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Behcet's Disease Mimicking Crohn Disease: A Pathological Approach to a Case

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Abstract

Bechet's Disease (BD) is a rare entity. It's a multi-systemic inflammatory disease of unknown etiology characterized by recurrent ulcers and vasculitis, mainly including oral cavity, eyes, gastrointestinal tract and joints. Crohn's Disease (CD) is a chronic inflammatory disorder that may affect the same organs as BD, however more frequently the GI tract. Distinguish BD from CD can be very challenge due to the overlapping clinical presentation and similar morphology features on pathology biopsy specimens. Herein, we present a rare and interesting case which BD mimics IBD on the GI biopsies but with some unique findings. This is a case report of a 32-year-old, female, who first presented at ED with fatigue, weight loss, arthralgia, and erythema nodosum. One month later, patient was admitted for oral ulcers, skin rash, genital ulcers and melena. Treatment with prednisone was started at the time and further workup for BD versus IBD was initiated. Her GI biopsies show granulomatous vasculitis of esophagus and colon biopsies demonstrate overlapping features with CD.

Keywords: Crohn disease • Inflammatory disease • BD mimics • Arthralgia

Introduction

BD is a chronic, multi-system, idiopathic inflammatory disease that can present with a wide range of symptoms [1]. Most of the patients, presents with recurrent oral and genital ulcers as well as skin lesions, thrombophlebitis and uveitis. Gastrointestinal involvement by BD is described to happen in 3% to 25% of the patients, being more common in East Asia and Japan.

In the United States (US), approximately 1/3 of the cases involve the GI tract [2]. Inflammatory Bowel Disease especially CD, usually presents with similar symptoms. However, is far more common in US than in Asia, what makes the differential diagnosis between those two entities challenging but necessary [3].

Currently, there are no specific laboratories tests that can be performed to give physicians a definitive diagnosis, the clinical presentation and pathologic diagnosis are the two major diagnostic tools. Unfortunately, given the morphological overlap between the two entities, even the pathologic approach of a biopsy can be challenging and the search for specific histopathological features in sum with clinical history is essential for a more specific diagnosis between BD and CD [4].

Case Presentation

The patient is a 32 years old, female who was transferred to our hospital for evaluation of possible Behcet's disease versus IBD. Patient first presented to emergency department with symptoms of erythema nodosum, fatigue, weight loss and arthralgias. Per history, those symptoms worsened after she got the COVID vaccine and she ended up admitted with a flare including oral ulcers, rash, genital ulcers, and bloody diarrhea. Uveitis was not identified.

In ER, abdominal CT showed colitis with reactive mesenteric lymphadenopathy. Inpatient EGD showed diffuse esophageal ulcers and erosions, and colonoscopy showed patchy colonic ulcers with patchy distribution (Figure 1).

Previous treatment with prednisone was continued and Remicade was initiated with some relief at time [5].

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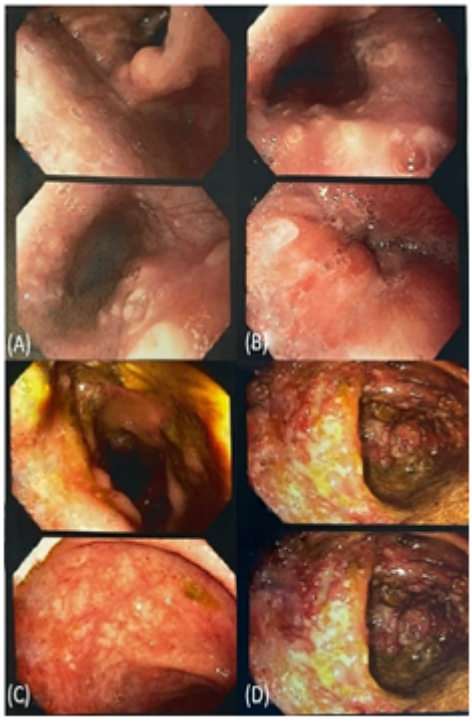

Figure 1. Upper gi endoscopy and colonoscopy: hypo pharynx (a), and esophagus (b) showing diffuse ulcers and erosions; stomach (c) and colon (d) showing patchy ulcers and mucosal erosions.

Patient then got hospitalized at outside hospital for surgical peri-rectal abscess drainage. She was then referred to our institute for better assessment for BD versus CD [6]. Same treatment with prednisone and IFX was continued at the time with possible introduction of Imuran. As part of the work up, a MRI was also performed, showing perianal fistula, presence of inter-sphincter abscess, focal recto sigmoid fixed narrowing and no defining evidence of active inflammatory bowel disease of the small or large bowel [7].

The biopsy slides were reviewed. The esophagus pathology revealed submucosal lymphohistiocytic infiltration involving vessels with associated vascular endothelial injury, consistent with granulomatous vasculitis (Figure 2). Duodenum biopsy showed focal mild active inflammation and focal villous blunting. Stomach biopsies demonstrate chronic active inflammation (Figure 3). Right colon biopsies show moderately active colitis with focal crypt abscess and vasculitis (Figure 4). However, the crypt architecture is reserved. Left colon biopsies demonstrate focal cryptitis, crypt abscess and ruptured crypt associated granulomas. Focal vasculitis is also present (Figure 5) [8].

