Chasing Albumin in Chronic Diarrhea - A New Subtype of Protein Losing Enteropathy: Autoimmune Cryptolytic Enterocolitis

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INTRODUCTION

Protein losing enteropathy (PLE) can be classified into erosive, non-erosive, and increased interstitial pressure PLE. Presenting signs are often chronic non-bloody diarrhea, anasarca, abdominal pain, and weight loss. We present a complex case of a PLE presentation with a novel underlying etiopathology.

CASE DESCRIPTION

30-year-old woman with a history of migraines presented with severe abdominal pain and chronic diarrhea.

Medications: long-term combined estrogen-progestin oral contraceptive pills (OCP) and chronic high dose non-steroidal anti-inflammatory (NSAIDs)

Exposures: +mycoplasma pneumoniae one month prior, without other infectious exposures, +psilocybin mushroom

Laboratory tests:

- Basic Serum: Hemoglobin 6 g/dL, Albumin 0.5 g/dL (Figure 1)
- Infectious Work up: stool pathogen PCR negative, Mycoplasma IgM+
- Inflammatory Biomarkers: fecal calprotectin >3000µg/g
- Alpha-1 anti-trypsin clearance >783 mL/ 24hr (ULN: 49mL/24hr)
- Celiac serologies: TTG-IgA, TTG-IgG and gliadin-IgG/IgA negative
- Neuroendocrine workup: glucagon and pancreatic polypeptide within normal limits, low gastrin, chromogranin A elevated (on proton pump inhibitor), calcitonin < 2.0
- Anti-*S. cerevisiae* Antibody: negative
- Coombs positive Warm autoimmune hemolytic anemia
- Normal B cell switching studies (common variable immunodeficiency ruled out)

Imaging:

- CT angiography: excluded ischemia and vasculitis revealed diffuse enterocolitis
- Magnetic resonance enterography revealed ileal ulcers.

Endoscopic Evaluation:

- Diffuse superficial ulcerations terminal ileum and colon with diffuse mucosal sloughing (Figure 2).

Figure 1:

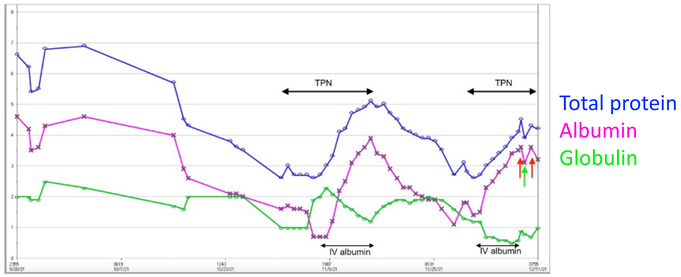


Figure 2:

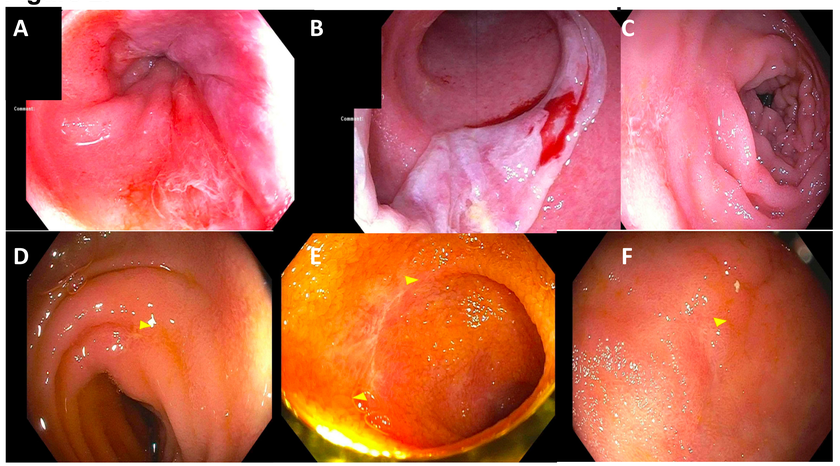
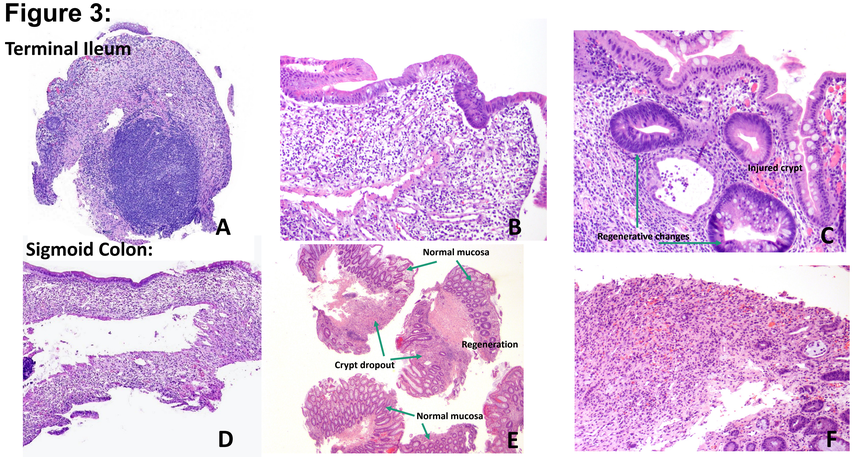


Figure 3:



HOSPITAL COURSE

Histopathology (Figure 3):

- Diffusely injured crypts with crypt drop-out and minimal inflammation
- No signs of infection, inflammatory bowel disease (IBD) or autoimmune enteritis.

Treatment:

- Albumin infusions for goal albumin >3g/dL
- Systemic and topical steroids were trialed without improvement
- Total Parenteral Nutrition (TPN)
- Vedolizumab empirically initiated with good symptomatic response and improved oral intake, however subsequent *Clostridiodes difficile* infection led worsening diarrhea and hypoalbuminemia leading to undetectable vedolizumab levels requiring resumption of IV albumin.

DISCUSSION

Given the absence of classic IBD findings, negative infectious and immunological testing, we present the first case of autoimmune cryptolytic enterocolitis, a histopathologic diagnosis of unclear etiology and pathogenesis. The patient has responded to vedolizumab infusions as an empiric treatment for an IBD-like entity, however medications such as monoclonal antibodies require adequate serum albumin levels, rendering the sustainability of such treatments challenging in PLE and thus proactive drug level monitoring necessary. Supporting the autoimmune component of this entity, is a possible concomitant potential pathway of molecular mimicry in the setting of post-mycoplasma infection along with chronic high-dose NSAID and OCP exposures.

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