CLINICAL VIGNETTE

Behçet's Disease with Rectovaginal Fistula: A Case Report

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Case Presentation

A 33-year-old female presented to the gastroenterology office for evaluation of diarrhea and lower abdominal cramps. She also reported significant weight loss, dark vaginal discharge, and passing gas from her vagina. Her history included genital and oral ulcers, arthralgias, atrial septal aneurysm, cholecystectomym, obesity, and Zoon vulvitis. She denied stool blood or mucus, rashes, joint aches, or eye redness. The patient is a social drinker and denied use if tobacco and other recreational drugs. Physical exam was notable for normal vital signs bilateral tenderness in the lower abdomen. Lab abnormalities included hemoglobin of 11.5 and a C-reactive protein of 1.4. She was scheduled for colonoscopy and imaging and a 3 mm rectal nodule was excised along with random biopsies. The colon and terminal ileum were normal. Biopsy of the rectal lesion revealed mild to moderately active proctitis with prominent lymphoid hyperplasia in the lamina propria favored to be reactive. There was no Paneth cell metaplasia or granulomas. No infectious agents were identified with immunostains for CMV, adenovirus, and spirochetes were negative. The random colon biopsies were normal. MRI of pelvis revealed fat saturation with fistulous tracts between the labia and anorectal junction. MR enterography showed a normal small bowel. She underwent an exam under anesthesia and seton placement. Daily azathioprine 50 mg and adalimumab induction were initiated followed by maintenance dosing at every two weeks, with improvement in fistula drainage.

Discussion

Behçet's disease (BD) is characterized as a chronic, multisystemic involvement of the joints, central nervous system, systemic vasculature, and gastrointestinal system. While the pathogenesis remains poorly understood, Behçet's is associated with HLA-B51 allele and MHC class 1 region, as well as circulating antibodies against alpha-enolase of endothelial cells, selenium binding protein and anti-saccharomyces cerevisiae antibodies.[1] Arthritis and arthralgias are commonly associated with BD joint disease, reported in about 50% of patients.[2] The systemic vasculature most affected are the superficial veins of the lower extremities, with vasculitis occurring in about one-third of patients. There may be involvement of the abdominal aorta, carotid, femoral, popliteal, and coronary arteries, although this is less often seen. Within the central nervous system, BD presents with parenchymal brain involvement, intracranial hypertension, and dural sinus thrombosis.[2] Gastrointestinal involvement involves mucosal ulceration in the ileum, cecum, and other parts of the colon. Other common clinical features of BD are recurrent cutaneous, oral (90-100% of patients), and urogenital ulcerations.[3] Less common findings including uveitis, secondary cataracts, glaucoma, and macular edema occur in about 3 percent of patients. There is no serologic marker or pathognomonic laboratory tests to diagnose BD. The diagnosis is established using clinical criteria following the revised International Criteria for Behcet's Disease (ICBD).[4]

Crohn's disease (CD) is a chronic inflammatory bowel disease that can affect any portion of the gastrointestinal tract, from the mouth to the anus.[5] CD typically manifests in young adulthood, ages 15-30, with increased prevalence in Caucasians.[6] Initial presentation for CD is often insidious but can include weight loss, fever, diarrhea, abdominal pain, and rectal bleeding. Physical examination of patients can determine urgency of care. Vital signs, abdominal and anorectal examinations are needed. Pelvic examinations should be considered due to the increased occurrence of abscesses, fistulas, and fissures.[5] Symptoms may be extra-intestinal involving eyes, joints and skin.[5] Diagnosis is usually made based on history and exam findings, with supporting laboratory, imaging, and histology.[6] Steroids initially are used to induce and maintain remission along with immunomodulators and biologics.[5] Surgical interventions may be needed for fistulas, abscesses and perianal disease.[5]

BD and CD are inflammatory diseases with potential extra-intestinal manifestations. The joint involvement of BD and CD are similar with both arthritis and arthralgias, although arthralgia is more common in BD. CNS manifestations are common in BD with lesions manifesting in aphasia, while CD rarely presents with pathological CNS involvement. While vasculitis is seen in both BD and CD, vasculitis in CD is limited to the gastrointestinal system. Another point of differentiation is that while BD and CD patients can both present with oral ulcers, BD patients more commonly manifest genital ulcers. Vasculitis most commonly involves the ileocecal area, although any part of the gastrointestinal tract may be involved in BD and CD. Both diseases have characteristic skipped lesions.[7] Lee et al., compared colonoscopy findings of 115 intestinal BD and 135 CD patients, and proposed diagnostic criteria. In their multivariate analysis, round shape, fewer numbers (≤5), focal distribution, discrete borders, deep penetrating, ileocecal location and absence of aphthous and cobblestone appearance were significantly dominant features of intestinal BD.[8] About a third

of patients with CD patients can develop anal complications such as stricture, fistula, and abscess formation, a complication seen in less than 1% BD patients.[9,10] Mesalamine medications (5-aminosalicylates) / sulfasalazine are indicated for mild intestinal BD and corticosteroids are used in moderate to severe disease. Thiopurines such as azathioprine or 6-mercaptopurine are used in patients with steroid dependent BD. Anti-TNF-α therapy has been used in CD as well BD with luminal involvement.[7]

