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An unusual etiology of diarrhea: primary intestinal lymphangiectasia

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Dear Editor,

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Primary intestinal lymphangiectasia (PIL) is a rare disorder characterized by malformed and dilated intestinal lymphatic vessels, resulting in impaired drainage and leakage of lymph fluid into the gastrointestinal lumen. This leads to protein-losing enteropathy, causing chronic diarrhea, peripheral edema, weight loss, and hypoalbuminemia due to loss of proteins, lymphocytes, and lipids[1]. Diagnosis relies on endoscopic visualization of pathognomonic white mucosal plaques (representing dilated lymphatics) and histopathological confirmation of lymphatic dilation, after excluding secondary causes like malignancy, surgery, or inflammatory conditions[2].

A 34-year-old woman presented with a 4-month history of persistent diarrhea, lethargy, and progressive weight loss. Physical examination revealed bilateral ankle edema. Laboratory tests showed normal complete blood count and renal function, with no albuminuria. Liver function tests were within normal ranges except for a serum albumin level of 26 g/L (reference 35-50 g/L). Stool examinations for ova, parasites, and enteric pathogens were negative on three consecutive tests. Clostridium difficile toxin B polymerase chain reaction (PCR) analysis was negative. Abdominal computed tomography demonstrated normal intestinal wall architecture. Gastroscopy revealed unremarkable esophageal and gastric mucosa. Colonoscopy identified multiple discrete white mucosal plaques distributed throughout the entire colon (Figures 1A-C). Histopathological evaluation of colonic biopsies showed villous enlargement with dilated lymphatic vessels (Figure 1D), confirming intestinal lymphangiectasia. Secondary causes including prior abdominal surgery, radiation exposure, and systemic diseases were excluded, establishing the diagnosis of primary intestinal lymphangiectasia. The patient was initiated on a high-protein, low-fat diet with medium-chain triglyceride supplementation. At 4-month follow-up, clinical improvement was evidenced by weight gain, resolution of edema, reduced diarrhea frequency, and increased serum albumin levels (38 g/L). Repeat colonoscopy demonstrated complete disappearance of the characteristic mucosal plaques.

This case provides two key clinical insights: Firstly, PIL must be considered in patients with unexplained hypoalbuminemia and chronic diarrhea when common etiologies are ruled out, emphasizing the diagnostic necessity of endoscopy with

targeted biopsies[3]. Secondly, dietary intervention with medium-chain triglyceride supplementation-which bypasses defective lymphatics via direct portal venous absorption-can achieve rapid clinical, biochemical, and endoscopic resolution[4], as demonstrated by this patient's normalized albumin (38 g/L), resolved edema, reduced diarrhea, weight gain, and complete disappearance of mucosal plaques within just 4 months. Early initiation of this targeted nutritional therapy is crucial for effective disease reversal.

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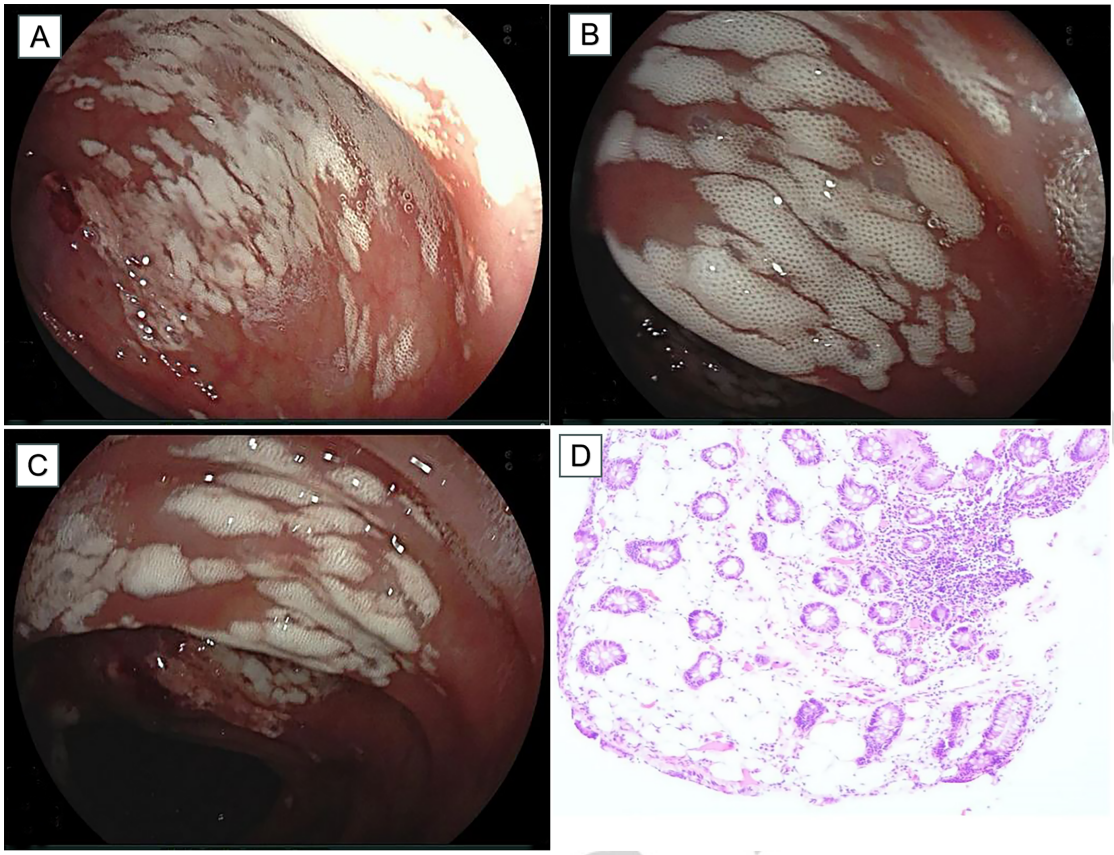

Figure 1: A,B,C: Colonoscopy identified multiple discrete white mucosal plaques distributed throughout the entire colon; **D:** Histopathological evaluation of colonic biopsies showed villous enlargement with dilated lymphatic vessels (HE, ×200).