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CASE REPORT

Chronic Vulval Ulcer as Crohn's Disease

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Introduction

Crohn's disease is chronic relapsing, remitting granulomatous bowel inflammation affecting any part of the gastrointestinal tract, from mouth to anus, with characteristic skin lesions. The etiology is a composite of several environmental, genetic, immune response-related, and bacterial factors; however, it is not an autoimmune disease. The onset of the disease is acute and is frequently seen among adolescents or young adults [1]. The disease is highly prevalent in females, especially affecting women in their peak reproductive ages. This disease is associated with a heightened long-term risk of colon and small bowel carcinoma and leads to a long delay in diagnosis once the disease sets in [2].

This disease usually includes gastrointestinal symptoms such as recurrent aphthoid ulcer, abdominal pain, and bloody or watery diarrhea, along with the presence of systemic symptoms like fever and weight loss [2]. The development of extra-intestinal manifestations affecting the joints, eyes, and skin is a significant feature reported in almost one-third (18–44%) of all patients [3]. The clinically observed extra-intestinal manifestations include arthritis, scleritis, episcleritis, and severe ulcerative skin lesions such as pyoderma gangrenosum or erythema nodosum [3]. Anemia is also a major extra-intestinal complication commonly experienced in patients with inflammatory bowel disease [4].

Vulval involvement in Crohn's disease is an extremely rare condition, wherein the vulvo-vaginal, perianal, perineal, or urological symptoms often mislead in the appropriate diagnosis. Vulval involvement of this disease can either be contiguous or non-contiguous with or without the presence of gastrointestinal symptoms [3]. Genital ulcers may affect the patient psychologically, further delaying the diagnosis among females, especially unmarried girls who resist revealing their condition. Diagnostic approaches include colonoscopy, in which the colon's cobblestone appearance indicates the disease, while magnetic resonance imaging helps rule out complications such as fistula. The primary management of vulvo-vaginal disease consists of combined pharmacological therapy of antibiotics, anti-inflammatory agents, corticosteroids, and immunosuppressants in conjunction with non-pharmacological and surgical interventions [3]. Here is a case report of a 17-year old unmarried girl diagnosed with Crohn's disease presented with chronic vulval ulcer, the diagnostic approach, and her treatment response are being reported and reviewed.

Case Report

A 17-year-old unmarried girl presented to the gynecological OPD with an acute flare-up of chronic vulval ulcer along with complaints of perineal pain and ambulatory difficulty. The patient had a 4-year history of painful pustule of spontaneous onset in the left labia majora region, which progressed as swelling and turned into an ulcer, and for which she was symptomatically treated elsewhere. In the light of persistent vulval ulcer, a biopsy was taken, followed by wound debridement and suturing, but later it turned out to be a chronic non-healing ulcer. The patient reported recurrent aphthous ulcer and intermittent spasmodic abdominal pain on and off since consultation. The menstrual cycle was regular throughout the time period. The patient had no weight gain, discharge or bleeding from ulcer or altered bowel habits. There was no history of contact with tuberculosis patients or significant

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drug history. The patient was psychologically affected due to the present condition.

On general examination, she was malnourished and anemic with no significant lymphadenopathy or ulcers in other body parts. Local examination revealed normal mons pubis, right labia majora, bilateral labia minora and perirectal area, but the presence of an ulcer of size 5 × 3 cm in the inferior end of left labia majora. There was no discharge or bleeding from the vulval ulcer site with no signs of healing (granulation tissue). No fistulous tract was seen inside or surrounding the ulcer.

Investigations showed leukocytosis and negative for tuberculosis, HIV and VDRL. MRI pelvis with fistulogram revealed residual collection in left labia and subcutaneous collection in gluteal cleft and enlarged unilateral inguinal lymph nodes. The patient was initiated antibiotic therapy. Considering the persistent abdominal pain, further ultrasound evaluation of the abdomen showed short segment bowel wall thickening in the right iliac fossa with adjacent omental thickening and mesenteric lymphadenitis, and the contrast-enhanced CT abdomen also confirmed the above findings. Gastroenterologist's opinion was obtained and the patient was advised colonoscopy. The results revealed multiple large transverse ulcers in the terminal ileum, caecum, ascending colon, transverse colon and rectum with intervening normal mucosa. Multiple biopsies were taken from ulcer edges and sent for gene expert and histopathology. Colon biopsy was indicative of moderate chronic active ileitis with ulceration and mild eosinophilia in the colon, along with edema and crypt loss in small bowel biopsy. The histopathology report confirmed a necrotizing lesion consistent with necrotizing fasciitis in the vulval ulcer, while the gene expert reported negative.

In view of the clinical findings and laboratory evidence, she was finally diagnosed with vulval Crohn's disease after ruling out vasculitis and autoimmune etiologies. She was started on mesalamine (5 amino-salicylic acids), prednisolone, and azathioprine and advised on restrictions in diet and lifestyle. The patient responded well to the therapy, and the vulval ulcer healed completely in 3 weeks, with remission of disease after a 1-month follow-up. The prednisolone dose was gradually tapered and stopped after 3 months. Mesalamine and azathioprine were continued along with strict adherence to changes in lifestyle (Fig. 1).

