Vulval Crohn's Disease in Childhood

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ABSTRACT

Vulval involvement in Crohn's disease (CD) is rare, particularly in children. The clinical features include erythema, edema, ulceration, and labial skin tags. The authors present two cases of children with vulval CD. In both cases, marked labial edema was the presenting feature. In one patient the immunomodulator tacrolimus ointment 0.03% was used with

success. In the second patient control was achieved with intralesional triamcinolone in combination with systemic metronidazole.

Keywords: Crohn's disease; Labial edema; Pediatric Crohn's disease; Pediatric dermatology; Vulval Crohn's disease

INTRODUCTION

Vulval involvement in Crohn's disease is rare, particularly in children. The clinical features include erythema, edema, ulceration, and labial skin tags. Cutaneous manifestations of Crohn's disease (CD) typically present as lesions that are contiguous with the gastrointestinal tract, such as peri-oral and peri-anal lesions [1, 2]. Vulval involvement, mostly due to distant spread of granulomata (metastatic CD), is rare, especially in children [2, 3]. Here, the authors report two cases of vulval lesions of CD in childhood.

METHODS

Informed consent was obtained from all patients for being included in the study.

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CASE SERIES

Patient 1

A 10-year-old girl presented to the dermatology department at York Hospital with a 12-month history of asymptomatic erythema and swelling of the vulva. The patient was systematically well, and had no gastrointestinal symptoms. There was no personal or family history of inflammatory bowel disease. On examination, the patient was a healthy-looking girl with normal height and weight for her age. Examination of the patient's vulva revealed a "peau d'orange" appearance, with a large skin tag in the anterior peri-anal area (Fig. 1). Examination of the oral mucosa, rectum, and the rest of the skin was unremarkable. Biopsy of the affected vulva showed non-caseating granulomata with negative Ziehl–Nielson stains and culture. The patient was treated with tacrolimus ointment 0.03% which resulted in good control of the disease. A year after the initial presentation, the patient developed mouth ulcers and bloody diarrhea. The patient was diagnosed with CD, confirmed on intestinal biopsies.

Patient 2

A 13-year-old girl presented with a 9-month history of asymptomatic vulval swelling. The patient was diagnosed with CD at the age of 5 years and had been well-controlled on methotrexate. Physical examination revealed a healthy-looking girl. Examination of the vulva demonstrated asymmetrical dusky-colored edematous skin with a "peau d'orange" appearance. Examination of the peri-anal skin showed fissures and skin tags (Fig. 2). Examination of the rest of the skin, oral mucosa, and other systems was unremarkable.

Fig. 1 Vulval swelling noted in patient 1

Biopsy of the affected vulval area showed typical features of non-caseating granulomata with negative stains and culture for organisms. The patient was treated with intralesional triamcinolone and oral metronidazole, which resulted in a clear improvement in terms of swelling after 3 months.

DISCUSSION

Metastatic CD in children is rare, and most present prior to the diagnosis of CD [1, 2]. Parks et al. [3] first reported vulval involvement of metastatic CD in 1965. There have only been 16 cases of vulval metastatic CD in children reported in the literature. Vulval CD may present as the first and only manifestation of CD, as seen in patient 1, or after development of

Fig. 2 Unilateral vulval swelling with erythema noted in patient 2

the disease, even if well-controlled otherwise, as in patient 2.

The clinical presentation of vulval CD is variable and may simply manifest as diffuse edema with infiltration or ulceration. The absence of gastrointestinal symptoms often makes diagnosis difficult in children; however, the presence of peri-anal fissures and skin tags should raise suspicion for vulval CD [4].

The differential diagnoses for vulval swelling should include sarcoidosis, infections (e.g. tuberculosis, cellulitis, lymphogranuloma venereum, actinomycosis, pyogenic infections, hidradenitis suppurativa, intertrigo, syphilitic lesions), foreign body reactions, contact dermatitis, acquired lymphangiectasia,

vascular malformations, and sexual abuse [4–6]. Biopsy of the lesion is often necessary to reach a definitive diagnosis, revealing the typical appearance of a non-caseating granulomatous inflammation as seen in CD [4].

The natural course of vulval CD is unpredictable. Some cases resolve spontaneously, while others are resistant to medical treatment [4]. As there is little correlation between the activity and/or severity of skin lesions and bowel disease, treatment of the underlying CD may not be effective against cutaneous CD [2], as was seen in patient 2. Currently, metronidazole alone and/or topical/oral steroids seem to be the most effective treatment for metastatic CD [4]. Other beneficial therapies include dapsone, tetracycline, azathioprine, 6-mercaptopurine, sulphasalazine, and oral zinc supplementation [4]. More recently, anti-tumor necrosis factor therapy has been used with success [5]. Surgical procedures, such as vulvectomy, simple excision, curettage, and debridement, are reserved for resistant cases.

Tacrolimus ointment is currently licensed for use as a second-line agent in the management of atopic eczema. In children, the licensed strength is 0.03%; however, it is increasingly used in other inflammatory conditions, including a pediatric vulval case of pemphigoid [7].

CONCLUSION

In conclusion, vulval swelling can be the first or subsequent manifestation of CD and dermatologists should have a high index of suspicion to facilitate early diagnosis and control of the disease. The use of tacrolimus ointment in vulval CD is novel.

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Conflict of interest. Dr Al-Niaimi and Calum Lyon declare no conflict of interest.

Compliance with ethics guidelines. Informed consent was obtained from all patients for being included in the study.

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