Metastatic Crohn's disease

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ABSTRACT

Crohn's disease, first described in 1922, is characterized by segmental granulomatous inflammation of the intestinal tract and frequently involves the cutaneous tissues as well. Cutaneous Crohn's disease (CCD) is synonymous with metastatic Crohn's disease (MSD). A case of CCD, without any gastrointestinal involvement is reported for its rarity.

Key words: Cutaneous crohn's disease, metastatic crohn's disease, non caseating granulomas

INTRODUCTION

Crohn's disease, first described in 1922, is characterized by segmental granulomatous inflammation of the intestinal tract that frequently involves the cutaneous tissues as well. Cutaneous Crohn's disease (CCD) is synonymous with metastatic Crohn's disease (MCD). A case of CCD without any gastrointestinal involvement has been reported for its rarity.

CASE REPORT

A 20-year-old woman, mother of a 1.5-year-old child, presented with a swelling in the genitalia of 6 months duration. Cutaneous examination revealed a single, non-tender, pedunculated, polypoid growth, about 6 × 7 cm, hanging from the left labium minus [Figure 1]. She had minimal swelling of the left labium majus. There was a tiny ulcer near the superior commissure of the labia majora. However, many scars were seen on the medial aspect of both the thighs, which were attributed to pruritic ulcers in childhood, details of which were unavailable. Her vagina, cervix, and uterus were normal. Systemic examination, including the eyes and oral mucosa, did not reveal any abnormality. Filariasis being endemic in Cuddalore district of Tamil Nadu, elephantiasis of external genitalia due to filariasis, besides, lymphogranuloma venereum and granuloma inguinale were also considered.

A clinical diagnosis of soft fibroma was entertained and an excision biopsy of the nodule was undertaken. Histolopathological examintion (HPE) revealed multiple non-caseating granulomas, edema, and dense lymphocytic infiltration in the dermis. Ziehl–Neelsen staining for *Mycobacterium tuberculosis* was negative [Figure 2]. Special stains for fungal organisms were not performed.

A week later, at the time of suture removal, the patient presented with multiple typical "knife-cut" ulcers on the external genitalia, in the inguino-crural fold, interlabial creases, and natal cleft, leaving no doubt in the diagnosis of CCD [Figure 3]. Biopsy from knife cut ulcers could not be carried out since the patient was not willing to undergo the procedure. She did not have any intestinal symptoms or pain abdomen at any time in the past or present.

On investigation, she was found to have hypochromic microcytic anemia (Hb: 10.5 gm/dl) and mild neutrophilic leucocytosis. Other relevant investigations including Mantoux test, ultrasonogram of the abdomen, and chest radiograph were not contributory. Both Enzyme-Linked ImmunoAssay (ELISA) test for HIV and VDRL test were negative.

The patient was referred to a gastroenterologist for further work up, and no gastrointestinal involvement was reported. With this background, a diagnosis of CCD was made.

The patient was managed with metronidazole 400 mg t.i.d. for 1 week and ciprofloxacin 500 mg bid. for 10 days in addition to oral prednisolone at 30 mg/day, which was gradually tapered over a period of 6 weeks.

Access this article online
Website: www.idoj.in
DOI: 10.4103/2229-5178.126029
Quick Response Code:

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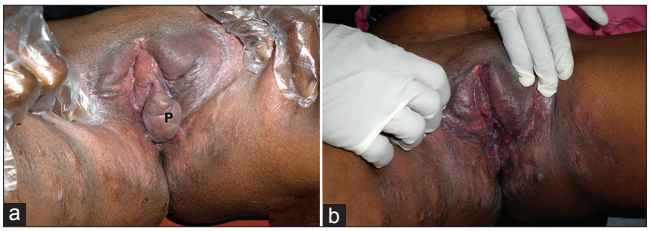

Figure 1: (a) Single, non-tender, pedunculated, polypoid growth, about 6 × 7 cm, hanging from the left labium minus (P) and minimal swelling of the left labium majus, (b) Multiple "knife-cut" ulcers on the external genitalia, in the inguino-crural fold, and in the interlabial creases

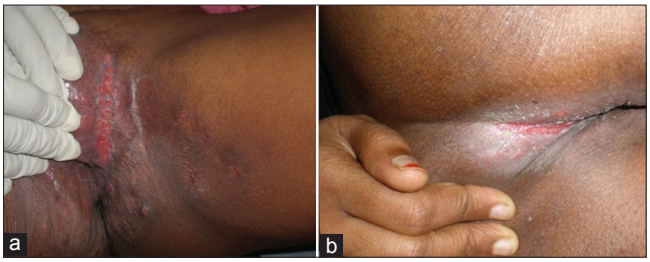

Figure 3: Multiple typical "knife-cut" ulcers on the external genitalia in the inguino-crural fold, (a) interlabial creases, and (b) natal cleft

She reported after 2 weeks with ulcers showing signs of healing, and by 6 weeks, they resolved. Later, the patient was lost on follow-up.

However, she reappeared 2 years later with massive swellings of both the labia majora, more marked on the left labium majus. She gave a history of having had a full-term normal child, 3 months earlier, delivered by an elective lower segment caesarean section (LSCS) in view of the edema of the labia majora [Figure 4].

DISCUSSION

CCD is relatively rare with fewer than 100 cases reported in the literature.[1] Two-thirds of the patients were women with a mean age of onset at 34.5 years, though our patient had lesions much earlier at 20 years of age.