Our patient initially presented with lower abdominal cramps, diarrhea, occasional constipation, as well as dark discharge and passing gas from her vagina. This constellation of symptoms did not allow discriminate between the two diseases of interest. In the absence of luminal involvement with the history of oral and genital ulcerations, the patient was believed to have BD, and satisfied the clinical criteria required for the diagnosis of BD.[6] Rectovaginal fistulas may be a complication of episiotomy or secondary to CD. Rectovaginal fistula caused by BD is extremely rare, with only 10 cases reported.[11-16] The main differentiating feature of CD is the typical granulomatous inflammation on pathology. However, the granulomatous change may not be seen in all patients with CD. This case represented a diagnostic challenge as the symptoms and signs of BD and CD often overlap.

Figures

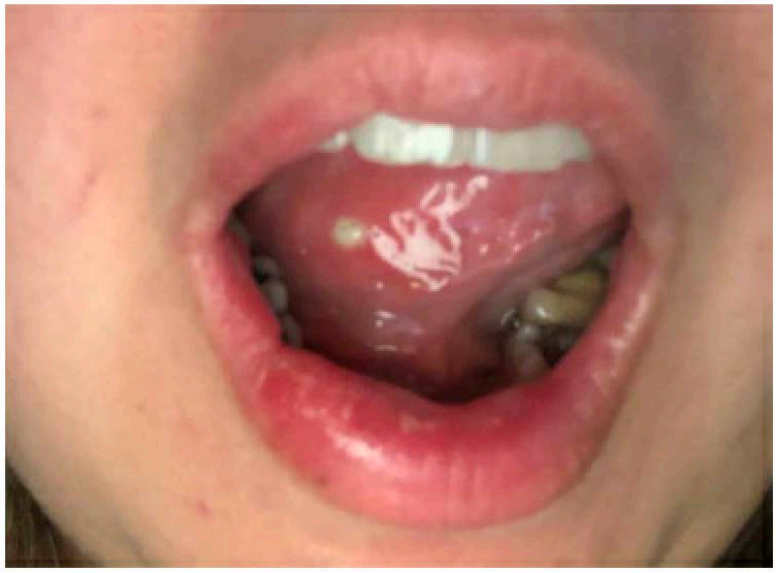

Figure 1: Oral ulcer on under surface of the tongue.

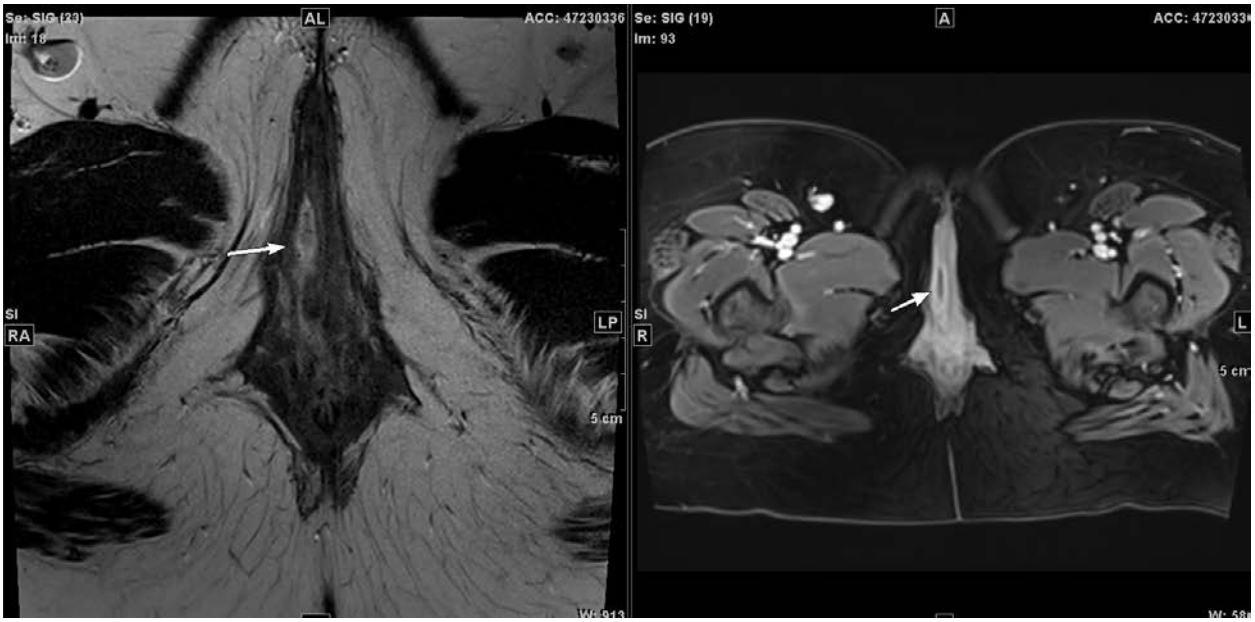

Figure 2: MRI with T2 axial images showing several vague to rim enhancing, curvilinear enhancing tracts in the perineum.

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