ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 12 | December - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

METASTATIC VULVAR CROHN'S DISEASE WITH "KNIFE LIKE" ULCERATION - A CASE REPORT.

Dermatology

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ABSTRACT

Crohn's disease (CD) is a chronic inflammatory disorder with relapsing and remitting course that can affect the entire length of gastrointestinal (GI) tract from oral cavity to anus and is characterized by abdominal pain, cramping, diarrhoea and weight loss. Besides GI manifestations, CD may also present with extraintestinal involvement, mucocutaneous being the most frequent (22-44%). Metastatic CD is defined as involvement of an area that is noncontiguous to the GI tract by granulomatous lesions of CD. We herein report a case of metastatic vulvar CD in a 30 year old female because of its rarity.

KEYWORDS

metastatic crohn's disease, knife like ulceration, extraintestinal crohn's disease

INTRODUCTION

Crohn's disease (CD) is a chronic inflammatory disorder with relapsing and remitting course and can affect the entire length of gastrointestinal (GI) tract from oral cavity to anus.[1,2] Crohn's disease may also present with extra-intestinal involvement, muco-cutaneous being the most frequent (22-44%).[3] Vulvar lesions occur in approximately 2% of women with CD of which asymmetrical labial swelling and oedema is the most common presentation.[3] It may occur either as direct extension of the perineal region or due to metastatic disease. Metastatic CD is defined as involvement an area that is non-contiguous to the GI tract by granulomatous lesions of CD.[2] We here report a case of metastatic vulvar CD with knife-like ulceration in both inguinal folds.

CASE REPORT

A 30 year old married woman reported to our dermatology outpatient clinic with complaints of a genital swelling since last 4 months. The swelling was progressive in nature and started with small nodules of 2-3 cm diameter at the right labia majora. Within a period of 2 months, it almost symmetrically involved both labia majora in form of swelling studded with multiple nodules. There was also ulceration in both the groins associated with severe pain and discharge. There was no history of premenstrual flare up of the lesions. Patient was housewife with one child. There was no history of any premarital and extramarital sexual exposure. There was no significant drug history. Patient was a diagnosed case of intestinal Crohn's disease and since 10 months she was on treatment with azathioprine and her disease was well controlled.

On general examination, the patient appeared normal. There was no lymphadenopathy or swelling of limbs.

Cutaneous examination of genital region revealed extremely tender bilateral grossly thickened labia majora studded with multiple almost symmetrical nodules, few of which were coalescing to form larger lesions. [Figure 1a] Localized areas with pus were also seen. Thickened labia majora almost occluded view of labia minora. On parting, normal looking labia minora, clitoris and periurethral region can be seen. Linear knife-like ulcerations were present in both the inguinal folds with discharge. [Figure 1b] There was no associated discharge per vagina. Per speculum examination did not reveal any abnormalities. Perianal area was normal.

Routine laboratory blood tests were normal apart from Haemoglobin which was 7.8 gm%. Total leukocyte count (TLC) was 9500 cells/cubic mm. Erythrocyte sedimentation rate (ESR) 38 mm at the end of first hour. Screening for human immunodeficiency virus (HIV), VDRL and serological tests for other STDs were negative. Angiotensin-converting enzyme level for sarcoidosis was normal. Mantoux's test and TB-interferon were negative.

On histopathology of lesion, epidermis showed ulceration and inflammatory infiltrate. Dermis showed loosely aggregated non-caseating granulomas composed of epithelioid cells and Langhans type of giant cells. [Figure 2a &2b] Interstitial oedema and mixed cell inflammatory infiltrate was present. The Ziehl Neelsen stain didn't show any acid fast bacilli. Based on clinical and pathological correlation a final diagnosis of metastatic vulvar crohn's disease was made.

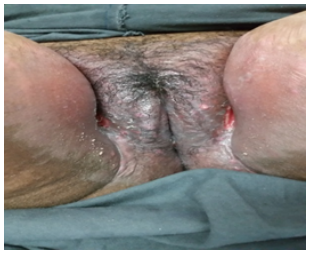

Figure 1a: bilateral grossly thickened labia majora

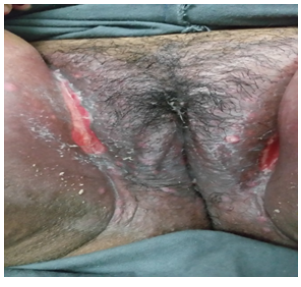

Figure 1b: Linear knife-like ulcerations were present in both the inguinal folds with discharge.

