Case Report

Crohn's Disease of the Vulva: A Unique Presentation with an Unconventional Treatment Approach

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Abstract

Vulva's Crohn's disease (CD) is a rare condition that can be easily misdiagnosed if not thoroughly investigated. The typical presentation of the disease is swelling, deep linear fissures, erythema and painful, and progressive ulceration of the labia. There are various treatment protocols, yet none of them have been established as gold standard. Surgery is not recommended, and medical treatment is the current standard of care. We aim to report an unusual presentation of vulvar CD and discuss the treatment option of surgery, a valuable alternative when medical therapy yields ineffective results. A 36-year-old patient, whose initial symptoms had begun 7 years ago, presented with labial fistulas and multiple tissue papillomas. During the disease course, the patient had previously received corticosteroid and antibiotic treatments of limited benefit. Vulvar hypertrophy was significant, and papilloma formation related to the extension of the knife-like ulcers had left the outer genitals dysmorphic. The patient underwent two consecutive partial vulvectomies with 1 year apart, along with oral moxifloxacin treatment. Partial vulvectomies reduced the amount of hypertrophic tissues and papillomas and allowed for asymptomatic urine output. The patient also had psychological relief and experienced less intervening intertriginous fungal infections. Due to the ongoing inflammatory process of CD, symptomatic and clinical relief by stabilizing the condition is of utmost importance. Although medical treatment is frequently sufficient to stabilize symptoms of vulvar CD, surgery certainly reserves a place in the treatment algorithm when the patient is unresponsive to other treatment modalities.

Keywords: Crohn's disease, genital surgery, vulvar granulomatosis

Introduction

Crohn's disease (CD) is a chronic granulomatous inflammatory disease that primarily affects the gastrointestinal tract. Extraintestinal manifestations can also be present in up to 45% of patients, including involvement of the joints, oral cavity, skin, and eyes.[1] Cutaneous manifestations are relatively common, as studies report dermatological findings in 14%–44% of patients, which may present as solitary skin lesions or involve various sites in the form of papules, plaques, nodules, or ulcers.[2] Among the most common cutaneous findings are fissures and fistulas arising through the direct extension of the gastrointestinal disease.[3] Cutaneous lesions can be peri-anal and peri-stromal lesions; skin lesions in association with CD like erythema nodosa, epidermolysis bullosa acquisita, pyoderma gangrenosum; or granulomatous lesions that are discontinuous with the diseased part of the gut.[4]

Although extraintestinal findings of CD frequently manifest itself through enteric fistulas, cutaneous involvement without a direct connection to the digestive tract may also be seen. Vulva's CD (VCD) is a rare condition with approximately 124 cases reported so far and can occur without any evidence of gastroduodenal CD.[2] Although VCD is frequently associated with luminal CD, this may not always be the case, as approximately 25% of patients with vulvar lesions precede the diagnosis of digestive CD.[4] This circumstance makes the differential diagnosis of the entity quite difficult, as these patients only present with vulvar symptoms.

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Vulvar involvement may be asymptomatic or present itself as swelling or edema, deep linear ("knife–cut") fissures, erythema, and progressive and painful ulceration of labia.[1,2,4] Due to the immunologic origin of the disease, ongoing chronic inflammation can cause lymphatic vessel obstruction, which can lead to lymphangiectasia.[4] Although pruritus and vulvar pain have been reported, the most common finding is unilateral labial hypertrophy and hypertrophic lesions.

Differential diagnoses of vulvar CD include other granulomatous disorders, including sarcoidosis, tuberculosis, lymphogranuloma venereum, pyogenic infections, hidradenitis suppurativa, intertrigo, and syphilitic lesions.[1] Diagnosis is made on a clinical and histopathological basis and requires an extensive multidisciplinary approach. Treatment is usually medical, as surgery is not recommended and is reserved only for refractory cases.

Herein, we report the case of an isolated vulvar CD, with a unique clinical presentation, and discuss the diagnostic and treatment approaches adopted for the patient.

