

Vulvo-Perineal Crohn's Disease: A Challenging Diagnosis!

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ABSTRACT

Crohn's disease (CD) is a chronic inflammatory bowel disease with polymorphous cutaneous manifestations. Vulvar localization is rare and may be either isolated, revealing the disease, or associated with digestive manifestations. Treatment is difficult and not well codified.

Based on 4 case reports of vulvo-perineal involvement, we present a reminder of the diagnostic and therapeutic challenges of this particular site.

In 2 out of 4 patients, vulvar involvement was incipient, with no gastrointestinal symptoms at the time of diagnosis. The vulva may present with many dermatological manifestations: knife-like ulcerations, oedema, labial hypertrophy, lymphangiectasia, abscesses and fistulae. The presence of non-caseating granulomas on histology may help to confirm the diagnosis.

Vulvar CD affects quality of life and requires long-term medical and surgical management with a multidisciplinary approach involving gastroenterologists, dermatologists, gynecologists and pathologists.

KEYWORDS: Vulvo-perineal- Crohn's disease- knife-shaped ulcerations- non-caseating granuloma.

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INTRODUCTION

Crohn's disease (CD) is a chronic inflammatory intestinal disease with polymorphous cutaneous manifestations. Vulvo-perineal localization is rare and may be either isolated, revealing the disease, or associated with digestive manifestations. However, cutaneous so-called metastatic lesions of the vulva have been reported in the literature, they are still under recognized and clinically challenging for dermatologists, gastroenterologists as well as for gynecologists, with numerous differential diagnoses, especially among venereal diseases, and require a multidisciplinary approach. The complexity of diagnosing vulvar Crohn's disease is also due to its overlap with other conditions such as hidradenitis suppurativa and vulvar lichen sclerosus [1]. Management is difficult and not well codified. We report on four cases of vulvo-perineal involvement of CD to review the different clinical aspects as well as the diagnostic and therapeutic challenges of this particular site [2].

REPORT OF CASES

Case 1:

A 23-year-old female patient presented with a 4-year history of chronic diarrhea and abdominal pain in flares with inflammatory nodular vulvo-perineal lesions. Physical examination revealed deep, linear, knife-shaped ulcers of the inguinal folds; lymphoedema of both vulva with a pseudo-tumoral appearance; pseudo condylomas in the vulvar region, ano-perineal region and around the ulcers associated with oral enanthema and perioral erythema (*Figure 1*).

Skin biopsy revealed a neutrophilic dermatitis with granuloma, upper gastrointestinal fibroscopy a gastrobulbitis with duodenitis, colonoscopy an oedematous inflammatory mucosa with polyploid lesions of the rectum and impermeable sigmoid stenosis. The patient was treated with sulfasalazine and metronidazole.

Case 2:

A 19-year-old female patient presented with a history of chronic diarrhea with vulvar lesions since the age of 10 years. Physical examination revealed inflammation and hypertrophy

Vulvo-Perineal Crohn's Disease: A Challenging Diagnosis!

of the labia majora; linear knife-like ulcerations of the inguinal and intergluteal folds; cheilitis with labial and nasal oedema (**Figure 2**).

Skin biopsy revealed epithelioid and giant cell granuloma of the dermis without necrosis. The patient was treated with metronidazole.

Case 3:

A 17-year-old female patient with no medical history presented with perianal lesions since the age of 13 years, followed by vulvar lesions 1 year ago. Physical examination revealed Perianal and labial pseudocondylomatous lesions (**Figure 3: a**).

Skin biopsy showed non-caseating granulomas. (**Figure 3: b,c**). Colonoscopy and upper fibroscopy were without abnormalities.

The patient was treated with metronidazole and azathioprine.

Case 4:

A 49-year-old woman with no previous medical history presented with papulo-nodular lesions and ulcerations of the vulva and gluteal folds. Physical examination revealed papulo-nodular lesions of the labia ; knife-shaped ulcerations in the inguinal and gluteal folds (**Figure 4**).

Skin biopsy revealed non-caseating granulomas. Colonoscopy and upper fibroscopy revealed no abnormalities.

1 month later, the patient presented with erythema nodosum on both legs and was put on metronidazole combined with corticosteroid therapy at a dose of 1 mg/kg/day with progressive degeneration over a period of 3 months with a complete disappearance of the dermo hypodermal nodules and partial regression of the inflammatory perineal lesions and ulcerations.

