International Journal of Research in Dermatology
Soukaina S et al. Int J Res Dermatol. 2023 May;9(3):130-132
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20231167

Case Report

A case of clitoral Crohn's disease

Sektaoui Soukaina*, Mehsas Zoubida, Benzekri Laila, Senouci Karima

Department of Dermatology and Venereology, Ibn Sina University Hospital, Faculty of medicine and pharmacy of rabat, Mohamed V University, Rabat, Morocco

Received: 24 February 2023
Accepted: 31 March 2023

***Correspondence:**
Dr. Sektaoui Soukaina,
E-mail: Soukaina.sektaoui@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Crohn's disease (CD) is an inflammatory bowel disease (IBD) that can present with cutaneous lesions, including metastatic lesions. Vulvar CD is a rare manifestation of the disease, with only a few reported cases in the literature. Here we present the case of a 45-year-old woman with metastatic vulvar CD who presented with a one-year history of painful genital swelling. Diagnosis was confirmed through histopathology of the genital lesion. Diagnosis of vulvar CD can be challenging, as the clinical presentation is highly variable. Clinicians should consider the possibility of vulvar CD in patients presenting with aphthous ulcerations, vulvar edema, lymphedema, or lymphangiectasia. Medical treatment options for vulvar CD include corticosteroids, azathioprine, systemic sulfasalazine, cyclosporine, ciprofloxacin, and long-term metronidazole, while surgical interventions include partial or total vulvectomy, laser vaporization, or excision of the lesion. Diagnosis of CD is important, and various diagnostic tools should be used to search for digestive CD. The evolution of vulvar CD can be unpredictable, and surgical intervention may be required when medical treatment fails.

Keywords: CD, IBD, Metastatic vulvar CD, Cutaneous lesions, Granulomatous lesions, Vulvar edema,

INTRODUCTION

Crohn's disease (CD) is an IBD that affects the digestive tract and is often complicated by cutaneous lesions that can be contiguous, metastatic, or associated with inflammation.[1] Metastatic CD is a particular manifestation of the disease that involves the presence of granulomatous lesions in locations outside of the gastrointestinal tract.[2] One such location where CD-related lesions can occur is the vulva, with approximately 2% of women with CD experiencing vulvar lesions.[3] The most common presentation of vulvar lesions in CD is asymmetrical labial swelling and edema.[4] Here we present the case of a 45-years old women with a metastatic vulvar CD.

CASE REPORT

A 45-years old women, with no medical history, presented to the emergency department with a one-year history of painful genital swelling. Cutaneous examination of genital area showed a swelling and edema involving labia majora that almost occluded the view of labia minora with a thickened and hypertrophied clitoris (Figure 1).

Per speculum examination was normal. There was no lymphadenopathy or swelling of limbs and routine laboratory test blood showed a microcytic hypochromic anemia (Hb 9.9 g/dl, MCV 72 fL, MCHC 28 g/dl, Ferritin 12 ng/ml). Ultrasonography was done to exclude an abscess, bacterial cultures and testing for sexually

transmitted infections were negative. Histopathology of the genital lesion (Clitoris) showed a dermis lined on the surface by a Malpighian epithelium hyperplasia overworked by orthokeratotic hyperkeratosis with the presence of a granular layer. The dermis is loose fibrous, widely traversed by thin-walled capillaries with a turgid endothelium. In depth, an inflammatory granulation tissue can be seen, containing numerous non-caseating epitheloid and gigantocellular follicles and surrounded by lymphocytes and plasma cells (Figure 2).

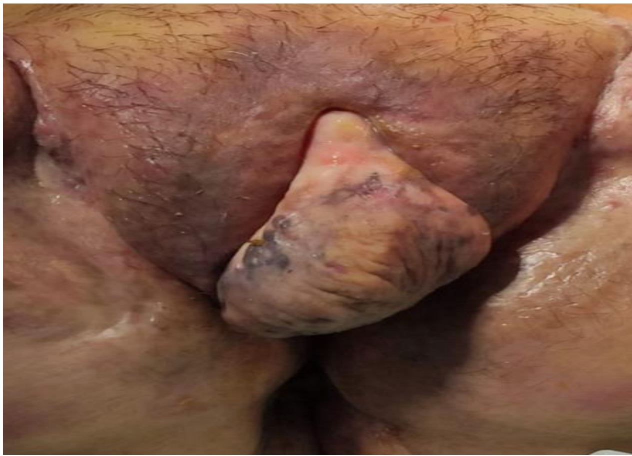

Figure 1: Clinical image showing a swelling and edema involving labia majora that almost occluded the view of labia minora with a thickened and hypertrophied clitoris.

