

A Case of Perianal Crohn's Disease with Atypical Digestive Symptoms and Latent Tuberculosis Infection

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1. Abstract

Cutaneous Crohn's disease (CCD) is a rare specific skin manifestation of Crohn's disease (CD) that can affect the skin of the external genitalia, abdomen, and inguinal regions without involving the affected gastrointestinal tract. There have been fewer than 200 reported cases in the literature. In this report, we present a case of a 59-year-old male with perianal skin erythema and ulceration persisting for four months. Skin biopsy revealed non-caseating granulomas with multinucleated giant cells and lymphocytic infiltration. Further investigations, including chest CT and PPD test, indicated latent tuberculosis infection. The initial diagnosis was CCD. After an in-depth medical history inquiry, we discovered that the patient experienced occasional loose stools following the consumption of spicy food. Colorectal endoscopy and MRI were subsequently performed, revealing the presence of CD complicated by an anal fistula. With the definitive diagnosis, the patient was initiated on intravenous infusion of ustekinumab (260 mg), leading to a significant improvement in the ulceration.

2. Introduction

Cutaneous Crohn's disease (CCD) is a specific dermatological manifestation of Crohn's disease (CD). Its characteristic feature is the presence of granulomatous lesions with a cheesy appearance, which can affect the skin of the genitalia, abdomen, and groin area. Unlike the affected gastrointestinal tract, CCD lesions are not continuous. The etiology of CCD remains unclear, but current research suggests that it may be related to immune system dysregulation, alteration in the microbial composition, genetic susceptibil-

ity, and environmental factors [1]. To date, there have been fewer than 200 reported cases of CCD in the medical literature. CCD is more commonly observed in young adults, with rare occurrences in children. The skin lesions can occur before, concurrently with, or after involvement of the gastrointestinal tract [2]. In 70% of cases in adults, CCD symptoms typically appear long after the initial diagnosis of CD and are rarely the first presenting symptom. When the rash presents as perianal ulcers, it can sometimes be misdiagnosed or overlooked, requiring the exclusion of conditions such as ulcerative skin tuberculosis, genital herpes virus, syphilis, perianal Langerhans cell histiocytosis, and Behçet's disease. Here, we report a case of CCD in a Chinese adult male with atypical gastrointestinal symptoms and concomitant latent tuberculosis infection, with written informed consent and institutional approval from Guilin Medical College Affiliated Hospital.

3. Case Report

A 59-year-old Chinese male presented to our hospital with a 4-month history of perianal erythema, ulcers, and pain. The patient reported occasional mild cough and sputum production, with occasional loose stools following the consumption of spicy food. He denied any fever, night sweats, chest pain, abdominal pain, diarrhea, or hematochezia. Physical examination revealed a large area of edematous erythema in the perianal and perineal region, with a central fissure and multiple scattered ulcers. The ulcers appeared shallow, with irregular shapes and varying sizes, the largest measuring 3 cm × 2.5 cm. The ulcer surfaces exhibited yellowish purulent discharge, a moist base, and irregular margins (Figure 1a). The

ulcers were tender but did not exhibit any odor, and there were no vesicles or bullae observed.

Laboratory tests have ruled out the possibility of syphilis, HIV, and herpes. The pathological examination of the anal skin suggests the presence of superficial dermal epithelial-like cells and lymphocytic infiltration, with focal multinucleated giant cell formation, indicative of granulomatous inflammation (Figure 1b). Immunohistochemistry and special staining results were positive for CD68 and negative for Periodic Acid-Schiff (PAS), Gomori Methenamine Silver (GMS), and acid-fast stains. A computed tomography (CT) scan of the chest indicated multiple bronchiectasis and solid nodules in both lungs, with nodules ranging in diameter from 3-6mm, with the largest one measuring approximately 6mm × 5mm in the anterior basal segment of the right lower lobe. Considering the distribution of the nodules in the lungs, which is a common site for tuberculosis, and the atypical clinical presentation of cutaneous tuberculosis, the diagnosis is challenging. Most patients have been vaccinated with Bacillus Calmette-Guérin (BCG), further complicating the diagnosis. To further investigate the presence of tuberculosis infection, additional tests were conducted. The results of the interferon-gamma release assay (IGRA) were positive, and the purified protein derivative (PPD) test indicated an induration size of 13mm×13mm, suggestive of latent tuberculosis infection. Both CD and tuberculosis can affect the gastrointestinal tract, with similar skin manifestations, making the differential diagnosis challenging. However, in this case, the pathological findings revealed non-caseating granulomatous inflammation with negative acid-fast staining, which leans towards a diagnosis of CCD.

The majority of CD patients experience gastrointestinal symptoms as the initial manifestation, such as abdominal pain and diarrhea. Although this patient occasionally has poorly formed stools after consuming spicy food, poorly balanced diet, endocrine and metabolic disorders, intestinal infections, non-infectious diseases, tumors, and neurological dysfunction can also cause poorly formed stools. While there are no typical clinical symptoms, this clue is still worth considering. Colonoscopy was performed to assess the involvement of the digestive system. The colonoscopy revealed multiple irregular shallow ulcers scattered in the cecum and ascending colon, as well as strictures at the ileocecal valve opening (Figure 1c). The perianal skin showed longitudinal shallow ulcers, without evidence of fistulas (Figure 1d). The histopathology of the ascending colon biopsy indicated severe active chronic inflammation with ulceration and crypt abscess formation, accompanied by significant infiltration of lymphocytes, plasma cells, neutrophils, and abundant eosinophils in the mucosal and submucosal layers. Granulation tissue and fibrous tissue proliferation were also observed. Pelvic magnetic resonance imaging (MRI) suggested a fistula-in-ano and perianal abscess formation through the puborectalis muscle. The skin manifestations in this patient may have preceded the gastrointestinal symptoms, and both skin and intestinal biopsies showed non-caseating granulomatous inflammation, consistent with the diagnosis of CD. Considering the involvement of the colon and perianal region in CD, the severity and activity were assessed as active and moderate. After confirming the diagnosis, ustekinumab 260mg, mesalamine, and prophylactic anti-tuberculosis treatment were administered.

