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Crohn's disease presenting as vulvar edema in a 15-year-old girl

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Abstract

Background: 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 can progresses to extensive ulcer formation. Vulval CD can appear before or after intestinal problems or it may occur simultaneously.

Observation: We present a 15-years-old girl with bilateral labial hypertrophy which revealed a Crohn's disease. The course of her lesion was independent of the intestinal disease and responded significantly to medical treatment including Mesalamine, corticosteroid and local care.

Conclusions: We emphasize that although vulval involvement in childhood is uncommon, Crohn's disease must be considered in the differential diagnosis of non-tender, red, edematous lesions of the genital area. (*J Dermatol Case Rep*. 2014; 8(3): 75-77)

Introduction

The skin is a common site of extraintestinal involvement in Crohn's disease. The most common site of cutaneous involvement in CD is the perineal areas.[1,2] Rarely, vulvar and groin areas may be affected.[2] 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 intestine.[3] 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

A 15-year-old girl with no past medical history presented to a dermatological consultation with an 18 months history of vulvar edema. She had consulted at several gynecologists and received empiric therapy including oral ampicillin, oral ketoconazole and topical antifungal but this therapy was unsuccessful. Physical examination showed failure to thrive, paleness, coated tongue, painful and asymmetric bilateral vulvar edema, deep erosions and multiple perianal skin tags (resembling condyloma acuminata) (Fig. 1). The vulvar skin was non-ulcerated, 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 any other symptoms. There was no history of trauma or sexual abuse. Vaginal cultures were negative for fungus and bacteria. Viral markers, VDRL and HIV were negative. Standard hematology and biochemistry showed an inflammatory syndrome. Biopsy of perianal lesions revealed chronic inflammation and numerous non-caseating granulomas (Fig. 2). Digestive endoscopy and biopsies showed an aspect in favor of crohn's disease. On the basis of the above clinical, laboratory and endoscopic data, these findings were evaluated as cutaneous CD, and the diagnosis of perianal and "metastatic" vulval CD was made. Clobetasol propionate was administered locally for vulval CD. One month later her intestinal CD flared. In addition

to the previous therapy, mesalamine was started. Three months later, both her vulval and intestinal CD had improved significantly. She has been maintained on mesalamine. On follow-up her vulval CD remained in remission.

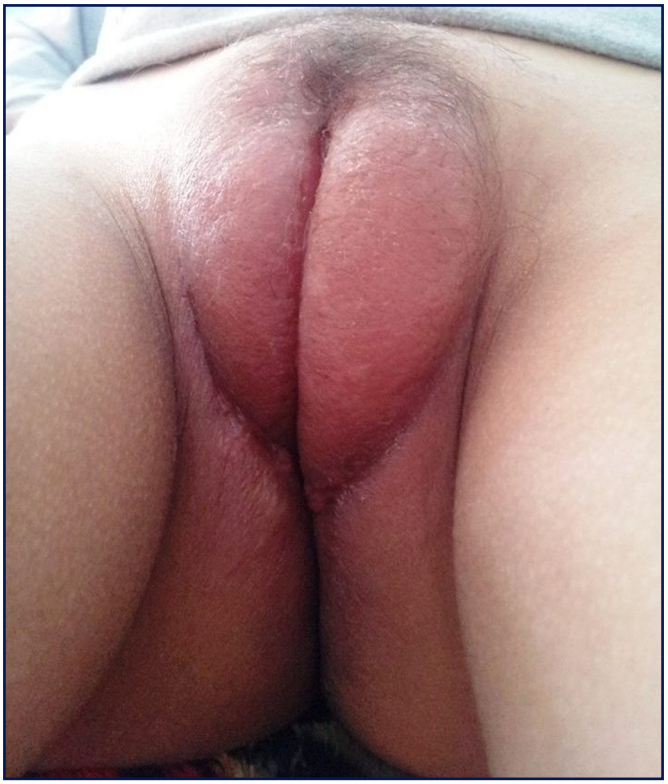

Figure 1
Marked erythema and edema of the labia bilaterally.

Discussion

Vulval CD is extremely rare in children.[5-9] Patients with vulval CD typically present with erythema and edema of the labia majora, but soon progress to extensive ulcer formation.[6] 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] The lesion in the vulva of our patient was a "metastatic" CD. As in our patient, perianal lesions accompany 90% of cases.[10] Vulval CD may develop before or after gastrointestinal tract symptoms or may occur simultaneously.[8] 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] The patient refused vulvar biopsy, but the perianal biopsy showed chronic inflammation and numerous non-caseating granulomas.

