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Vulvar Swelling in a 12-Year-Old Girl: An Early Manifestation of Crohn's Disease

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Abstract

Vulvar Crohn's disease (VCD) is a rare complication of Crohn's disease, especially in pediatric population. An early diagnosis can result difficult if the complication does not present in conjunction with the classic gastrointestinal symptoms that characterize this disease. In this study we present the case of a 12-year-old girl whose initial symptom of Crohn's disease was a symptomatic vulvar swelling promoted by a rectovaginal fistula. We also provide an overview of Crohn's disease and the vulvar changes found in the course of this disease.

Keywords

Recto-Vaginal Fistula, Vulvar Swelling, Metastatic Crohn's Disease

1. Introduction

Crohn's disease is a chronic inflammatory condition of the gastrointestinal tract with relapsing and remitting symptoms [1].

It presents a bimodal peak of onset, the first between the ages of 15 and 30 years, the second at ages 60 to 70 years [2] [3]. It can affect any portion of the gastrointestinal tract, but the terminal ileum represents the preferred location especially in pediatric age [4].

Gynecologic manifestations in young females may include genital manifestations, delayed puberty, and menstrual irregularities [5].

The two forms of presentation of vulvar Crohn's disease (VCD) include lesions contiguous to the gastrointestinal tract, such as fistulas and fissures; the other form includes noncontiguous vulvar lesions, termed metastatic VCD [5].

Vulva involvement from CD is very rare and mostly due to distant spread (metastatic). Diagnosis is difficult due to the rarity of the condition and the great variety of signs and symptoms [6].

Vulva CD typically manifests with swelling, pruritus, erythema and as a progressive and painful ulceration of the labia. These can often evolve into condyloma-like lesions and/or skin tags.

The 75% of these patients have gastrointestinal symptoms of CD [6].

Approximately 10% of children at diagnosis will have fistulizing disease including perianal, rectovestibular, and rectovaginal types. [7]

Unfortunately, because of the rarity of VCD, patients may not be accurately diagnosed or the diagnosis is initially delayed, and the patient may unintentionally receive localized clinical or surgical treatment without success.

Definitive diagnosis can only be achieved by biopsy which would reveal a non-caseating granulomatous lesion[6].

We present a case of VCD with contiguous lesion to the gastrointestinal tract (recto-vaginal fistula), in an adolescent who had no gastrointestinal symptoms.

2. Case Report

Here, we present the case of a 12-year-old girl who came for observation at PO Santobono because of painful vulvar swelling. The patient reported perineal pain and discomfort when sitting. On physical examination, the patient's lips appeared oedematous and erythematous with an irregularly thickened skin surface (**Figure 1**).

Figure 1. Labial hypertrophy with a large area of skin dyschromia in the perineal region.

No other notable signs or symptoms were highlighted by the visit and past medical history did not show anything relevant. No genetic disorder history was found. Clinical findings pointed in the first hypothesis to bartholinitis.

Lab tests showed modest lymphocytosis, a relevant increase of CRP and ESR

as well as a slight reduction in serum iron levels (17 ug/dL). Vaginal secretions were used for cultures which resulted negative.

To better characterize the vulvar lesion, the patient was examined using transperineal ultrasonography (US). The exam showed, in the context of the subcutaneous plane, posteriorly and deeply to the right major labrum, an area of ecostructural inhomogeneity with a central fluid-dense component, suspicious for small abscess collection. At the upper end of this area was evident a hypoechogenic lineal image directed towards the anterior wall of the rectum, likely a fistulous tract (Figure 2).

Performing transperineal US an important fluid flap in the pelvic cavity was noticed so an US of the whole abdomen was performed; it showed a marked thickening with loss of the normal layering involving the wall of the last ileal loop, the cecum and the ascending colon. These regions resulted markedly hyperemic at the Color Doppler investigation (Figure 3(a) & Figure 3(b)).

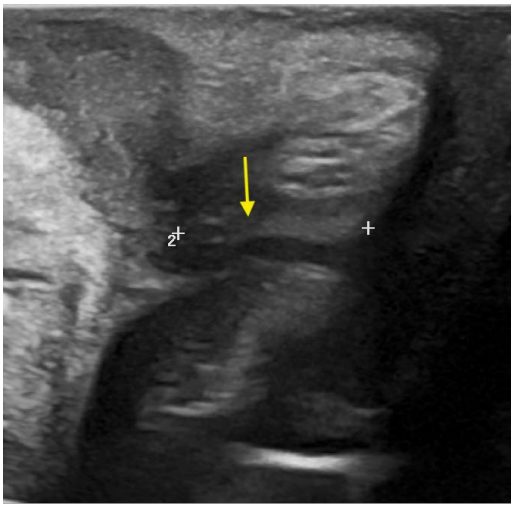
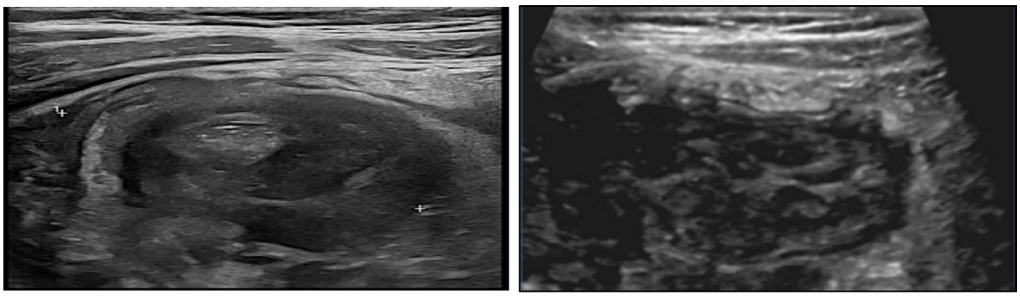