Vulvar involvement in CD may be by virtue of contiguity as a direct extension of intestinal involvement or non-contiguous (metastatic) in which there is no connection between the vulva and the bowel.[2] In a review by Andreani *et al.*, 25% of vulvar CD did not have any intestinal involvement at the time of vulvar lesion.[3] It is in these cases that making a correct diagnosis becomes difficult, as was the experience of the present authors, who also missed the correct clinical diagnosis initially.

Mucocutaneous manifestations occur in up to 44% of patients,[4] but they do not necessarily mirror intestinal involvement[5] akin to our patient, who did not have any intestinal symptoms

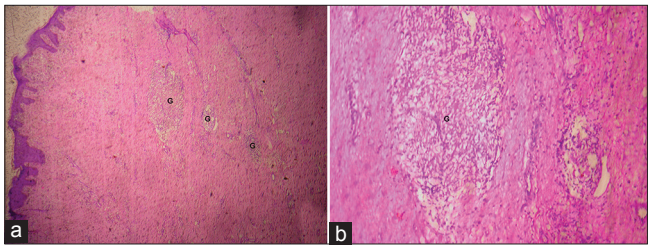

Figure 2: Multiple non-caseating granulomas in the dermis (G) (a) H and E, ×20; (b) H and E, ×40

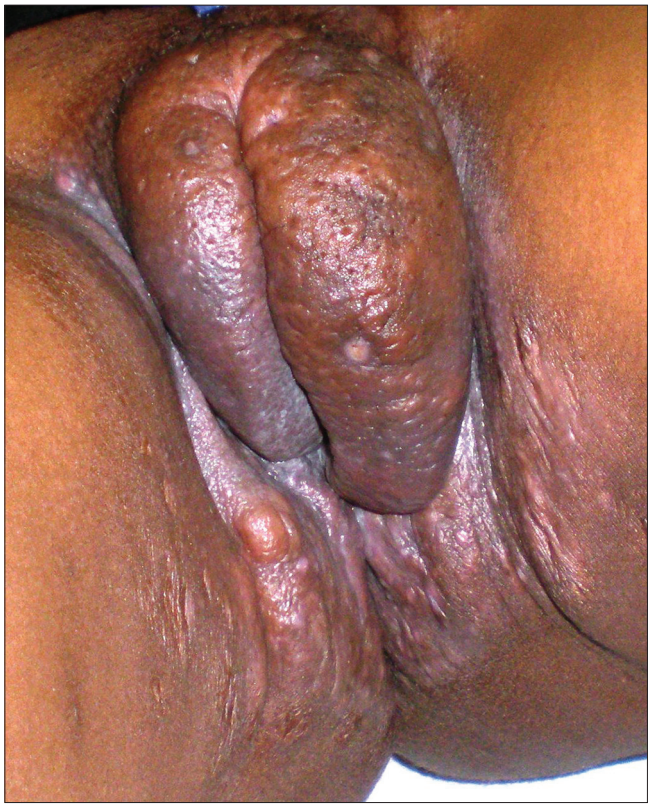

Figure 4: Massive edema of genitalia more marked on the left labium majus

at any time. CCD can occur in association with or preceding intestinal Crohn's disease. Vulvar ulcers may precede intestinal manifestations by up to 18 years.[6] No intestinal involvement was found in the present case, similar to that in other reports.[3,5,7]

Skin manifestations in CD can be categorized as either histologically specific or non-specific (reactive). Specific lesions are characterized by non–caseating granulomas, similar to those seen in the intestinal lesions. Nonspecific or reactive skin manifestations include entities like pyoderma gangrenosum, erythema nodosum, and oral aphthae.[4] However, such non-specific manifestations were not observed in the present case.

Among patients with CCD, 70% present with genital lesions. Vulvar CD presents with erythema and unilateral or bilateral

labial swelling. Chronic edema leads to firm coalescing papules and fibrotic nodules, which could be mistaken for lymphatic obstruction.[8] Tuberculosis and filariasis, more common conditions in this part of South India, were also considered initially, before the biopsy report was available.

The differential diagnosis of granulomatous vulvar lesions includes conditions like sarcoidosis, foreign body implantation, hidradenitis suppurativa, mycobacterial, or deep fungal infections and granuloma inguinale that were ruled out by the HPE and other relevant investigations.

Treatment of CD requires systemic administration of various antibiotics, immunosuppressive agents, and anti-inflammatory agents such as corticosteroids, azathioprene, sulfasalazine, or 6-mercaptopurine. CCD is rare and treatment trials have not been performed. Infliximab and adalimumab, cyclosporine, and mycophenolate mofetil have been found beneficial by various authors in addition to surgical intervention for fistulae.[9] Surgical excision may often be complicated by wound dehiscence and recurrence of the disease.

Our patient responded well to administration of metronidazole and systemic corticosteroids. She was advised frequent reviews by the gastroenterologist to help in early identification of gastrointestinal involvement and complications and its management, but she was lost to follow-up; however, she resurfaced 2 years later with another child of 3 months age and no specific complications except for the massive swelling of the labia majora.

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Cite this article as: Lanka P, Lanka LR, Sylvester N, Lakshmi MD, Ethirajan N. Metastatic Crohn's disease. Indian Dermatol Online J 2014;5:41-3.

Source of Support: Nil, **Conflict of Interest:** None declared.