi-nucleate giant cells. Negative fungal stains ruled out deep fungal infections. A biopsy was performed from the genital lesions which revealed epidermal ulceration, presence of perivascular lymphocytic inflammatory infiltrate, thickened and mildly fibrotic dermis and non-caseating epithelioid granulomas which were strongly suggestive of genital CD. (Figure 3[A-D]) Upper GI endoscopy and colonoscopy did not reveal any abnormalities. X-rays of the chest and pelvic region were normal. The patient subsequently underwent CT scans of the abdomen and pelvis which did not show any evidence of Crohn’s colitis or enteritis and also ruled out genital tuberculosis. Based on these findings a diagnosis of cutaneous Crohn’s disease (metastatic vulvar CD) was made. The patient was started on tablet doxycycline 100 mg BD and topical clobetasol propionate cream and mupirocin ointment under occlusion with a gauze dressing. The patient was followed up regularly and showed gradual improvement with complete resolution of lesions after three months, following which she was put on maintenance therapy of clofazimine 100 mg BD for a month. (Figure 4)
FIGURE 1 AND 2: Dermatological examination of the genital region revealed multiple shallow ulcers and erosions ranging from 1 x 1cm to 2 x 1cm present over bilateral inguinal folds, perianal region and mons pubis. The ulcers were oval, superficial with irregular margins, sloping edges and a slough with minimal granulation tissue was present on the floor. Localized hyperpigmentation, edema and erythema were present around the erosions. The labia majora, labia minora, clitoris and periurethral region were normal.

FIGURE 3: HISTOPATHOLOGY FINDINGS: A biopsy was performed from the genital lesions which revealed epidermal ulceration, presence of perivascular lymphocytic inflammatory infiltrate, thickened and mildly fibrotic dermis and non-caseating epithelioid granulomas which were strongly suggestive of genital Crohn’s disease (A, B). Haematoxylin and eosin stain. 10x magnification.

The black arrows represent epithelioid histiocytes in Image D. Image C shows a high-power view of the perivascular infiltrate. Haematoxylin and eosin stain. 40x magnification.

FIGURE 4: HEALING KNIFE CUT ULCERS: The patient was followed up regularly and showed gradual improvement with complete resolution of lesions after three months following which she was put on maintenance therapy of clofazimine 100mg BD for a month.

Discussion

The diverse array of clinical presentations in genital CD can include ulcers, swellings, exophytic lesions, fissures, fistulas, edema and abnormal pap smears. Perianal lesions accompany 90 per cent of cases of a metastatic vulval CD which may develop before or after GI symptoms or may occur simultaneously. Our diagnosis was supported by the clinical presentation, chronicity of the lesions, biopsy findings and exclusion of other granulomatous and infectious diseases. This case illustrates the importance of considering CD as a possible diagnosis in women who present with cutaneous vulvar lesions as the only complaint without any coexisting gastrointestinal symptoms because these features may be erroneously attributed to a gynaecologic disorder. Endoscopy guided biopsy is helpful to establish the diagnosis of CD in these patients but the classical pathognomonic histological features of non-caseating granulomas are found in only 50 per cent of GI CD cases adding to the diagnostic dilemma. Other granulomatous disorders such as cutaneous sarcoidosis, deep fungal infections, mycobacterial infections, actinomycosis, lymphogranuloma venereum (Esthiomene), granuloma inguinale, cellulitis, granulomatous cheilitis, chronic lymphedema, schistosomiasis, hidradenitis suppurativa, and foreign body reactions should be considered in the differential diagnosis of vulval CD. These were ruled out in our patient by using the respective biochemical, serological and radiological investigations which included the use of stains and smears to eliminate infective etiology. The initial management of vulval CD is medical, including metronidazole, topical and intralesional corticosteroids, systemic steroids, sulfasalazine, cytotoxic agents such as azathioprine or 6-mercaptopurine and cyclosporine. A combination of oral doxycycline and topical CS can be used as an effective management strategy in the cutaneous CD without concomitant GIT
(gastrointestinal tract) symptoms as underlined in this case report.³

References