Autoimmune Enteropathy, A Rare Cause of Intractable Diarrhea, Presenting in an 80-Year-Old-Man
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Case Presentation
An 80 year-old-male presented with a four month history of profound diarrhea amounting to 8 to 10 watery, non-bloody bowel movements a day resulting in hypokalemia and a 60 pound weight loss. Gastrin, calcitonin, thyroid function panel, celiac work up, urine 5-HIAA, metanephrines, catecholamines and infectious stool studies were negative.

The patient underwent an upper endoscopy, enteroscopy, and colonoscopy. Duodenal biopsies revealed chronic active inflammation with villous atrophy. Jejunal biopsies were consistent with a diagnosis of autoimmune enteropathy (AIE).

The patient commenced with systemic steroid and immunomodulator therapy. His symptoms improved to one bowel movement a day with an increase in his weight. However, as the steroids were tapered, the diarrhea returned. Thereafter, therapy with intravenous infusion of infliximab was initiated. The patient is currently receiving azathioprine and infliximab while re-attempting to taper steroid therapy in an effort to control symptoms.

Introduction
AIE is a rare disease characterized by intractable diarrhea, malabsorption and anorexia leading to severe weight loss. Dietary modifications are generally ineffective and patients may require total parenteral nutrition. This condition more often affects infants within the first 6 months of life and is described only in case reports in adults. In adults, the average age at diagnosis is 55 and patients usually require immuno-suppressive therapy.

Diagnostic Criteria
The diagnostic criteria for AIE includes chronic diarrhea of more than 6 weeks that does not improve with dietary modification, malabsorption, partial or complete blunting of the small bowel villi, deep crypt lymphocytosis, increased crypt apoptotic bodies, minimal intraepithelial lymphocytosis, and exclusion of other causes of villous atrophy. The presence of anti-enterocyte or anti-goblet cell antibodies is supportive of the diagnosis, although not confirmatory.

Extraintestinal manifestations include hypothyroidism, nephritic and nephrotic syndrome, autoimmune hemolytic anemia, rheumatoid arthritis, dermatitis/atopic eczema, autoimmune hepatitis, and chronic pancreatitis. The differential diagnosis of AIE includes celiac disease and common variable immune deficiency (CVID). The presence of intraepithelial cell lymphocytosis is found in celiac disease, while immunoglobulin deficiency and a normal albumin suggest CVID.

Management
Management of AIE is clinically challenging because of its uncommon nature and paucity of evidence based clinical guidelines. Corticosteroids including budesonide and prednisone are first line therapies in the treatment of AIE. In patients refractory to steroids, treatment and maintenance of remission with azathioprine, 6-mercaptopurine, cyclosporine, tacrolimus, mycophenolate mofetil, sirolimus, infliximab, and rituximab has been described.

We present a case of AIE, a rare entity, diagnosed in an 80 year old male whose symptoms became steroid refractory and required the use of an immuno-modulator and a biologic to control symptoms of malabsorption, diarrhea, and weight loss.