A Rare Triad of Ulcerative Colitis, Large Vessel Vasculitis and Celiac Disease

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INTRODUCTION

Inflammatory bowel disease (IBD) has been associated with large-vessel vasculitis (LVV), with the diagnosis of IBD preceding that of LVV by years. We present for the first time in known literature a triad of concurrent ulcerative colitis (UC), aortitis and celiac disease.

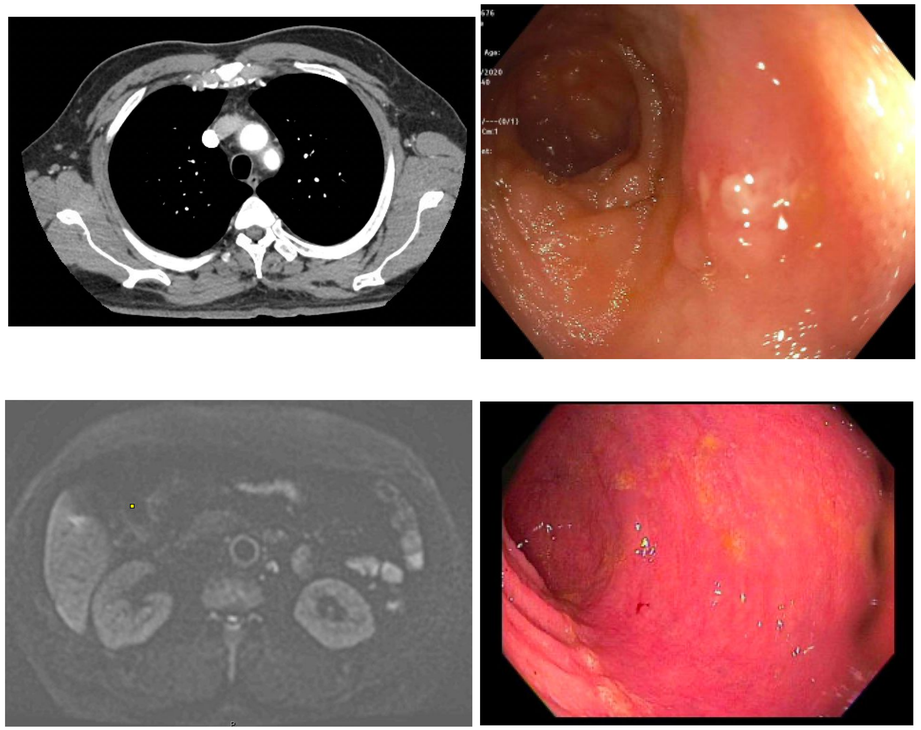
CASE STUDY

A 58 year old Hispanic man with a history of hypertension and gout presented with two weeks of intractable temporomandibular headaches, and two months of non-bloody diarrhea and weight loss.

Vitals were normal. Exam was significant for pale conjunctiva, normal cardiopulmonary exam. Abdomen soft and nontender to palpation. Brown patches on anterior shins bilaterally.

Labs significant for: Hgb 6.9, ESR 120, CRP 281, AST 47, ALT 75, Alk Phos 146, LDH 363. Anti gliadin IgA 50, Anti TTG IgA 23. ANA, C3, C4, proteinase-3 and myeloperoxidase antibodies were normal.

Initial endoscopy showed patchy, ulcerated mucosa involving the terminal ileum, sigmoid and descending colon and rectum. Patient was started on steroids. Pathology showed chronic active duodenitis with focal villous atrophy and diffuse intraepithelial lymphocytosis, and diffuse inactive chronic colitis from cecum to sigmoid colon, and severely active chronic proctitis. Abdominal MRI to evaluate a liver lesion incidentally showed hyperintense signal within the wall of the aorta at the level below the renal arteries to the common iliac bifurcation, suggesting abdominal aortitis. Patient underwent head, neck and chest imaging which subsequently revealed thoracic aortic wall thickening and edema, suggestive of aortitis/vasculitis.



Left figures: (Top) CT chest with contrast showing thoracic aortic wall edema; (Bottom) MRI Abdomen showing hyperintensity of abdominal aortic wall

Right figures: (Top) Initial colonoscopy showing skipped lesions; (Bottom) Repeat colonoscopy 4 months later showing active pancolitis

FURTHER DIAGNOSTIC STUDIES/TREATMENT

Patient was given pulse steroids. He underwent temporal artery biopsy was which was negative, and was discharged on a steroid taper with close GI and rheumatology follow up. Repeat endoscopy four months later to confirm IBD and celiac disease revealed patchy mild increased in intraepithelial lymphocytes in the duodenal mucosa with preserved villous architecture, as well as mild to moderate active colitis from ascending colon to rectum. He was started on a gluten free diet and adalimumab in combination with methotrexate for ulcerative colitis (UC) and large vessel vasculitis (LVV).

DISCUSSION

- About 10 case reports of patients with both UC and either Takayasu (TAK) or giant cell arteritis (GCA) have been described, with UC typically diagnosed 15-45 years before the vasculitis.
- Vasculitis in the GI tract can mimic IBD, making colonoscopy and biopsy crucial for diagnosis.
- HLA haplotypes A24, B52, and DR2 are associated with both UC and aortitis and interleukin-9, observed in temporal arteritis lesions, may be implicated in the pathogenesis of UC.
- Shared chromosomal variants between patients with UC and celiac disease may explain why IBD risk is up to 9-fold higher in patients with celiac disease.
- Our patient may have presented with isolated aortitis or an early form of GCA.
- Methotrexate is used to treat LVV and is combined with an anti-TNF agent to treat UC, as in our case.

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