

# Eosinophilic Gastroenteritis Misdiagnosed as Common Variable Immunodeficiency

by Beverton Moxey, Alison Schneider, Daniela Allende, Fernando Castro

## INTRODUCTION

**E**osinophilic gastroenteritis (EG) is an uncommon disorder characterized by either local or diffuse eosinophilic infiltration of the gastrointestinal tract. The stomach and small bowel is most commonly involved, but EG can occur in the esophagus, colon, pancreas and gallbladder. Clinical presentation varies depending on the anatomical location and the layer of the gastrointestinal tract involved.<sup>1</sup> Protein losing enteropathy is a manifestation of this disease secondary to damage of the gastrointestinal epithelium. Proteins such as albumin and most gamma globulins do not have rapid catabolic turnover rates and as such may be limited in their ability to respond to increased gastrointestinal losses.<sup>2</sup> We report a case of a patient diagnosed with common variable immunodeficiency (CVID) not responding to therapy. Evaluation determined that his suspected refractory CVID was in fact a protein losing enteropathy due to eosinophilic gastroenteritis.

## Case Report

A 23 year-old male recently diagnosed with CVID was referred for chronic diarrhea, abdominal pain and hypoalbuminemia. He denied recent travel or ill contacts. Over the past year he had recurrent bronchitis and sinusitis, which led to his diagnosis of CVID. Monthly intravenous gammaglobulin infusions were started, however, the regimen was changed to every three weeks due to persistently low serum immunoglobulins. He denied drug and food allergies and had no other

allergic conditions. His only medication was a daily multivitamin. Physical examination revealed 1+ pedal edema and was otherwise normal. Laboratory studies revealed a white blood cell count of 6.6 K/ul with absolute eosinophils of 1881 cells/uL (15-500), hemoglobin of 13.3 g/dl and platelets of 256 K/ul. Serum albumin was 2.8 g/dl and total protein was 5.4 g/dl. Serum IgG was 223 mg/dl (700-1600), IgA 58 mg/dl (70-400) and IgM 36 mg/dl (40-230). Autoimmune panel, tuberculosis (TB) quantiferon gold and celiac serology were negative. Stool studies for parasites were negative. Endoscopic evaluation found esophagitis, superficial erosions in the antrum and duodenitis. (Figure A and B). Pathology revealed chronic active gastritis with increased eosinophils >100/high power field (hpf), epithelial injury and muscularis mucosa involvement. *Helicobacter pylori* was not identified. There was also eosinophilic esophagitis (>58 eosinophils/hpf), degranulation and eosinophilic microabscesses. The duodenum exhibited slight aggregates of eosinophils in the lamina propria (16-18/hpf). The villous architecture was preserved and plasma cells were present in the lamina propria.<sup>3</sup> No evidence of parasites was noted. (Figure C and D). The patient was started on Prednisone 50 mg/day for EG with a taper by 10 mg every 4 weeks. At 8 week follow-up he had normalization of his eosinophil count, serum albumin and immunoglobulins. Follow up endoscopy and biopsies of the stomach and esophagus were normal.

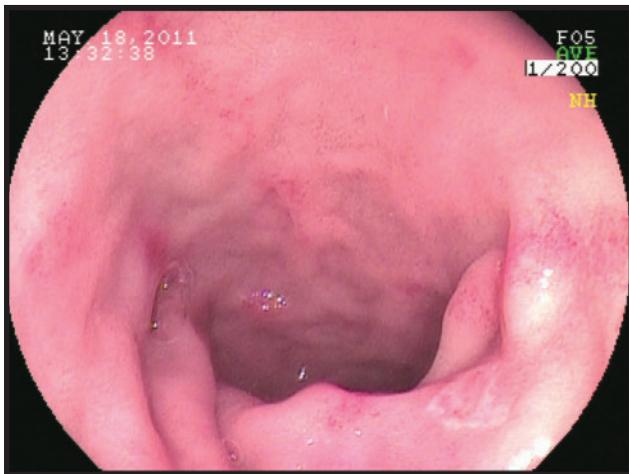
## DISCUSSION

This is the first case reported to the authors' knowledge in which the presentation of a very different entity (CVID) was in fact due to EG. Eosinophilic gastroenteritis, first described by Kaijser in 1937, is a rare condition