Case Report

A 36-year-old female patient was referred from the general surgery department with symptoms of diffuse vulvar edema, ulcerations, and hypertrophic lesions. She was in routine follow-up for widespread perianal fistulas. No pathological abnormality was found in colonoscopy and gastroduodenoscopy. Patient's current complaints started right after perianal involvement 7 years ago and showed a postero-anterior progression, all of which significantly deterred her quality of life.

During the physical examination, largely oedemated and endurated bilateral labia majora was noted. Numerous chronic suppurative and nodular lesions invaded the labia majora [Figure 1]. The most striking and unique characteristic of the patient's clinical presentation was the presence of numerous mucosal papillomas caused by the extension of contralateral fissures leading to the detachment of tissue from the labia and clitoral hood. No internal genital pathology was found in the gynecological workup and the cervical pap-smear was normal. The patient stated that she was initially diagnosed with nonspesific granulomatous disease and received oral prednisolone and adalimumab treatment but did not benefit from it. In addition, she received metronidazole treatment but discontinued it after no improvement was observed.

Due to the multiple papillary mucosal extensions caused by the adjoinment of the contralateral linear ulcers, a partial vulvectomy was performed. Excision of these pedunculated tissues from the labia majoras, labia minoras and the perineal endurated areas was undertaken. Secondary closure was intended when primary closure was not suitable. The patient was followed up with daily Bactigras® (Smith and Nephew) wound dressings until epithelialization was complete [Figure 2].

In the early postoperative phase, ongoing inflammation was observed. Intraoperative vaginal culture was reported back as *Escherichia coli* and *Enterococcus faecalis* infection and the patient was started on 4 mg × 500 mg intravenous imipenem regime. Once vaginal discharge and vulvar swelling subsided, the patient was discharged with 1 mg × 400 mg oral moxifloxacin treatment. During the follow-ups, the patient improved dramatically under moxifloxacin treatment which she continued for 2 months. Patient's intertriginous linear ulcers healed spontaneously, and pain subsided.

The histopathological analysis revealed active chronic inflammation characterized by abscesses, fistulas, and dispersed noncaseified granuloma formation, as well as secondary vasculitic findings. Despite the fact that the patient had no prior inflammatory bowel disease, these findings were concordant with cutaneous/genital CD. Unfortunately, the patient lived abroad and could not present for routine follow-ups after surgery.

At the patient's 1st year follow-up, a bulky mucosal papilloma of the right labia majora adjoining multiple gapes in the right labia minora was observed as a result of the deepening of



Figure 1: Preoperative presentation of the patient demonstrating generalized vulvar erythema, hypertrophy and edema, bilateral linear fissures, and papillomatous tissue extensions along the labias and clitoral hood

linear fissures from the previous disease process. The patient had recurrent linear ulcers in the bilateral intertriginous area of the genitalia and a circular ulcer with a 1 cm diameter at the vaginal opening [Figure 3]. The clitoral hood was aberrant with hypertrophy. Both labia majora had swelled again.

The patient underwent a second operation in which disfigured parts of the right labia majora and minora were excised and the ulcers were gently debrided. Intraoperative smear was taken from the ulcers and revealed Klebsiella Pneumonia and Candida Albicans infection. The patient was prescribed 150 mg of single-dose oral fluconazole and intravenous ceftriaxone treatment of 1 g/day for 14 days. In the postoperative 1st week, spontaneous healing of linear ulcers was noted [Figure 4]. The pain had completely subsided apart from the mild macerations in the groin region which was due to the healing Candida infection. The overall appearance was normal and the surgical incisions healed uneventfully. The patient has been followed up without clinical progression for 4 months.

Discussion

Vulvar involvement in CD was first described by Parks in 1965.[5] VCD may accompany gastrointestinal CD or may be a separate entity. Among the signs of VCD are edematous labia majora and minora, vulvar ulcerations, and hypertrophic lesions, all of which were present in our patient. In the differential diagnosis, other infectious, inflammatory, and malignant diseases of the vulva must be ruled out with meticulous investigation. While noncaseating granulomas are pathognomic, diagnosis is made according to clinicopathologic findings.