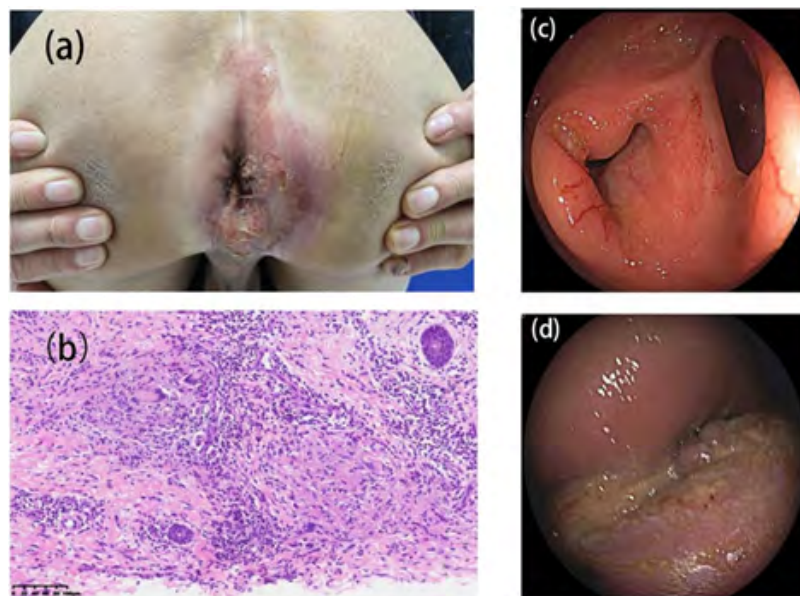


Figure 1: (a) Erythema, fissure and scattered ulcers of the perianal region. (b) The biopsy specimen from perianal skin shows a focal infiltration of epithelioid cells and lymphocytes in the shallow dermis, with the formation of multinucleated giant cells. The finding confirmed granulomatous inflammation. (hematoxylin–eosin stain [HE], original magnification×100). (c) Irregular shallow ulcers scattered in the cecum and ascending colon and internal fistula is likely formed. (d) Shallow longitudinal ulcers were observed in the perianal mucosa.

4. Discussion

CCD is a specific cutaneous manifestation of CD, characterized by granulomatous lesions with a cheesy consistency that can affect the skin of the genitalia, abdomen, and groin area. In 1970, Mountain first introduced the concept of metastatic Crohn's disease [3]. However, in 1999, Anadoh and others believed that the term metastatic Crohn's disease was misleading, and considering the inflammatory nature and diverse clinical manifestations of the disease, renamed it Cutaneous Crohn's Disease [4]. CCD has often been mistaken as an imitator, leading to misdiagnosis and inadequate understanding. CCD can be divided into two main categories: genital and extragenital. It can present as skin erythema, nodules, ulcers, secondary crusts, and scars. On the mucosa of the genitalia, it mainly manifests as ulcers or fissures, and less frequently as isolated lymphedema, or even resembling condyloma acuminatum with papules or patches [5]. Pathologically, CCD is characterized by non-caseating granulomas involving the papillary and reticular dermis, extending into the subcutaneous fat. The granulomas consist of epithelioid cells and multinucleated giant cells, accompanied by significant lymphocytic infiltration, with the presence of plasma cells and eosinophils, while neutrophils and necrotic areas are usually absent [6]. This non-caseating granulomatous lesion is a distinguishing feature from cutaneous tuberculosis.

Currently, there are no uniform diagnostic criteria for CCD, and diagnostic clues come from clinical manifestations, histopathology, and exclusionary tests. Previously reported cases of CCD were mostly found in patients with a history of CD, especially those with colonic involvement. A small percentage of CD patients develop cutaneous lesions several months to years before intestinal involvement [7]. Perianal lesions other than hemorrhoids are one of the warning signs of adult CD, and CCD cases with perianal skin ulcers are mostly reported in individual cases, with a high misdiagnosis rate upon initial diagnosis. Research has found that at least 23% of CD patients develop anal fistulas within the first 20 years after diagnosis, especially in patients with colonic and rectal involvement [8]. In approximately 10% of patients, anal fistulas are the initial manifestation of CD, and their formation may occur several years earlier than the onset of intestinal CD.

Perianal CD presents with various similarities to multiple disease manifestations. In these patients, perianal lesions appear before gastrointestinal symptoms, and the skin lesions, as well as clinical presentations, lack specificity. Dermatologists have limited knowledge of this disease, and if patients seek initial consultation with a dermatologist without focusing on digestive symptoms, it is prone to misdiagnosis or missed diagnosis. A detailed history taking and necessary investigations are essential for an accurate diagnosis.

5. Conclusion

We have reported a case of CCD with concomitant latent tuberculosis infection and atypical gastrointestinal symptoms. A retrospective study of patients with perianal skin ulcers demonstrated the importance of early comprehensive systemic examinations for early diagnosis and treatment of the disease.

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