Discussion

Crohn's disease is a chronic granulomatous gastrointestinal disorder of unknown etiopathogenesis. The Ileocecal junction is the commonly affected site of the disease, although it involves the entire alimentary canal. In addition, it has extra-intestinal cutaneous manifestations such as perianal and peristomal lesions, pyoderma gangrenosum, Sweet's syndrome, acrodermatitis enteropathica, epidermolysis, bullosa acquisita and granulomatous cutaneous lesions are relatively infrequent, affecting only 22–44% of the cases [1, 2]. Vulvar Crohn's disease as a clinical manifestation is an extremely rare condition, and only 124 cases have been reported in literature worldwide. Vulval Crohn's disease is presented in two forms on account of its contiguity, as a direct extension of the gastrointestinal involvement, such as fistulas or fissures; the other form is non-contiguous that has no connection between the vulva and the bowel, which is also known as metastatic Crohn's disease [3].

Multiple differential diagnoses including genital tuberculosis, sarcoidosis, hidradenitis suppurativa, lymphogranuloma venereum, syphilitic lesion, fungal, bacterial or parasitic infectious vulvovaginitis, donovanosis, herpetic vulvovaginitis, Behcet's disease, pyoderma gangrenosum, condyloma, foreign body reaction and vulval neoplasia must be taken into consideration before the final diagnosis is made. Histopathology report revealing inflammatory cells with non-caseating epithelioid granulomas remains the cornerstone diagnostic criteria to make a definitive diagnosis [2].

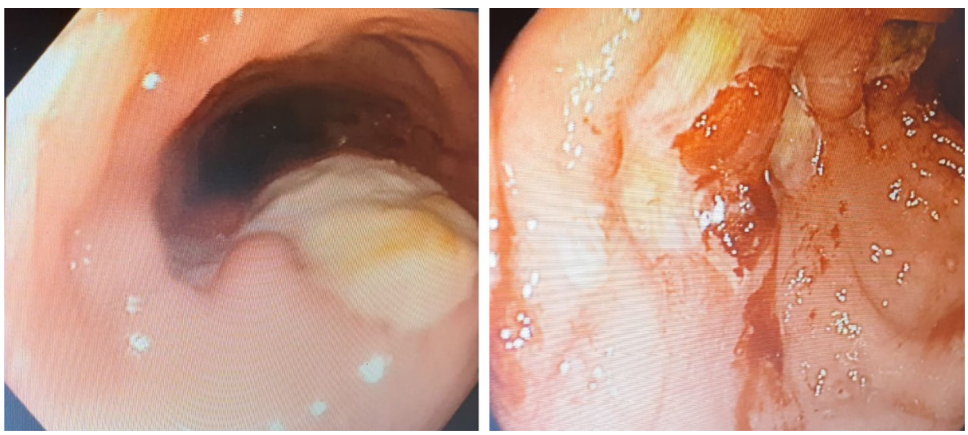

Fig. 1 Colonoscopic view

In our case, the patient had symptoms of a persistent non-healing vulval ulcer perineal pain but no gastrointestinal symptoms except for on-and-off abdominal pain. The conventional pharmacological approach consisting of combined therapy of mesalamine, corticosteroids and azathioprine was found to be successful in healing vulval ulcers relatively faster despite a lack of gastrointestinal manifestations. Kim et al. [2] reported a case of vulval ulcer relapse in an adolescent female taking medications for Crohn's disease for 10 weeks. The delay in diagnosing disease of the vulva resulted in the recurrence even after a surgical procedure, and resolution of symptoms was finally achieved after re-operation [3].

Bondarenko et al. [4], reported the case of a 47-year-old female having recurrent severe vulvar edema with severe symptoms. The gynecological assessment revealed bilateral asymmetric hypertrophy, and prominent edema of the labia majora, predominantly on the right side. Fissures with clear liquid discharge. The diagnosis followed by antimicrobial, anti-inflammatory, immunomodulating, and other agents were reported [4].

Vulval Crohn's disease is an uncommon entity, and differential diagnosis of vulvar lesions is based on infectious diseases such as tuberculosis, lymphogranuloma venereum, syphilitic chancre, and herpetic lesions. Before initiation of therapy, non-infectious causes fistulas and abscesses should be ruled out by proctoscopy, sigmoidoscopy, colonoscopy, and imaging techniques like USG/CT/MRI [3]. Initial pharmacological management includes systemic sulfasalazine or mesalamine, corticosteroids such as prednisolone, and immunosuppressants like azathioprine. Metronidazole monotherapy or in combination with steroids are effectively used to manage vulval Crohn's disease with a high success rate. Anti-TNF agents and surgical procedures such as vulvectomy (partial/total), laser vaporization, and lesion excision are also common in clinical practice [2].

Conclusion

Vulval ulcer as a primary presentation of without fistulous tract in an unmarried girl at adolescent age is a very rare condition. The case reports that the patient had a quick and effective remission of the disease when treated with mesalamine and azathioprine and lifestyle modifications. Further research is warranted for assessing the effectiveness of first-line agents in vulval Crohn's disease and long-term follow-up to analyze the risk for any relapse in therapy.

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Declarations

Conflict of interest Nil.

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Informed Consent Obtained.

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