**CLINICAL VIGNETTE**

# A Case of Collagenous Gastritis in a Patient with Newly Diagnosed Common Immunodeficiency Disease

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***Abstract***

Collagenous gastritis (CG) is an uncommon histopathologic disorder characterized by subepithelial collagen deposition. Adult CG is heterogeneous in its presentation and often diffusely involves the GI tract. Most patients present with watery diarrhea and can have underlying autoimmune, infiltrative lymphocytic, and/or malabsorptive conditions. While it is understood that hypogammaglobinemia may cause various pathological conditions in the intestine due to disruption of the gut mucosal immune barrier, the association between collagenous infiltrative disorders and primary immunodeficiency is rarely described. This case highlights a patient with newly diagnosed common immunodeficiency disease (CVID) who developed diarrhea, and was subsequently diagnosed with CG.

***Introduction***

Collagenous gastritis is a rare disorder characterized endoscopically by nodularity, and histologically by deposition of collagen bands within the subepithelial layer.[1] Collagenous gastritis was first described in 1989 in a pediatric patient presenting with recurrent abdominal pain and upper gastrointestinal bleeding.[2] Although the annual incidence rate of collagenous colitis is thought to be around 1.1-5.2 cases per 100,000 people, the incidence rate of collagenous gastritis is much more difficult to estimate as there have only been around 60 cases described in the literature.[3–6]

There are two described phenotypes of collagenous gastritis: pediatric-onset and adult-onset.[1] The pediatric phenotype typically manifests with recurrent abdominal pain and iron-deficiency anemia secondary to inflammation specific to the stomach.[1,7] Alternatively, the adult phenotype is characterized by abdominal pain, malabsorption, and watery diarrhea.[7] Although the pediatric-onset phenotype is limited to the stomach, the adult-onset phenotype is more heterogenous in presentation and often diffusely involves the gastrointestinal tract.[8] Adult patients with collagenous gastritis may also have collagenous sprue and collagenous colitis.[7]

This report describes a patient with collagenous gastritis and newly diagnosed CVID. Since the etiology of collagenous gastritis remains unknown, this case serves as a possible mechanism for the pathogenesis of disease.

***Case Report***

An 18-year-old male was referred for outpatient evaluation of watery diarrhea and intermittent, upper abdominal pain of 6 months duration. His history was notable for frequent infections, specifically upper respiratory infections, requiring recurrent antibiotic use since infancy. He had been previously evaluated by his primary care physician for his symptoms and undergone an extensive negative infectious evaluation for his diarrhea. His biochemical profile was notable for hypogammaglobulinemia with serum IgG level of 521 mg/dL and serum IgA level of 45 mg/dL. Upper endoscopy and colonoscopy were performed as additional evaluation of his symptoms.

The upper endoscopy demonstrated edematous mucosa with nodularity throughout the stomach (Figure 1). The colonoscopy did not demonstrate any gross abnormalities. Gastric biopsies demonstrated collagenous gastritis (Figure 2). They were negative for Helicobacter pylori, celiac disease, and intestinal metaplasia. Random biopsies throughout the colon were negative for active or chronic inflammation, collagenous or lymphocytic colitis, and did not show any histopathologic abnormalities.

The patient was treated with proton-pump-inhibitor (PPI) for an eight-week course, which did improve his symptoms. Additionally, he was referred to an immunologist and diagnosed with CVID. Treatment with IVIG was offered, but the decision was made to clinically monitor the patient.

***Discussion***

Collagenous gastritis is an uncommon disorder characterized by subepithelial deposition of collagen and inflammatory infiltrates in the lamina propria[1]. The clinical associations of adult collagenous gastritis have been described in various case reports and include autoimmune disorders such as Sjogren's syndrome, Graves' disease and systemic lupus erythematosus, and malabsorptive disorders, such as celiac disease.[9] Collagenous gastritis shares similar histopathological features with

collagenous sprue and colitis and these three entities have been associated with each other in some adult cases.[8] Finally, some case reports suggest an association between the development of collagenous gastritis and the use of antidepressants and olmesartan, an angiotensin II inhibitor.[10] However, the mechanisms by which these associated systemic diseases and medications predispose adults to collagenous gastritis is not well understood.[10]

Clinical symptoms of collagenous gastritis include abdominal pain, anemia, diarrhea, nausea, vomiting, weight loss, gastrointestinal bleeding, and fatigue.[1] The most prominent endoscopic finding appears to be mucosal nodularity, which is thought due to uneven distribution of inflammation causing glandular atrophy, and collagen deposition in the depressed mucosa surrounding the mucosal nodules.[1,11] Other endoscopic features include mucosal erythema, ulcerations, erosions, and hypertrophic rugal folds.[7]

Due to the rarity of this disorder, no well-established standardized therapy exists for collagenous gastritis. However, antisecretory therapy, steroids, sucralfate, bismuth subsalicylate, 5-aminosalicylic acid, and iron supplementation have been utilized with limited and varied success.[1,7] It is postulated that a dietary antigen may play a role in the pathogenesis of this disorder, but such an antigen is yet to be identified.[12]

The natural history of collagenous gastritis has not been well described. In the adult-onset phenotype of the disorder, the disease is chronic and intermittent without severe progression or significant mortality risk.[12] Patients with hypogammaglobulinemia or CVID may have gastrointestinal manifestations due to the disruption in the gut mucosal immune barrier.[13] In addition, there is a high rate of inflammatory, infectious and malignant gastrointestinal disorders in patients with either IgA deficiency or CVID.[14] The association between collagenous infiltrative gastrointestinal disorders and primary immunodeficiency has not been well described. Therefore, the mechanism by which patients with primary immunodeficiency develop collagenous gastritis remains poorly understood. However, it is postulated that recurrent infection with subsequent epithelial damage and accumulation of intraepithelial lymphocytosis with progression over time to subepithelial collagen accumulation could serve as a possible mechanism of disease.[15]

In conclusion, collagenous gastritis is a rare and complex clinicopathological condition. A high clinical suspicion in patients with immunodeficiency and gastrointestinal symptoms can lead to early diagnosis and management.

