CASE REPORT

Intestinal goblet cell autoantibody associated enteropathy

K Hori, Y Fukuda, T Tomita, T Kosaka, K Tamura, T Nishigami, A Kubota, T Shimoyama

This report describes a case of refractory enteropathy with circulating intestinal goblet cell autoantibodies (IGA). A 19 year old man with hyperthyroidism had suffered from protracted diarrhoea for nearly 10 years. Histological examination showed evidence of collagenous enterocolitis. The diarrhoea did not improve despite fasting under total parenteral nutrition. An immunofluorescence assay demonstrated IGA without anti-enterocyte autoantibodies, the hallmark of autoimmune enteropathy, although other criteria were fulfilled. None of 109 controls, including 55 cases of inflammatory bowel disease and one of lymphocytic colitis, had IGA. This case is considered to be a variant of autoimmune enteropathy, and might be a distinct entity.

Collagenous enterocolitis (CE) is a rare condition characterised by collagenous colitis with involvement of the small intestine. Autoimmune enteropathy (AE) is also a rare disorder defined by the following criteria: protracted diarrhoea, no response to total parenteral nutrition (TPN), predisposition to autoimmune disease, and no immunodeficiency. We report the first case of CE with circulating autoantibodies to mucin in intestinal goblet cells but not autoantibodies to the cytoplasm of enterocytes, which are the hallmark of AE.

CASE REPORT

A 19 year old Japanese man was admitted to our hospital in July 2001 with a long-term history of continuous watery diarrhoea, which had averaged five to six times daily since December 1991 at the age of 10 years. His past medical history included atopic dermatitis and uveitis at the age of 10 and 17 years, respectively. He suffered from hyperthyroidism two years before admission, and has been treated with thiamazole. His mother has systemic lupus erythematosus and Sjögren's syndrome, and his brother has hypophysial dwarfism and idiopathic leucopenia.

He was 146 cm tall and weighed 32 kg. His bowel movements had increased with distention. Laboratory data on admission showed severe hypokalaemia at a concentration of 1.5 mmol/litre. There were mild liver and pancreatic dysfunction (alanine aminotransferase, 64 U/litre; aspartate aminotransferase, 64 U/litre; serum amylase, 198 U/litre; and trypsin, 675 ng/ml). He had no viral hepatitis B or C infection, but anti-adult T cell leukaemia virus antibody was positive. Quantitative immunoglobulins were normal (IgG, 9340 mg/litre; IgA, 1170 mg/litre; and IgM, 370 mg/litre). All stool cultures were negative. An α1 antitrypsin test showed no evidence of protein losing enteropathy. A 5-h D-xylene (5 g) absorption test was consistent with severe malabsorption, with a five hour urine collection of 0.3 g (normal, > 1.5 g). Because the diarrhoea continued at an average of 2200 g/day over a six week period, despite fasting under TPN, a diagnosis of coeliac disease could not be made.

Abdominal ultrasonography and computed tomography showed no abnormalities. Barium studies of the small intestine and colorectum were normal. Upper and lower endoscopical examinations revealed no gross abnormalities, except for reactive lymphoid hyperplasia in the terminal ileum. However, biopsies of the duodenum and entire colorectum demonstrated subepithelial collagen deposits in the sigmoid colon with a striking depletion of goblet cells (bar, 100 μm).

Figure 1  [A] Subepithelial collagen bands around tubules are seen in the duodenal mucosa with goblet cell depletion and increased lymphoplasmacytic infiltrate (bar, 50 μm; haematoxylin and eosin stain). [B] The Masson-trichrome stain showed subepithelial collagen deposits in the sigmoid colon with a striking depletion of goblet cells (bar, 100 μm).

Abbreviations: AE, autoimmune enteropathy; CE, collagenous enterocolitis; IE, intestinal goblet cell autoantibody associated enteropathy; IGA, intestinal goblet cell autoantibodies; TPN, total parenteral nutrition
autoantibodies in patients with Crohn’s disease (n = 32), the pancreas at a titre of 1/40. We also tested for the above colonic goblet cells at a titre of 1/80, and in the acinar cells of diagnosis of CE was made.

The duodenum and terminal ileum. There was crypt hyperplasia in the duodenum, but hypoplasia in the terminal ileum. A diagnosis of CE was made.

This case is thought to be a variant of autoimmune enteropathy, and might be a distinct entity — IGA associated enteropathy.

**REFERENCES**


