Vulvar lesion in Crohn's disease: a new case report


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ABSTRACT

The mucocutaneous manifestations are, with the osteo-articular attacks, the most frequent of the extradigestive attacks of the Crohn’s disease. While some of these events evolve alongside the digestive disease, others evolve on their own. Sometimes they can even precede the appearance of intestinal manifestations by several months, which then poses diagnostic problems.

Recto-vaginal fistulas and ovarian involvement in Crohn’s disease have been widely reported in the literature (1; 2). However, there are few series in the literature reporting the genital complications of CD, let alone regarding vulvar lesion.

We report a new case of vulvar lesion in Crohn's disease.

Keywords: Vulvar lesion, Crohn’s disease, case report
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**Case report**

A 30-year-old divorced woman with 2 children was followed for Crohn's disease evolving for 7 years and treated with 5 ASA. The patient was lost for 5 years. She came back with a polyfistulous perineum and vulvar swelling and ulceration. During the interview, the patient reported bloody diarrhea at the rate of 3 to 4 stools / day associated with a rectal syndrome, evolving by flares/remission, and appearance of several perineal fistulous orifices and vulvar swelling and ulcerations for 4 years for which she has never consulted.

Clinically, the perineum assessment found a reshaped perineum, retracted with multiple external fistulous orifices, some of which are budding. There is also a remodeled vulva, a painful edema of the labia majora with a hypertrophied clitoris (figure 1).

Anatomopathological study of vulvar biopsies revealed a giganto-epithelioid granuloma without caseous necrosis, compatible with Crohn's disease (figure 2).

Colonoscopy showed an erythematous mucosa, seat of ulcers wide in geographical map without healthy mucosal interval at the recto-sigmoid level, beyond, the mucosa is discreetly erythematous with individualization of healthy mucosal interval. The anamopathology of colon biopsies showed a morphological aspect of an epithelio-giganto-cellular reaction without caseous necrosis in favor of Crohn's disease.

Pelvic and enteral MRI showed multiple ano-perineal complex fistulas with perineal and pelvic infiltration, as well as circumferential and regular thickening of the extended colon to the sigmoid colon.

Therapeutically, the patient received infliximab at 5 mg / kg (including three injections at weeks 0, 2 and 6 and then every 8 weeks) with onset of significant decrease in digestive, ano-perineal and vulvar manifestations. Surgical treatment consisting of an excision of tumor masses with perineal plasty and colostomy has been proposed.

**Figure 1 : Remodeled vulva, edema of the labia majora with hypertrophied clitoris and multiple perineal fistulas**
Discussion:
The Crohn's disease (CD) is a chronic granulomatous inflammatory disease that can affect any part of the digestive tract. It is associated with frequent and polymorphic extraintestinal manifestations, the most frequent of which are musculoskeletal, dermatological, ophthalmological and hepatobiliary.

The dermatological manifestations during Crohn’s disease include reactive dermatosis, autoimmune dermatosis, diseases related to nutritional deficiencies, cutaneous manifestations secondary to treatments and specific granulomatous lesions [3]. The latter form can affect the genitals, but also other parts of the skin [4;5].

The vulvar Crohn’s disease was described for the first time by Park et al. [6], and there are few series in the literature reporting genital complications of CD, let alone vulvar lesion.

The involvement of the vulva during Crohn's disease can occur either by direct extension from the perineal region or by metastasis. Metastatic Crohn's disease is defined as granulomatous lesions separated from the affected regions of the digestive tract by a territory of healthy skin [7; 8].

In a review of the literature, Andreani et al found that 91% of cases of vulvar Crohn's disease were metastatic, while only 5% were contiguous from the perineal region. [10] In the same study, 25% of Vulvar Crohn’s cases had no digestive tract at the time of diagnosis.

Clinically, vulvar lesions can be associated to varying degrees with edema, papules, painful nodules, endovaginal fistulas, Bartholin's gland abscesses and ulcerations [1,2]. The diagnosis must be evoked in the presence of deep vulvar linear ulcers called "stab wounds" or painful indurated labial edema, often asymmetrical.

The differential diagnoses of vulvar localization include sarcoidosis, tuberculosis, lymphogranuloma venereum, pyogenic infections, hidradenitis suppurativa, intertrigo and syphilitic lesions. The definitive diagnosis can only be made by performing a biopsy that reveals a non-caseating granulomatous lesion [9].

The natural evolution of vulvar Crohn's disease is unpredictable. Although some cases resolve spontaneously, the majority of cases persist [12, 13].

Due to its rarity, there are no randomized trials that suggest special treatment for vulvar Crohn's disease [14].

The oral steroids were associated with long-term improvement [15], but other medications, oral metronidazole, azathioprine and 6-mercaptopurine also showed good results [16].

The Infliximab has been effective in cases of vulvar Crohn’s disease resistant to other immunosuppressants, including prednisolone, azathioprine and oral cyclosporine [8].

The adalimumab and certolizumab have also been shown to be effective in cutaneous Crohn's disease [17,18].

The surgery may be necessary for vulvar Crohn's disease refractory to all medical treatments. Localized surgical excision frequently leads to localized recurrence and suboptimal wound healing. Radical vulvectomy is usually necessary in difficult cases [9].

Conclusion:
The vulvar localization is a rare extra-intestinal manifestation of Crohn's disease. A multidisciplinary approach involving gastroenterologists, gynecologists, pathologists and dermatologists is recommended to identify this entity and to initiate appropriate treatment.

As standard therapy is lacking, the current therapeutic approach is case-by-case and can be escalated according to the severity of the disease.

References: