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Dermatological insight as the key to diagnosing intestinal Behçet's disease misdiagnosed as Crohn's: a case report

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Abstract

This report presents the case of a 22-year-old male with recurrent genital ulcers, oral aphthae, and gastrointestinal ulcerations initially diagnosed as Crohn's disease by gastroenterologists. Despite overlapping clinical and histological features between Crohn's disease and intestinal Behçet's disease, a dermatological consultation raised suspicion for Behçet's, leading to a revised diagnosis. This case highlights the helpful role of dermatologists in avoiding misdiagnosis and ensuring appropriate treatments for complex inflammatory conditions.

Introduction

Behçet's disease is a chronic, multisystemic, autoinflammatory disorder that features recurrent oral and genital ulcers, skin lesions, and potential gastrointestinal involvement.¹ Its clinical presentation can overlap with Crohn's disease, especially when gastrointestinal symptoms dominate, leading to diagnostic delays and suboptimal treatments.² Differentiating these two conditions is crucial, as their therapeutic approaches, while partially overlapping, differ significantly in key areas. For instance, some medications like infliximab and adalimumab are effective in both diseases, but others, such as ustekinumab or vedolizumab, are more specific to Crohn's. This report emphasizes the importance of dermatological assessment in distinguishing Behçet's disease from Crohn's, especially when systemic signs are minimal or absent.³

Case Report

A 22-year-old male presented with a one-year history of chronic abdominal pain, diarrhea, and cyclical rectal bleeding. Given these symptoms, an initial diagnosis of Crohn's disease was made based on gastrointestinal presentation. Colonoscopy revealed deep ulcerations at the ileocecal valve and in the ascending colon, findings commonly associated with Crohn's.¹ Histopathological analysis showed mucosal ulceration, lymphoplasmacytic infiltration, and sporadic neutrophilic granulomas, supporting the diagnosis.²

Magnetic resonance enterography (MRE) revealed inflammatory changes in the terminal ileum, including segmental thickening and ulceration, without fistulas, strictures, or significant mesenteric lymphadenopathy. While these findings were consistent with inflammatory bowel disease (IBD), they were not specific to Crohn's disease.²

The patient later reported a history of recurrent painful ulcers of the glans (Figure 1) and persistent oral aphthae. No systemic signs commonly associated with Crohn's, such as arthritis or uveitis, were present.

However, he mentioned progressive night blindness (hemeralopia), a subtle but recognized manifestation of Behçet's disease.³

Dermatological evaluation raised suspicion for Behçet's disease based on the patient's recurrent genital and oral ulcers. This suspicion prompted a re-evaluation of the diagnosis, leading to a clinical and histological re-evaluation.

Re-examination of biopsy samples revealed deep non-caseating granulomas and neutrophilic infiltration. Vasculitic aspects, including perivascular lymphocytic infiltration and endothelial swelling in the submucosa, were observed.

These findings, combined with the absence of transmural inflammation, supported the diagnosis of Behçet's disease rather than Crohn's.^{4,5}

The combination of recurrent genital and oral ulcers, gastrointestinal ulcerations with vasculitic aspects, and ocular symptoms led to a diagnosis of Behçet's disease.¹ The initial diagnosis of Crohn's was revised following dermatological input, showcasing the need for a multidisciplinary approach in complex cases of IBD.⁵

The initial Crohn's diagnosis led gastroenterologists to propose ustekinumab, a biologic drug more specific for Crohn's disease.⁴ Following the revised diagnosis, the patient was treated with oral corticosteroids and azathioprine, resulting in a marked improvement of gastrointestinal symptoms and a reduction of oral and genital ulcers. Topical corticosteroids were also prescribed for genital ulcers.

Discussion

This case underscores the significant clinical overlap between Behçet's disease and Crohn's disease, especially when gastrointestinal symptoms are prominent.¹ Misdiagnosis can result in inappropriate treatments and delays in proper management.² Clinicians should consider Behçet's disease in patients with recurring oral and genital ulcers, even if systemic signs such as arthritis or uveitis are absent.⁵ Histology plays a pivotal role in distinguishing these conditions.⁴ Vasculitis and perivascular infiltration are more characteristic of Behçet's disease, while Crohn's typically involves transmural inflammation and granulomas without vasculitic features.⁶

Therapeutically, both diseases benefit from anti-TNF agents like infliximab and adalimumab. However, while first-line immunosuppressive therapies such as corticosteroids, azathioprine, and colchicine are effective for Behçet's disease,⁴ biologics like ustekinumab and vedolizumab are primarily indicated for Crohn's.³

Conclusions

This case highlights the importance of considering Behçet's disease in the differential diagnosis of patients presenting with gastrointestinal ulcerations and recurrent mucocutaneous symptoms.⁵ A multidisciplinary approach, including dermatological evaluation, is critical to avoid misdiagnosis and ensure appropriate management.²

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Figure 1. Large painful ulcers of the glans at first evaluation.