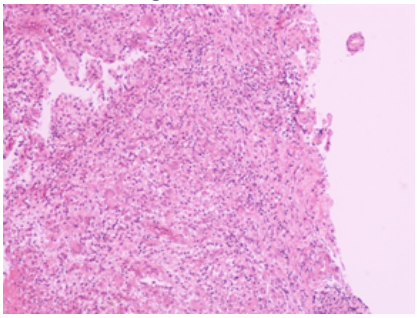

Figure 2a: Epidermis showed ulceration and inflammatory infiltrate. Dermis showed loosely aggregated granulomas, interstitial oedema and mixed cell inflammatory infiltrate. (H & E, 4X)

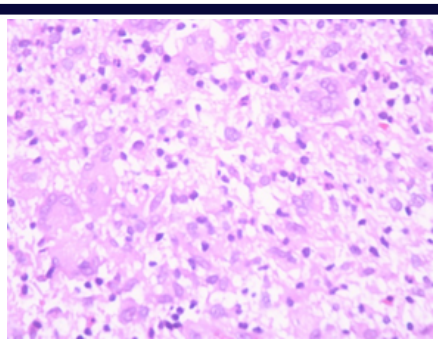

Figure 2b: non-caseating granulomas composed of epithelioid cells and Langhans type of giant cells. (H & E, 40X)

DISCUSSION

CD, also known as Crohn's syndrome and regional enteritis, is a type of inflammatory bowel disease (IBD) that may affect any part of the GI tract from mouth to anus. [1] Cutaneous involvement is present in 22-44% of cases. [3]

Cutaneous manifestations are not uncommon and may be classified into (i) peri-anal and peri-stomal lesion (the most common presentation); (ii) different skin lesions associated with CD include erythema nodosum, pyoderma gangrenosum, sweet's syndrome, acrodermatitis enteropathica, epidermolysis bullosa acquisita; (iii) granulomatous cutaneous lesions separated from the affected gut by healthy tissue known as metastatic lesions.[4]

Vulvar metastatic Crohn's disease was first described by Parks et al in 1965, and this entity was labeled as metastatic Crohn disease first in 1970 by Mountain.[3] Overall, only about 100 cases of vulvar CD have been reported in the literature since the first description.[3] In 25% of cases it may precede the diagnosis of intestinal CD [5,6]

Barret et al [4] clinically classified vulvar CD into following four major types:

The most prevalent type is vulvar swelling or edema which is mostly asymmetrical and inflammatory. Ulceration can occur which can be single or multiple, asymptomatic or painful, aphthoid and superficial or deep linear knife-like. Hypertrophic and exophytic lesions emerge from chronic inflammation and recurrent infections causing destruction of lymphatic vessels. The final type is chronic suppuration and abscess formation.

Patients are frequently asymptomatic but they can also have a different range of symptoms such as vulvar pain, pruritus, vulvar discharge or dyspareunia.

As the vulvar metastatic CD (MCD) is a relatively rare entity and clinical lesions are nonspecific and cause diagnostic dilemma, histopathological analysis is a cornerstone of diagnosis along with clinical correlation. A complete diagnostic workup by a multidisciplinary team is necessary before making a definitive diagnosis. Differential diagnosis includes other granulomatous diseases such as sarcoidosis, mycobacterial diseases, fungal infections, lymphogranuloma venereum, pyogenic infections, hidradenitis suppurativa, and syphilis.

The evolution of vulvar CD is unpredictable, ranging from cases of spontaneous healing to lesions refractory to medical therapy that require surgery. Medical treatment is the first line of treatment and surgery should be reserved for refractory lesions. Several medical regimens have been proposed with variable results and include topical, intralesional and systemic corticosteroids, antibiotics, 5-aminosalicylates, immunosupressors, and anti-tumor necrosis factor alpha (TNF-α) agents. [7]

CONCLUSION

Metastatic CD represents another 'great imitator' for which a complete diagnostic workup and involvement of multiple specialities is required to correctly recognize and treat this condition which has significant physical, mental and social morbidity to the patient and has a severe impact on quality of life.

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