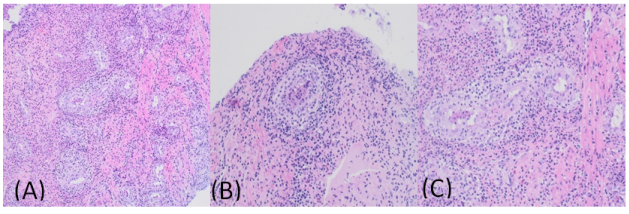

Figure 2. Esophagus biopsies demonstrate diffuse granulomatous vasculitis (A, 10x) and high power view (B, C, 20x).

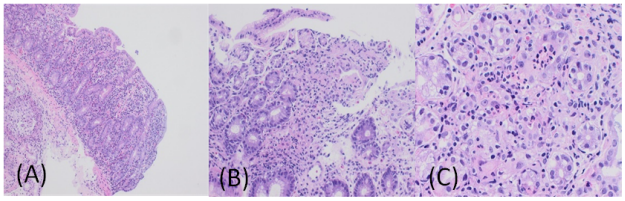

Figure 3. Duodenal biopsy shows focal villous blunting and mild acute inflammation (A, 10x). Stomach biopsy shows chronic active gastritis (B, 20x; C, 40x).

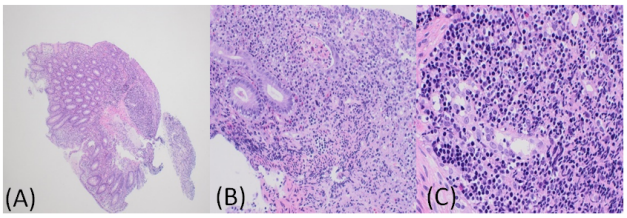

Figure 4. Right Colon biopsies show reserved crypt architecture at low power view (A, 4x). High power view shows cryptitis and crypt abscess (B, 20x) and focal vasculitis (C, 40X).

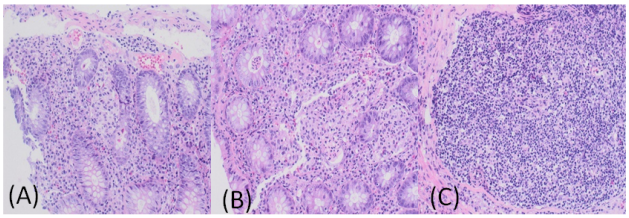

Figure 5. Left Colon biopsies show cryptic abscess with associated granuloma (A and B, 20x) and vasculitis (C, 20x).

The findings were most prominent in esophagus biopsies which demonstrate granulomatous-like vasculitis. Colon biopsies, did not show prominent chronicity changes (such as architectural distortion or Paneth cell metaplasia). Overall, in the setting of patient's history of oral and anal ulcer, perianal abscess and GI symptoms, the findings at the current biopsies were most compatible with vasculitis etiology such as Bechet's disease [9].

The patient continued follow up with general surgery and rheumatology and treatment with prednisone (12.5 mg/day) and remicade (every 8 weeks) was maintained. Imuran (175 mg/day) was introduced as well for her drug therapy. Patient is currently on follow up for perianal fistula drain with general surgery, as well as following up with GI and rheumatology for her symptoms and drugs optimization. Her symptoms is significantly improved [10].

Results and Discussion

BD is a chronic, multi-system, idiopathic inflammatory disease that presents with a wide range of symptoms, mainly including recurrent oral and genital ulcers, skin lesions, thrombophlebitis and uveitis. GI involvement is described to be more common in East Asia and Japan, whereas in the United States (US), only 1/3 of the cases involve the GI tract. BD is thought to be an autoimmune disease in genetically susceptible individuals. HLA-B51 is the allele which may play a role in this disease pathogenesis [11].

The clinical manifestations of the two entities can be very similar. There are no pathognomonic laboratory tests. Serum markers such as C-reactive protein and erythrocytes sedimentation rate are elevated in both conditions. Two potential serum markers for diagnosis BD are Anti-Saccharomyces Cerevisiae Antibody (ASCA) and Anit-α-Enolase Antibody (AAEA). The clinical presentation and pathologic diagnosis are two major diagnostic tools. Clinically, the BD patients usually present with recurrent aphthous stomatitis, oral and genital ulcers, erythema nodosum-like lesions. Ocular involvement including pan-uveitits and optic neuritis are common. Cardiac, neurologic and musculoskeletal manifestations are not uncommon. Gastrointestinal symptoms have been reported in 3% to 26% of BD patients and usually occur 4.5-6 years later than the onset of oral ulcerations. Rarely, intestinal lesions can occurs first. Anal strictures, fistula are frequently seen in CD patients but are rare in BD patients (1%) [12].

Endoscopic findings might be useful clues for distinguish the two entity. Large, patchy longitudinal ulcers, cobblestone appearance are more suggestive of CD. While round deep ulcers with sharp borders and ileocecal location are suggestive of BD. GI biopsies are the most common type of tissue available for pathology evaluation. The findings can be subtle with only non-specific inflammation and ulceration of the mucosa seen. The active cryptitis and cryptic abscess closely mimics infectious or drug induced colitis. The ruptured crypt associated granulomas closely mimic Crohn' disease. However, the chronicity change including architecture distortion and paneth cell metaplasia are essential to make a diagnosis of IBD. Prominent vasculitis is the most common feature for BD. In our case, this patient's perirectal abscess and crypt granulomas are closely mimicking CD, but the prominent granulomatous vasculitis is a unique finding and helpful clue.

Conclusion

In conclusion, our study described a rare case of BD with its endoscopic and pathologic findings, its potential diagnosis pitfall which mimicking IBD. Detailed history query, thorough physical examination, auto-antibody serology testing, in conjunction with possible GI tract endoscopy biopsy are important tools to establish the correct diagnosis.

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