As the lesions persisted, the patient was treated with a TNF inhibitor.

DISCUSSION

Vulvar Crohn's disease is a chronic recalcitrant condition that affects patients' quality of life and requires long-term

multidisciplinary management. In 2 out of 4 patients, vulvar involvement was incipient, with no gastrointestinal symptoms at the time of diagnosis. The presence of non-caseating granulomas on histology may help to confirm the diagnosis [3]

The vulva may present with other dermatological manifestations: oedema, labial hypertrophy, lymphangiectasia, knife-shaped ulcerations, abscesses and fistulae. Histological findings such as the presence of non-caseating granulomas, granulomatous vasculitis or dermal lymphangitis may help to confirm the diagnosis. The main differential diagnoses to consider are condyloma, vulvar intraepithelial neoplasia and acquired lymphangiectasia.

Cutaneous symptoms may precede, coincide with, or follow gastrointestinal involvement. Topical, intralesional and systemic corticosteroids, TNF inhibition, interleukin 12/23 inhibition, azathioprine, antibiotics, and surgery have shown benefits in some patients; however, not all patients experience improvement with these treatments [4-5].

Despite the variety of traditional therapeutics for vulvo-perineal CD, inconsistent outcomes demonstrate the need for more effective treatments; Upadacitinib, a Janus kinase (JAK)1 inhibitor, was approved in 2023 as a potential therapeutic option for these patients [6-7].

CONCLUSION

Vulvar CD affects quality of life and requires long-term medical and surgical management. It requires a multidisciplinary approach involving gastroenterologists, dermatologists, gynecologists and pathologists.

Vulvar involvement may reveal de diagnosis and the presence of non-caseating granulomas on histology may help to confirm the diagnosis.

Surgery remains limited to medical treatment failure or resection of unsightly lesions. Prospective studies or case series with long-term follow-up are still missing to guide the treatment of this condition.

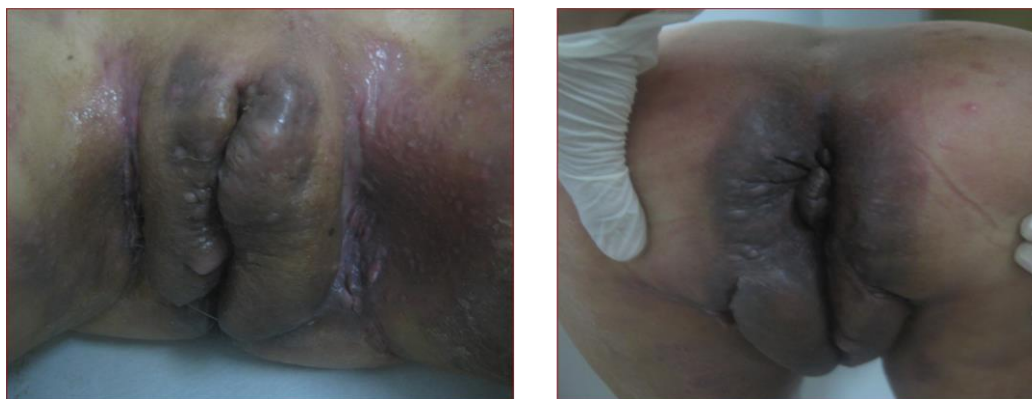


Figure 1: Clinical aspect of vulvo-perineal Crohn's disease of the first patient

Deep, linear, knife-shaped ulcers of the inguinal folds; lymphoedema of both vulva with a pseudo-tumoral appearance

Vulvo-Perineal Crohn's Disease: A Challenging Diagnosis!



Figure 2: Clinical aspect of vulvo-perineal Crohn's disease of the second patient

Inflammation of the intergluteal region with hypertrophy and inflammation of the labia; linear knife-like ulcerations of the inguinal and intergluteal folds.