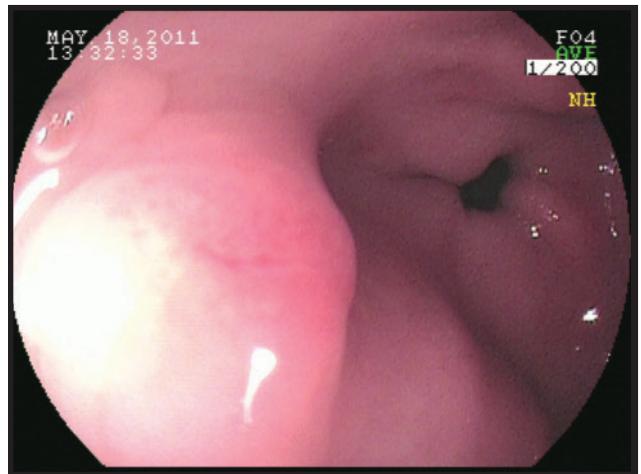
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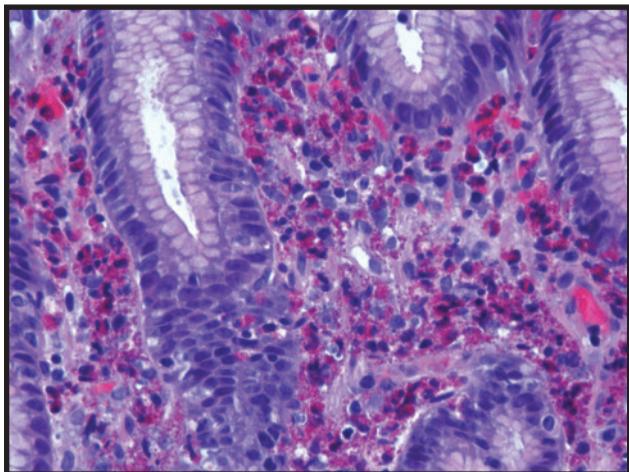
A CASE REPORT



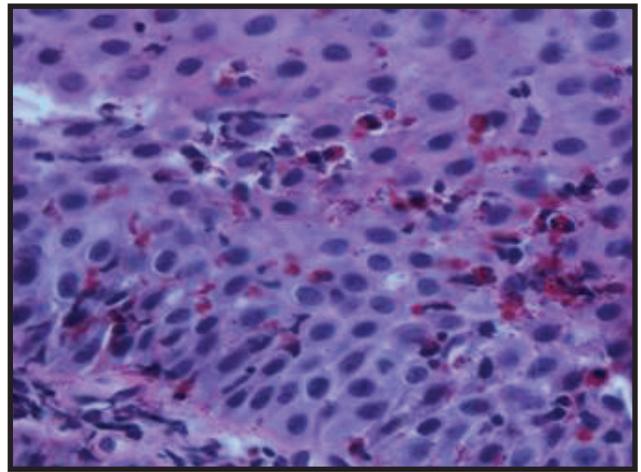
**Figure A.** Duodenal Bulb revealing patchy erythema and inflammation



**Figure B.** Superficial erosion in the antrum



**Figure C.** Eosinophilic gastritis with significant eosinophilic infiltrates and epithelial injury from antrum (HE, 400x).



**Figure D.** Eosinophilic esophagitis with prominent degranulation of eosinophils and microabscesses (HE, 400x).

involving eosinophilic infiltrates in the gastrointestinal tract. Klein et al developed a classification based on maximal depth of tissue involvement: mucosal, muscle layer or subserosal layer and showed the clinical features of EG vary based on the region and depth of eosinophilic infiltration.<sup>4</sup> Patients with mucosal layer disease commonly have abdominal pain, nausea, vomiting and diarrhea. They may also present with a protein losing enteropathy as in this patient. Those with muscularis invasion typically have obstructive symptoms and patients with serosal involvement may present with ascites. Current knowledge of treatment is derived mostly from small studies and anecdotal experience using dietary therapy, steroids, mast cell inhibitors, leukotriene receptor antagonists, surgery and biologics.<sup>1</sup> Corticosteroids are often the mainstay of treatment. The natural history is not well described

but clinical studies show that about 40% patients have spontaneous remission without relapse after treatment and 50% have a more complex, chronic course characterized by unpredictable relapses.<sup>5</sup> ■

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