Figure 2. Transperineal ultrasound scan performed with a linear probe showed a small subcutaneous abscess collection, located dorsally and deeply to the right labia majora and a fistulous tract between rectum and vagina (arrow).



(a) (b)

Figure 3. (a) Abdominal US with bowel investigation showed marked wall thickening with loss of the normal layering of right colon; (b) Wall of the last ileal loop.

The patient underwent a magnetic resonance (MRI), to assess and quantify the extent of the changes found on US. The MRI showed overdistension of ce-

cum and ascending colon with marked and irregular wall thickening and extensive loss of haustra; a stenotic area was found near the hepatic flexure, surrounded by thickened and hyperemic perivisceral adipose tissue with multiple lymphadenomegalies (Figure 4).

The exam confirmed the presence of the abscess in the perineum, posterior to the right major labrum, as well as the recto-vaginal fistula visualized by US examination (Figure 5(a) & Figure 5(b)).

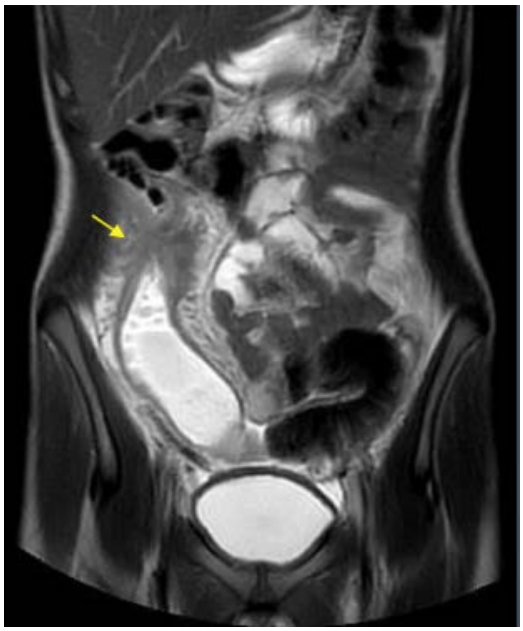
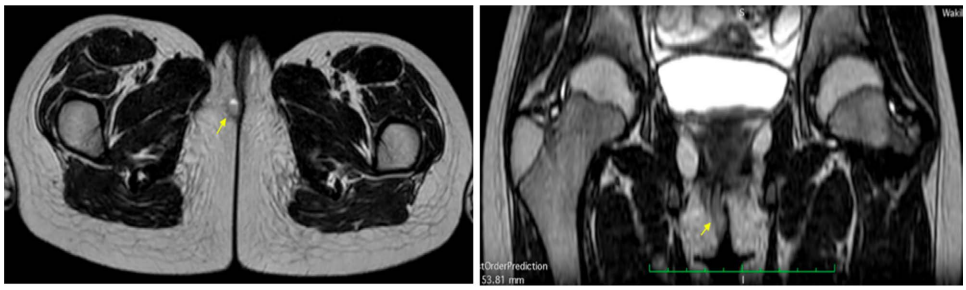


Figure 4. MRI Coronal plane T2W sequence with fat saturation: overdistension of cecum and ascending colon with marked wall thickening and stenotic area near the hepatic flexure (arrow), and extensive loss of haustra.



(a) (b)

Figure 5. MRI Axial (a) and coronal (b) T2w sequence confirmed the presence of perineal abscess posterior to the right major labrum (arrow).

Thickened colic tracts showed a discrete contrast enhacement. Recto-vaginal fistula appeared as a linear image with a low signal in all sequences and a modest contrast uptake. Imaging findings pointed towards an inflammatory bowel disease (IBD) complicated by recto-vaginal fistula. It was surprising, as the patient showed a complete lack of gastrointestinal symptoms.

The patient was then referred to the pediatric Gastroenterology Department.