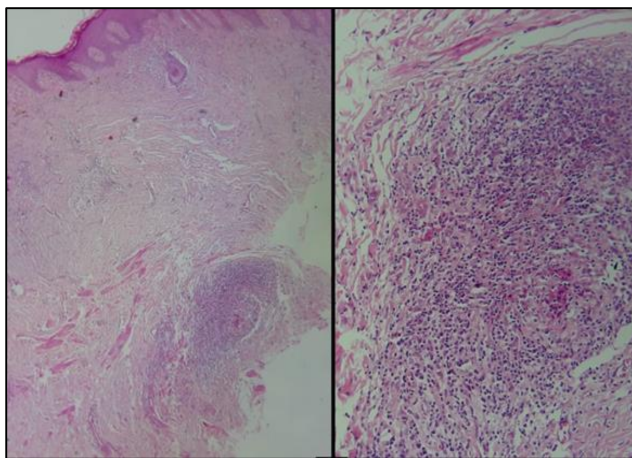

Figure 2: Histopathological image confirming the diagnosis of metastatic vulvar (Clitoris) CD.

A final diagnosis of metastatic vulvar (Clitoris) CD was done and the patient was referred to a gastroenterologist and gynecologist for treatment.

DISCUSSION

CD is a chronic, relapsing IBD that affects the digestive tract, from the oral cavity to the anus. Along with gastrointestinal symptoms, CD can also be associated with inflammatory cutaneous lesions that can be classified into three categories: (A) peri-anal and peri-stomal lesions; (B) specific cutaneous manifestations, including erythema nodosum, pyoderma gangrenosum, Sweet's syndrome, acrodermatitis enteropathica, and epidermolysis bullosa acquisita; and (C) metastatic lesions.[1,5,6] Metastatic vulvar CD is an extremely rare manifestation, with only a few reported cases in the literature.[1,7,8] It has been observed to precede the diagnosis of digestive CD in approximately 25% of cases, and it is more common in women, accounting for around 50% of cases.[9] The clinical presentation of vulvar CD is highly variable, and diagnosis can be challenging.[10] Clinicians should consider the possibility of vulvar CD in patients presenting with aphthous ulcerations, vulvar edema, lymphedema or lymphangiectasia, knife-cut ulcers, recurrent aphthous ulcerations, perianal tags or fistulae, and suppurative nodules.[1] Metastatic vulvar CD is a rare manifestation, and its clinical lesions are nonspecific, leading to a diagnostic dilemma for clinicians. In such cases, clinical-histopathological correlation is essential for accurate diagnosis.[1] When CD is suspected clinically or histopathologically, it is important to search for digestive Crohn's using various diagnostic tools, such as proctoscopy, sigmoidoscopy, colonoscopy, and upper gastrointestinal endoscopy, along with imaging studies such as ultrasonography, computed tomography (CT) scan, and magnetic resonance imaging (MRI) of the abdomen and pelvis.[6] In addition, patients with vulvar CD can develop unrecognized streptococcal cellulitis, so bacterial cultures should be obtained when fluctuant lesions do not improve. Ultrasonography or magnetic resonance enterography may also be useful for detecting abscesses or fistulae.[11] The evolution of vulvar CD can be unpredictable, and surgical intervention may be required when medical treatment fails, although some cases of spontaneous healing have been reported in the literature.[12] Medical treatment options for vulvar CD include corticosteroids, azathioprine, systemic sulfasalazine, cyclosporine, ciprofloxacin, and long-term metronidazole.[2-6] Corticosteroids are considered a mainstay of CD treatment, while ciprofloxacin and metronidazole can be used in combination to treat active inflammatory, fistulizing, and perianal CD.[2,5] Sulfasalazine is more effective in treating bowel disease rather than perianal disease.[4] Surgical interventions for vulvar CD include partial or total vulvectomy, laser vaporization, or excision of the lesion.[8,12,13] Surgery may be necessary when lesions are refractory to medical treatment and have caused significant aesthetic disfigurement.