Clinical signs can be misleading as they can occur in most inflammatory cases. Tissue biopsies are therefore required to elicit the origin of the cause. The initial work-up of a vulvar granulomatous disorder includes gynecologic examination, cervical smear, and serologic tests, which aid in ruling out malignancies and venereal diseases. Spirochetes, herpes simplex virus, and varicella zoster virus can be ruled out with immunofluorescence staining.[6] In case of vaginal discharge, a vaginal smear and culture must be obtained to determine the infectious cause. If abscess or fistulated lesions are present, ultrasound or pelvic MR might be considered. In addition, the patient must be questioned for any abdominal complaints, sexual activity, and eye or joint dysfunctions. Our patient presented with a unique clinical finding of mucosal papillomas caused by the extension of contralateral fissures leading to the detachment of tissue from the labias.

First-line treatment of VCD is medical. Topical corticosteroids are the initial treatment modality for the majority of cases. They can be used alone or combined with oral antibiotics.

Prolonged use of antibiotics such as metronidazole or immunosuppressive drugs such as corticosteroids, azathioprine, and infliximab have been reported in the literature.[4,7] Ciprofloxacin has also been reported as a treatment alternative for VCD and may provide transient relief.[8] However, oral moxifloxacin use has not been previously mentioned. Apart from bactericidal effects, the anti-inflammatory property



Figure 2: Postoperative immediate (left) and 1 week (right) view after partial vulvectomy. Areas that displayed tension were left to heal secondarily with daily wound dressings. The patient displayed rapid healing under oral moxifloxacin treatment

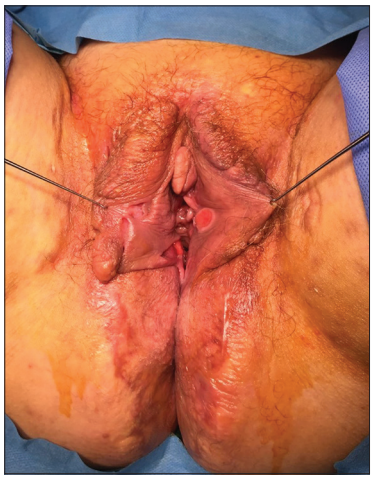


Figure 3: Presentation 1 year after surgery without routine postoperative follow-up, showing multiple fistulas, recurrence of linear fissures and papillomas, along with an ulcer at the vaginal opening. The internal gynecological examination was unremarkable at the same time frame



Figure 4: Postoperative appearance of the patient at 3 weeks, displaying a general resolution of edema, pain, and intertriginous fissures

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of moxifloxacin can explain our patient's good response to treatment.[9] Our patient did not benefit from metronidazole, prednisolone, and adalimumab. Therefore surgery was carried out to provide relief to the patient's symptoms and psychological discomfort. After partial vulvectomy, the addition of moxifloxacin resulted in dramatic improvement. Ulcers not addressed during the first surgery all healed spontaneously. Unfortunately, the patient was unable to comply with postoperative follow-ups after the first line of surgery and treatment due to her overseas residency and therefore had recurrence, which was left untreated until her visit in the 1st year. Although surgery can provide benefits for VCD, it is crucial that patients are followed up meticulously, as the surgery itself can lead to additional deformation if not healed properly. This was the reason for limiting the amount of surgical resection at the second operation, as a similar postoperative follow-up was expected. It is important to sustain regular postoperative follow-up visits. Secondary infections due to colonizations in the area can disrupt the treatment process. Therefore, frequent wound cultures and appropriate antibiotic treatment, as well as meticulous wound care, are essential. Once the patient is confirmed to have VCD, ensuring close contact with a gastroenterologist is also important as VCD might be the first manifestations of the underlying disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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