***Figures***

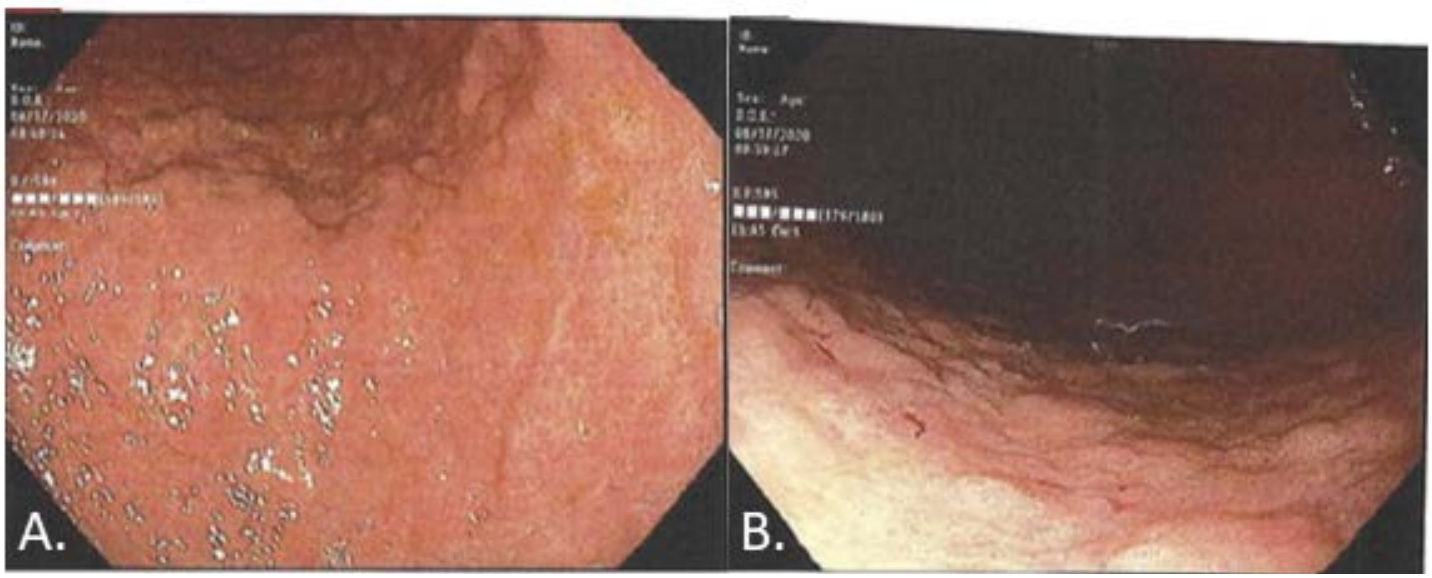


Figure 1. Diffuse nodularity and mild diffuse erythema in the gastric body (image B) and gastric antrum (image A).

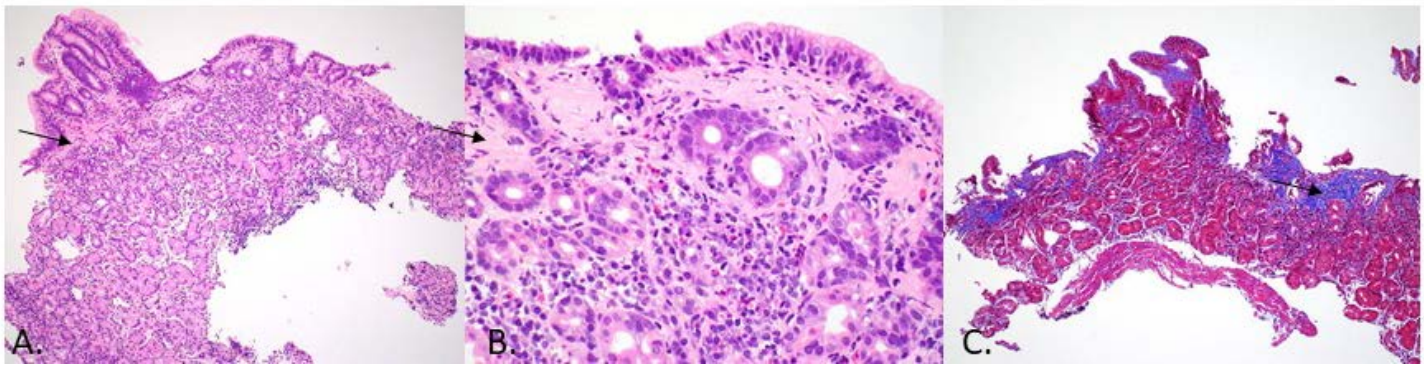

Figure 2. Gastric pathology displaying collagen band in the subepithelial region in low magnification (image A), high magnification (image B) and Trichrome stain (image C) with black arrow pointing to the collagen band.

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