Whipple's Disease without Arthropathy in an Immunocompromised Patient

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INTRODUCTION

- Whipple's disease (WD) is a rare disorder caused by the pathogen *Tropheryma whipplei*.
- WD presents with non-specific symptoms which can mimic the presentation of other gastrointestinal pathogens, especially in immunocompromised patients.
- We report a unique presentation of WD involving an immunocompromised patient who experienced non-specific gastrointestinal symptoms without arthralgia, which is unique given that joint involvement is characteristic of this disease.

CASE DESCRIPTION/METHODS

- A 67-year-old man with a history of chronic hepatitis B infection and Human immunodeficiency virus (HIV) infection presented with weight loss, nausea, vomiting and myalgia.
- Of note, patient did not have arthralgia.
- Physical exam revealed cervical lymphadenopathy and diffuse abdominal tenderness.
- Patient was anemic (hemoglobin of 8.6 g/dL) with a normal white count.
- Endoscopy demonstrated erythema in the gastric body, lymphangiectasia of the duodenum, and increased granularity of the terminal ileum.

CASE DESCRIPTION/METHODS (cont.)

- Mucosal biopsies revealed macrophages in the lamina propria with focal histiocytic aggregates throughout the small bowel and cecum (Figure 1), with positive PAS staining (Figure 2), consistent with Whipple's disease.
- Confirmatory *T. whipplei* PCR testing was positive.

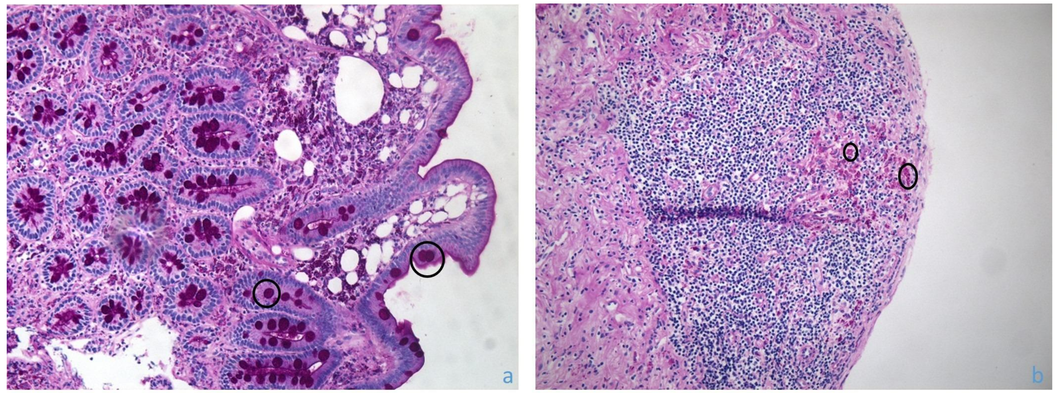

Figure 1 Periodic Acid-Schiff-Staining Macrophages (circles) Within the Lamina Propria of the Duodenal Bulb (a) and Cecum (b) as Seen via Light Microscopy with a 20x Objective Lens.

CONCLUSIONS

- WD is a rare diagnosis that must be considered in the differential diagnoses of patients presenting with unexplained nausea, vomiting, diarrhea and anemia. Furthermore, in patients with HIV, the possibilities would also include opportunistic gastrointestinal pathogens.
- Classic WD is characterized by diarrhea, weight loss, abdominal pain and extra-intestinal involvement manifesting as joint pain, endocarditis, dementia, supranuclear gaze palsy, and mediastinal lymphadenopathy.
- Diagnosis involves biopsy of affected tissue demonstrating foamy macrophages with PAS (+) substance, which can be confirmed by PCR or immunohistochemistry.
- Treatment involves an initial course of penicillin or ceftriaxone followed by a prolonged course of Bactrim, or a one-year course of doxycycline and hydroxychloroquine, followed by lifelong doxycycline therapy, which is more likely to prevent disease relapse.
- In the absence of suppressive therapy, relapse of WD is common and may lead to further complications, including neurological involvement.