In conclusion, metastatic vulvar CD is a rare manifestation that can present with a variety of clinical symptoms, including vulvar edema and swelling. Diagnosis can be challenging, but histopathological examination is essential for accurate diagnosis. The treatment needs the involvement of gynecologists, gastroenterologists, and dermatologists to achieve disease control, ensure optimization of both intestinal and

cutaneous disease and improve patient quality of life. Medical treatment options include corticosteroids, azathioprine, systemic sulfasalazine, cyclosporine, ciprofloxacin, and long-term metronidazole, while surgical interventions may be necessary when medical treatment fails. Clinicians should consider the possibility of vulvar CD in patients presenting with vulvar lesions, and diagnostic tools should be used to search for digestive CD.

ACKNOWLEDGEMENTS

Author would like to thanks to all the individuals who contributed to this work. First and foremost, we are grateful to the patient for her cooperation and willingness to share her medical history. We would also like to thank the staff at the department of dermatology and venereology at Ibn Sina university hospital, faculty of medicine and pharmacy of Rabat, Mohamed V university for their support and assistance in carrying out this research.

Finally, we would like to thank all of our colleagues and peers who provided feedback and support throughout the course of this research. Their insights and suggestions have been instrumental in shaping our work and improving its quality.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Shields BE, Richardson C, Arkin L, Kornik R. Vulvar Crohn disease: Diagnostic challenges and approach to therapy. Int J Womens Dermatol. 2020;6(5):390-4.
2. Makhija S, Trotter M, Wagner E, Coderre S, Panaccione R. Refractory Crohn'sdisease of the vulva treated with infliximab: A case report. Can J Gastroenterol. 2007;21:835-7.
3. Feller ER, Ribaubo S, Jackson ND. Gynecolgic aspects of Crohn'sdisease. Am Fam Physician. 2001;64:1725-8.
4. Amankwah Y, Haefner H. Vulvaredema. Dermatol Clin. 2010;28:775-7.
5. Friedman S, Blumberg RS. Inflammatory bowel disease. In: Longo DL, editor. Harrison's Principle of Internal Medicine. 18th ed. New York: McGraw-Hill. 2012;2477-95.
6. Barret M, deParades V, Battistella M, Sokol H, Lemarchand N, Marteau P. Crohn'sdisease of the vulva. J Crohnscolitis. 2014;8:563-7.
7. Andreani SM, Ratnasingham K, Dang HH, Gravante G, Giordano P. Crohn'sdisease of the vulva. Int J Surg. 2010;8:2-5.
8. Guerrieri C, Ohlsson E, Ryden G, Westermark P. Vulv it is granulomatosa: Acryptogenic chronic inflammatory hypertrophy of vulvar labia related to cheilitis granulomatosa and Crohn's disease. Int J Gynecol Pathol. 1995;14:352-9.
9. Loftus EV Jr. Clinical epidemiology of inflammatory bowel disease: Incidence, prevalence, and environmental influences. Gastroenterology. 2004;126:1504-17.
10. Moreno AC, Goje O, Piliang MP, Batur P. Vulvar and gluteal manifestations of Crohn disease. Cleve Clin J Med. 2019;86(10):645-6.
11. Shen RN, Cybulska BA, Thin RN, McKee PH. Vulval Crohn'sdisease mimicking genital herpes. Int J STD AIDS. 1993;4:54-6.
12. Reyman L, Milano A, Demopoulos R, Mayron J, Schuster S. Metastatic vulvarul creation in Crohn'sdisease. Am J Gastroenterol. 1986;81:46-9.
13. Dap RF, vander Meijden WI. Vulvar lymphangiectasias in Crohn'sdisease. Ned Tijdschr Geneeskd. 2000;144:1692-5.

Cite this article as: Soukaina S, Zoubida M, Laila B, Karima S. A case of clitoral Crohn's disease. Int J Res Dermatol 2023;9:130-2.