She underwent a colonoscopy hindered by an intestinal substenosis at the distal third of the ascending colon. This exam documented oedema and hyperemia of the mucosa and a approx. 2 cm long serpiginous ulcer in the ascending colon, nodular hyperplasia of the sigma and hyperemia and aphthoid ulceration of the rectum. The fistula observed by US examination and confirmed by MRI was initially not detected; a later confirmation was obtained by a tampon test. Four colonic mucosa biopsies were taken for surgical pathology analysis, which showed glandular distortion, fibrosis and moderate, chronic-active inflammatory infiltrate (**Figure 6**).

Clinical, biochemical, histological and radiological findings confirmed the diagnosis of IBD, in particular Crohn's disease (CD). Thus, the patient received drainage of the perineal fistula with seton placement.

After diagnosis, patient started therapy with infliximab (5 mg/kg) until complete remission.

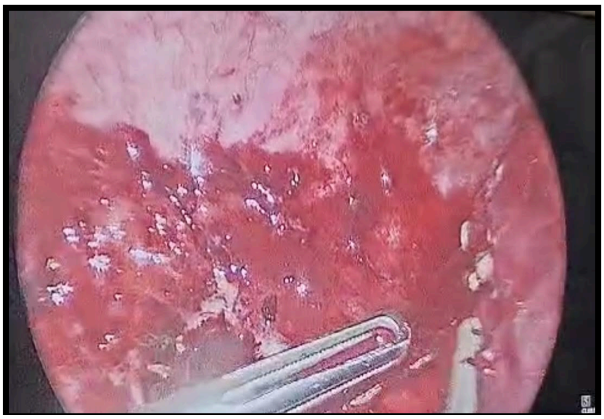


Figure 6. Colonoscopy revealed erythema, edema and pseudopolyps.

3. Discussion

Ours is one of the few pediatric cases reported in literature of vulvar swelling sustained by a recto-vaginal fistula as an early manifestation of Crohn's disease in a patient without gastrointestinal symptoms. This clinical presentation occurs in 9% of female patients with rectal CD [8], almost always accompanied by gastrointestinal symptoms.

Vulvar swelling, such as that observed in our patient, can be secondary to several clinical conditions such as infectious diseases (cellulitis, tuberculosis, fungal infections, lymphogranuloma venereum, tertiary syphilis, actinomycosis) as well as contact dermatitis, sexual abuse, acquired lymphangiectasia, vascular malformations, sarcoidosis or bartolinitis [9], which was our initial suspect.

Vulvar manifestations of Crohn's can be related to two conditions: gastrointestinal complications that may induce the development of recto-vaginal fistulae as in our patient and metastatic cutaneous manifestations which may cause vulvar skin changes independent from the gastrointestinal disease [10].

In children, genitalia represent the most common site for metastatic cutane-

ous disease, one of the rarest complications linked to Crohn's. Metastatic vulvar CD is a rare syndrome characterized by granulomatous genital inflammation with no direct link to fistulous CD [11] [12].

In our case, the fistula was evident on both transperineal US scan and MRI but, it was not immediately evident during colonoscopy, which only documented a few typical features of IBD as serpiginous ulcers and substenosis of the ascending colon. The absence of a frank inflammation in the rectum at the time of colonoscopy was probably due, at least in part, to the remitting nature of this condition [1]. The presence of the fistula was confirmed by tampon test or methylene blue test: a tampon is inserted into the vagina while approx. 20 cc of methylene blue are injected into the rectum with a syringe. 24 hours later, the swab is removed; a test is considered positive if traces of the blue dye are found on the swab.

Although colonoscopy is still the gold standard for the diagnosis of Crohn's disease, imaging makes possible to assess in detail the exact extent of the pathology, to assess the degree of impairment of the loops downstream of a stenotic process and it allows to identify the presence of extraintestinal complications. Ultrasonography is an operator-dependent method, but it is fast and completely free of the risks associated with ionising radiation; it allows the study of intestinal loops and certain findings associated with IBD such as abscesses, fistulas, wall thickening [13]-[15]. It is a very useful method especially in paediatric patients and in our clinical case it was the first useful method to identify the fistulous tract. MRI allows a detailed morphological and quantitative evaluation of the main anatomopathological and clinical findings of intestinal and extraintestinal Crohn's disease [15];

It is able to precisely define the sites and extent of the pathology, its degree of activity and the main complications; As ultrasonography, MRI is completely free of ionising radiation [16]-[18].

4. Conclusions

Our experience, should advice to consider a vulvar presentation of CD in patients with vulvar inflammatory lesions of unknown origin that do not respond to antibiotics or surgical treatment.

Ultrasound and MRI can supplement and, sometimes, replace colonoscopy in the quiescent phase and in the presence of complications (stenosis, subocclusion, fistulae) of Crohn's disease.

Early diagnosis and treatment of recto-vaginal fistulae is crucial, as delayed therapy may lead to permanent vulvar dysmorphism and decreased quality of life.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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