AUTOIMMUNE ENTEROPATHY: A RARE CAUSE OF SMALL INTESTINAL VILLOUS ATROPHY

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INTRODUCTION

Autoimmune enteropathy (AIE) is a rare cause of immune-mediated small intestinal villous atrophy that was initially recognized in children for causing intractable diarrhea. Onset in adulthood is extraordinarily rare and can pose a diagnostic challenge due to its similarities with celiac disease.

CASE PRESENTATION

- A 54 year-old previously healthy woman was hospitalized with profuse watery diarrhea, abdominal pain, and weight loss for six months.
- CT enterography showed hyperenhancement of the small bowel consistent with enteritis, without evidence of stricture or abscess.
- EGD showed diffuse edema of the duodenal mucosa with villous blunting and cracked-earth appearance **(Figure 1A, 1B**). The distal small bowel and colon appeared normal.
- Pathology from duodenal biopsies showed chronic active duodenitis with characteristics detailed in **Figure 1C**. Similar findings were seen in random ileal biopsies, while random colon biopsies were normal.
- Anti-enterocyte antibodies and anti-transglutaminase antibodies were negative.
- Patient was treated for AIE with intravenous methylprednisolone with subsequent improvement in her diarrhea. She required temporary parenteral nutrition for her severe malabsorption and malnutrition.

CASE IMAGES

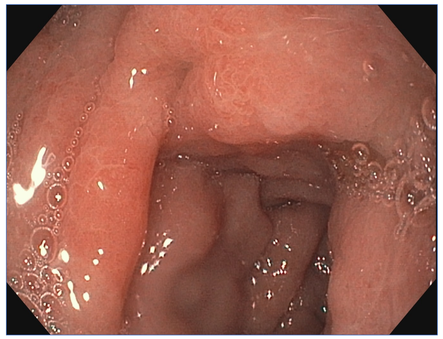

Figure 1A. EGD showing villous blunting and edema of duodenal mucosa.

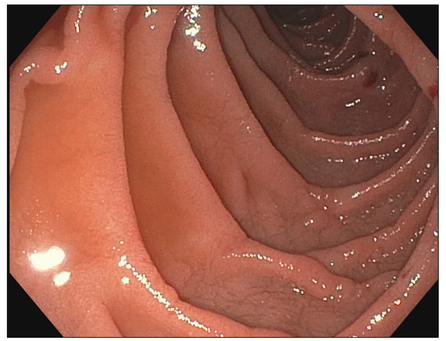

Figure 1B. EGD showing cracked-earth appearance of duodenal mucosa.

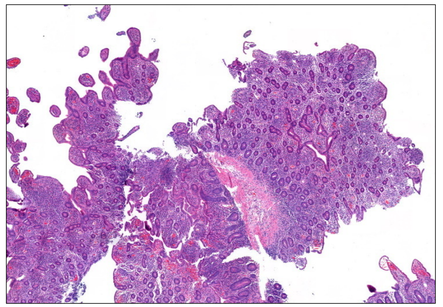

Figure 1C. Duodenal biopsy showing chronic active duodenitis characterized by marked villous blunting, reduced goblet and Paneth cells, neutrophilic and lymphocytic infiltration of deep crypts, and basal layer apoptosis.

DISCUSSION

- AIE should be suspected in a patient with:
  - chronic diarrhea
  - signs of severe malabsorption
  - intestinal biopsies showing villous blunting, absence of goblet or Paneth cells, and increased crypt apoptotic bodies
  - absence of other causes of villous atrophy such as Celiac disease, tropical sprue, Whipple disease, HIV enteropathy, and CVID
- Anti-enterocyte antibodies are associated with AIE (55-80%), but are not required for diagnosis.
- Steroids are the mainstay of therapy and have variable effect.
- Case reports have described benefit from steroid-sparing therapies (azathioprine, anti-TNF).

REFERENCES

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