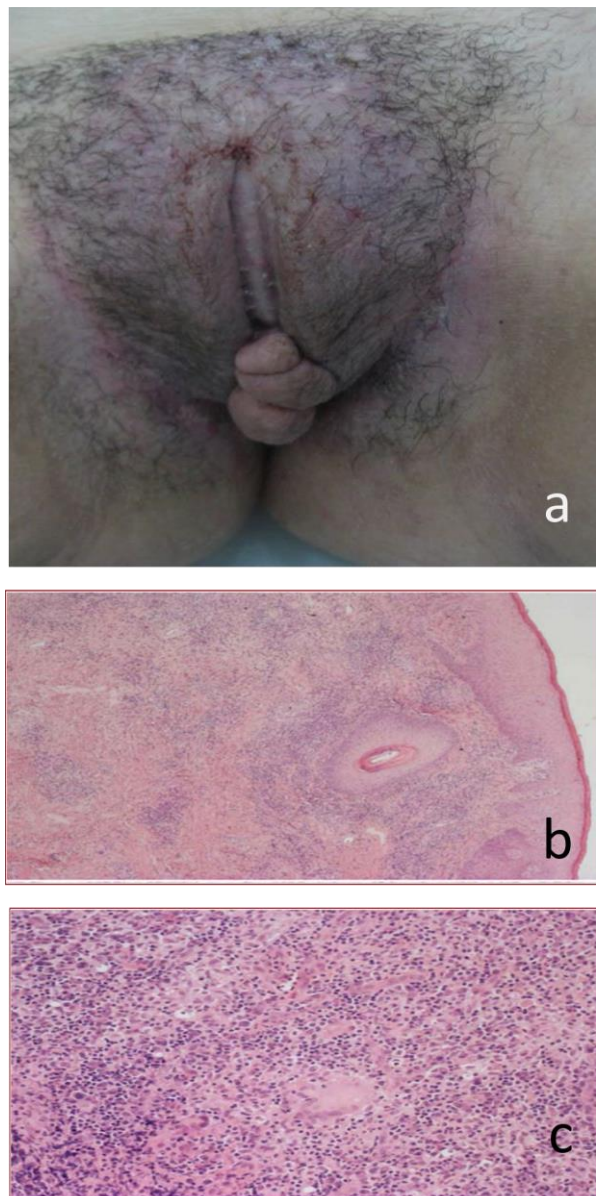


Figure 3: Clinical aspect of vulvo-perineal Crohn's disease of the third patient

Perianal and labial pseudocondylomatous lesions with vulvar and inguinal erythema

Vulvo-Perineal Crohn's Disease: A Challenging Diagnosis!

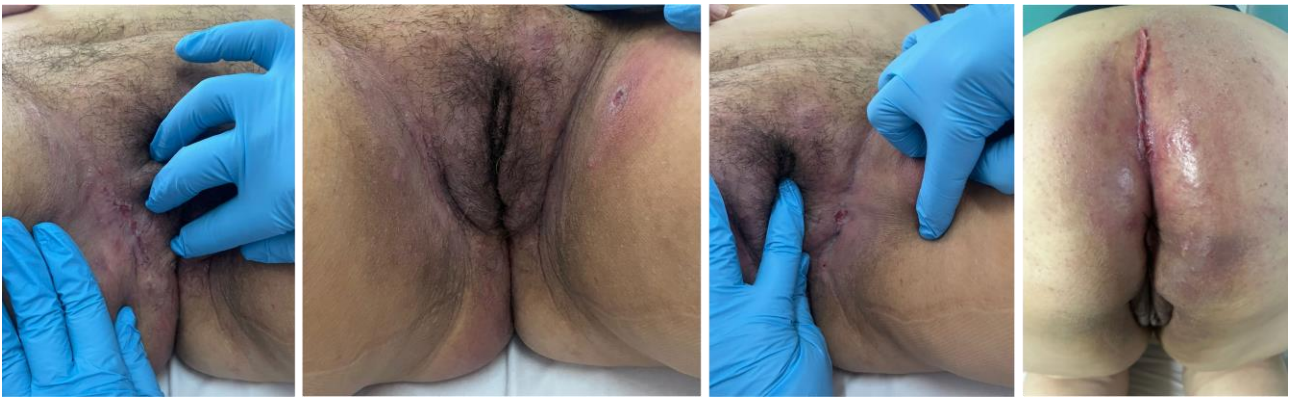


Figure 4: Clinical aspect of vulvo-perineal Crohn's disease of the fourth patient:

Papulo-nodular lesions of the labia ; knife-shaped ulcerations in the inguinal and gluteal folds

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