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Vulvar Presentation of Crohn's Disease

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Abstract: *Crohn's disease is an inflammatory bowel disorder with several well-known extraintestinal manifestations such as erythema nodosum, uveitis, and arthritis and rarely observed is vulvar manifestation. A case of 24 years old female G1P1 with regular periods with vulvo-anal involvement without bowel manifestation was diagnosed as a case of vulvar presentation of Crohn's disease and was put on multiple drug treatment metronidazole & prednisolone. Patient was reviewed after 3 weeks with no improvement but with flare up of disease. Patient was admitted and biopsy was taken at multiple sites from vulvo-anal region to confirm diagnosis. Histopathology report shows chronic granulomatous inflammatory disease. Azathioprine was added to prior treatment and responded.*

Keywords: *vulva, Crohn's disease, azathioprine, cutaneous*

I. Case Report

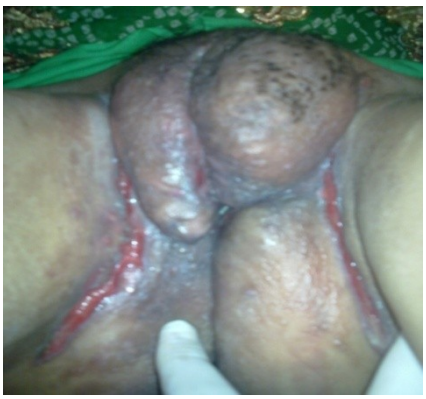
A 24 years old female G1P1 presented to gynecology OPD in Mahatma Gandhi Hospital Jaipur, with complain of pain and swelling of vulva 3 years back. She started with vulvar edema in the late 2nd trimester of her pregnancy which gradually increased in the size till term and she was delivered by LSCS. After delivery there was some regression of swelling and patient disappeared. Six months after delivery she noticed thickening & hyperpigmentation of vulvar skin for which she attended surgical OPD & some surgical procedure was done, but no reports available of the surgery .Two and half yrs later she noticed vesicular rash in perianal region, purulent vaginal discharge, besides pain and further increase in labial swelling.

On examination vitals were stable, chest, cardiovascular system and abdominal examination were clinically normal. Inspection of the vulva and perineum revealed huge bilateral labial swelling, with thickened hyperpigmented skin and multiple areas of induration both over vulva and perianal region involving groin and gluteal folds with knife cut ulceration over groin.

All routine investigation were advised and the result of our initial laboratory investigations were normal, including a complete blood count (hemoglobin: 10.4gm/dl; platelet: 257*10^9/L; white cell count: 8.1*10^9/L) ; ESR:10 mm/hr, HIV and VDRL was negative. An upper gastrointestinal small bowel follow-through was unremarkable. Montoux test and chest radiograph were negative. An ultrasound of the region revealed diffusely enlarged and echogenic soft tissue of the labia, clitoris and surrounding perineum, with possible increased regional blood flow.

Patient was put on oral metronidazole, prednisolone and topical antibiotics. Patient reported back after 3 weeks without any improvement in signs and symptoms. Patient was admitted for biopsy after consultation with dermatologist, biopsy was taken from multiple sites and dose of metronidazole and prednisolone was increased. Histopathology report revealed chronic nonnecrotizing granulomatous inflammatory disease which raised suspicion of vulvo-anal Crohn's disease.

In addition to topical and oral metronidazole and oral prednisolone she was put on Azathioprine. Patient responded to the treatment with regression of her signs and symptoms. After 2 months her groin and perianal ulcers completely healed and patient is symptoms free except the huge, ugly vulva with thickened and hyperpigmented skin for which vulvectomy is advised.



II. Discussion

Crohn's disease is an autoimmune disorder with hereditary component more prevalent among adolescent and young adults between ages 15 and 35 years and symptoms predominantly relate to the diseased a gastrointestinal system and include abdominal pain, diarhoea and weight loss. Associated mucosal and skin finding are less common with Crohn's disease, affecting 20%-40% of patients and may precede the onset of gastrointestinal symptoms. There is no consistent correlation between the appearance of cutaneous lesion and intestinal disease activity.

Crohn's disease is a chronic granulomatous, inflammatory disorder of unknown pathogenesis. Although it can involve any section of the bowel, the ileo-caecal junction is most commonly involved. Extra-intestinal cutaneous manifestations of Crohn's disease, like erythema nodosum and pyoderma gangrenosum, are well known. [2] Ulcerative vulvar Crohn's disease was first described by Park *et al*. [3]It is very rare, and less than 60 such cases have been described. [4] Although the average age of presentation is 34 years, there have been reports in children as young as 8 years of age. [5] Patients may initially present with swelling, erythema, pruritus or pain, and subsequently develop unilateral vulvar hypertrophy, vulvar mass, vulvar edema, draining sinuses, ulceration, or abscess formation. "Knife-cut" ulcers which resemble lacerations are almost pathognomonic of Crohn's disease although they have been reported in herpetic infections in the immunocompromised and in cutaneous tuberculosis. Vulvar involvement in Crohn's disease may be by virtue of contiguity, as a direct extension of intestinal involvement, or non-contiguous (metastatic) in which there is no connection between the vulva and the bowel. [6] In a review by Andreani *et al*., 91% of cases of vulvar Crohn's disease had metastatic spread, while only 5% had contiguous spread. It is in these cases that making a correct diagnosis becomes difficult. Werlin*et al*., have reported that vulvar ulcers may precede intestinal manifestations by up to 18 years. [7] Initial stages of vulvar Crohn's disease can be medically managed. Metronidazole alone or in combination with steroids has been the most effective treatment with a success rate of 87.5%. [4] The optimal recommended dose of metronidazole is 20 mg/kg/day for at least 12 to 36 months. [8] Bilateral pedal paresthesia is a complication reported with long-term metronidazole. Other drugs like sulfasalazine, azathioprine, infliximab, and thalidomide have been used with varied response. Advanced cases may require vulvectomy, but local excision has been reported to show recurrence of the disease.

III. Conclusion

Crohn's disease may often be overlooked as a cause of cutaneous vulvar symptoms. Awareness that such involvement may precede gastrointestinal symptoms is important, because it occurs infrequently, and makes diagnosis challenging, because genital manifestation of Crohn's disease may be erroneously attributed to gynecological disorder.

References

[1]. Kingsland CR, Alderman B. Crohn's disease of the vulva. J R Soc Med 1991;84:236-7.
[2]. Leu S, Sun PK, Collyer J, Smidt A, Stika CS, Schlosser B, et al. Clinical Spectrum of Vulva Metastatic Crohn's Disease. Dig Dis Sci 2009;54:1565-71
[3]. Parks AG, Morson BC, Pegum JS. Crohn's disease with cutaneous involvement. Proc R Soc Med 1965;58:241-2.
[4]. Andreani SM, Ratnasingham K, Dang HH, Gravante G, Giordano P. Crohn's disease of the vulva. Int J Surg 2010;8:2-5.
[5]. Lally MR, Orenstein SR, Cohen BA. Crohn's disease of the Vulva in an 8-year-old girl. Pediatr Dermatol 1988;5:103-6.
[6]. Bohl TG. Vulvar ulcers and erosions-a dermatologist's viewpoint. Dermatol Ther 2004;17:55-67.
[7]. Werlin SL, Easterly NB, Oechler H. Crohn's disease presenting as unilateral labial hypertrophy. J Am Acad Dermatol 1992;27:893-5
[8]. Brandt LJ, Bernstein LH, Boley SJ, Frank MS. Metronidazole therapy for perineal Crohn's disease: A follow-up study. Gastroenterology 1982;83:383-7