

Crohn's disease of the vulva in a 10-year-old girl

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Crohn's disease may involve all parts of the gastrointestinal tract and may often involve other organs as well. These non-intestinal affections are termed extraintestinal manifestations. Vulval involvement is an uncommon extraintestinal manifestation of Crohn's disease, and it is very rare in children. Patients with vulval CD typically present with erythema and edema of the labia majora, which progresses to extensive ulcer formation. Vulval Crohn's disease can appear before or after intestinal problems or it may occur simultaneously. We present a 10-year-old girl with intestinal Crohn's disease complicated with perianal skin tags and asymptomatic unilateral labial hypertrophy. The course of her lesion was independent of the intestinal disease and responded significantly to medical treatment including azathioprine and topical steroid.

We emphasize that although vulval involvement in childhood is uncommon, Crohn's disease must be considered in the differential diagnosis of nontender, red, edematous lesions of the genital area.

Key words: Crohn's disease, vulva, children.

The skin is a common site of extraintestinal involvement in Crohn's disease (CD). The most common site of cutaneous involvement in CD is the perineal and perianal areas^{1,2}. Rarely, vulvar and groin areas may be affected². Skin can be affected by this granulomatous disease either through direct extension of the intestinal lesions or with manifestations that are totally separated from the intestine³. The latter kind of lesion is located at a distance from the intestine, surrounded by healthy skin, not connected to the intestine, and also known in the literature as "metastatic CD"¹⁻⁴. We present a rare case of "metastatic" vulval CD in a child.

Case Report

In May 2003, a 10-year-old girl was evaluated for a six-month history of persistent unilateral vulvar erythema and swelling. She had CD of the large bowel for eight months, which was well controlled by mesalamine and corticosteroid. For her vulvar problem, she had received

empiric therapy including oral ampicillin, oral ketoconazole, topical antifungal and local care, but this therapy was unsuccessful. Physical examination showed failure to thrive, paleness, clubbing of fingers, nail dystrophy, left vulvar erythema and edema, and multiple perianal skin tags (resembling condyloma acuminata) (Fig. 1). The vulvar skin was non-ulcerated and



Fig. 1. Patient's vulva pretreatment showing marked reddish-purple colored edema without ulceration.

non-tender, and had softly elastic consistency by palpation. The architecture was normal in the vulvar area, and there was no discharge. She had no inguinal lymphadenopathy, and no pain or any other symptoms. There was no history of trauma or sexual abuse.

Standard hematology and biochemistry were normal. Vaginal cultures were negative for fungus and bacteria. Viral markers, VDRL and rapid plasma reagin tests were negative. Pelvic ultrasonography, vaginoscopy and chest X-ray were normal. Tuberculin skin test was negative. Radiological studies of bowel showed no fistulas. Biopsy of vulva showed perivascular inflammatory infiltrate in the dermis, but no granulomas were identified. Special staining and bacterial cultures were all negative. Specimen of perianal lesions revealed chronic inflammation and numerous non-caseating granulomas. On the basis of the above clinical and laboratory data, these findings were evaluated as cutaneous CD, and the diagnosis of perianal and "metastatic" vulval CD was made. Oral metronidazole was started for vulval CD; however, this therapy was discontinued one month later due to her elevated liver enzymes. Clobetasol propionate was then administered locally. One month later her intestinal CD flared. In addition to the previous therapy, azathioprine (2 mg/kg/day) was started. Three months later, both her vulval and intestinal CD had improved significantly. She has been maintained on azathioprine. On follow-up, although her bowel CD flared twice during the following two years, her vulval CD remained in remission.

Discussion

Vulval CD is extremely rare in children⁵⁻⁹. Patients with vulval CD typically present with erythema and edema of the labia majora, but soon progress to extensive ulcer formation⁶. The labia majora and clitoris are involved in most cases, and there may be gross architectural destruction of the vulval region. In our case, erythema and edema of the labium majus were present, but there was no ulcer or architectural destruction in the vulval region^{5-9,10}. As we were unable to find any fistulization by barium radiographic studies and vaginoscopy, we believe the lesion in the vulva of our patient was a "metastatic" CD. As in our patient,

perianal lesions accompany 90% of cases¹⁰. Vulval CD may develop before or after gastrointestinal tract symptoms or may occur simultaneously⁸.

The recognition of CD of the vulva can be made easily in the presence of intestinal involvement. Histopathologically, vulval CD is characterized by discrete, non-caseating granulomas with Langhans type giant cells. Non-caseating granulomas may not be seen in 10% of cases. Other granulomatous disorders such as cutaneous sarcoidosis, mycobacterial infections, actinomycosis, deep fungal infections, lymphogranuloma venereum, granuloma inguinale, cellulitis, chronic lymphedema, schistosomiasis, hidradenitis suppurativa, and foreign body reactions should be considered in the differential diagnosis of vulval CD^{1,10}. No granulomas were identified in the vulval biopsy of our case, but chronic inflammation and numerous non-caseating granulomas were present in perianal biopsy. In our case, the diagnosis of "metastatic" vulval CD was supported by the gastrointestinal and perianal manifestations, nail dystrophy, chronicity of the lesions, and exclusion of other granulomatous and infectious disease.

The initial management of vulval CD is medical, including metronidazole, topical and intralesional corticosteroids, systemic steroids, sulfasalazine, cytotoxic agents such as azathioprine or 6-mercaptoprine, and cyclosporine⁵⁻⁹. The recovery of skin lesions is variable and does not parallel that of intestinal lesions. In our patient, treatment of vulval CD was disappointing with corticosteroid and mesalamine. Metronidazole had to be stopped because of adverse effect. Azathioprine and topical clobetasol propionate combination significantly improved the vulval lesions in our case. When aggressive medical management fails, radical excision may be considered in patients with severe symptoms and for cosmetic reasons^{3,5-9}.

In conclusion, although vulval CD in childhood is uncommon, CD must be considered in the differential diagnosis of vulval lesions.

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