In our case, the diagnosis of "metastatic" vulval CD was supported by the gastrointestinal and perianal manifestations, coated tongue, chronicity of the lesions, histological founds and exclusion of other granulomatous and infectious disease.

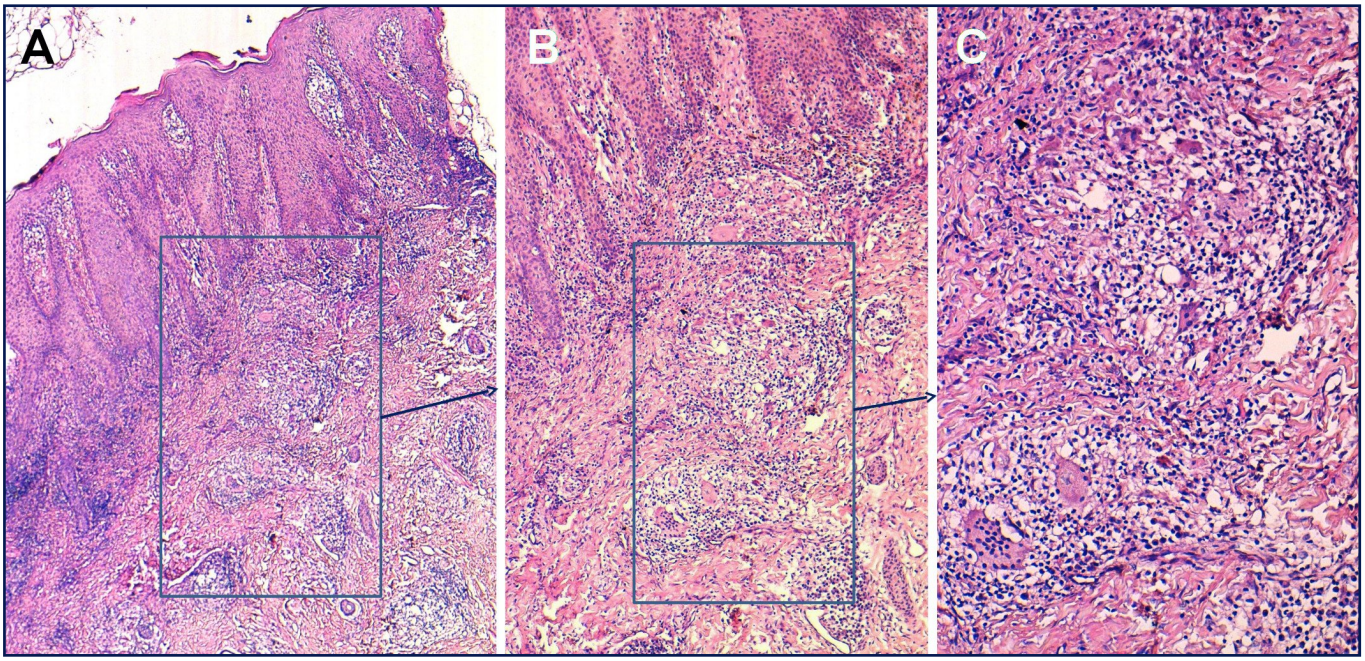

Figure 2
Histopathology of the perianal area demonstrating non-caseating granulomas commonly seen in metastatic Crohn's disease. A. x5, B. x10, C. x20.

In addition to the inherent complications of Crohn's disease that are dominated in children by delayed growth and puberty, the anoperineal manifestations can lead to mutilation real cause of a psychological impact.

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 cyclosporine.[5-9] 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. They 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]

Conclusions

This case illustrates the difficulty in early recognition of CD in children who present with cutaneous vulvar lesions as their initial clinical manifestation. Awareness that such involvement may precede gastrointestinal symptoms is important, because vulvar manifestations of CD may be erroneously attributed to a gynecologic disorder, which highlights the role of endoscopy in helping to establish the diagnosis of CD in the absence of gastrointestinal symptoms. This case also emphasizes the need to be vigilant about metastatic CD symptomatology, and about vulvar symptoms in particular, as a rare but possible first manifestation of CD.

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