Primary Intestinal Lymphangiectasia in a Middle-Aged Female

Primary intestinal lymphangiectasia (PIL) is a rare disorder caused by impaired intestinal lymphatic drainage due to congenital lymphatic hypoplasia. It was first described by Waldman *et al.* in 1961,[1] about 205 cases have been reported.[2] The disease usually manifests before 3 years of age, and only occasionally diagnosed in adulthood. Here, we report a case of a middle-aged female who presented with chronic diarrhea and was finally diagnosed as a case of PIL.

A 49-year-old female presented to the Outpatient Department of our hospital with complaints of diarrhea for 2 years. The patient reported an occurrence of diarrhea four to five times daily, which was semisolid to watery in consistency with absence of blood, mucus, or oil. The diarrhea was also not associated with tenesmus, urgency, or pain in the abdomen. On examination, her body mass index was 18 kg/m^2 and there was presence of bilateral pitting edema in the lower limbs. Routine laboratory investigation revealed hemoglobin of 9 gm/dl, total leukocyte count of 4100/mm^3 with 14% of lymphocytes, platelet count of 1.6 lakh/mm^3 and hypoproteinemia (serum protein, 3.90 gm/dl; serum albumin, 2.10 gm/dl; serum globulin, 1.8 gm/dl). She was non-reactive for HIV and hepatitis B. Stool routine and microscopic examination was negative on three occasions and culture was negative. Upper gastrointestinal endoscopy revealed multiple scattered whitish spots in the second part of the duodenum [Figure 1]. Multiple biopsies from it revealed dilated lymphatic channels in lamina propria, muscularis mucosae and submucosa [Figure 2]. Therefore, a diagnosis of intestinal lymphangiectasia (IL) was made and further work up was done to rule out secondary causes. Her colonoscopy with terminal ileoscopy, contrast-enhanced computed tomography of chest and abdomen, echocardiography, autoimmune profile, immunoglobulin levels and hormonal profile were all normal except for reduced immunoglobulin levels. Accordingly, a final diagnosis of PIL was made and the patient was started on a low-fat, high-protein diet with medium-chain triglyceride (MCT) supplementation. Subsequently, her condition improved, and the diarrhea stopped after 1 month. The upper gastrointestinal endoscopy was repeated after 6 months, and the findings were normal with no whitish spots in the duodenum [Figure 3].

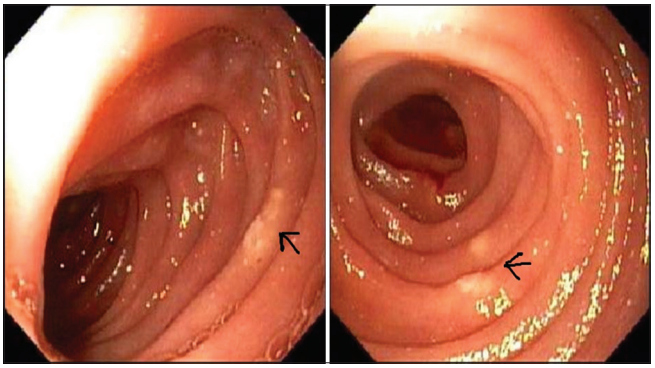

Figure 1: Upper gastrointestinal endoscopy showing whitish spots in the second part of duodenum

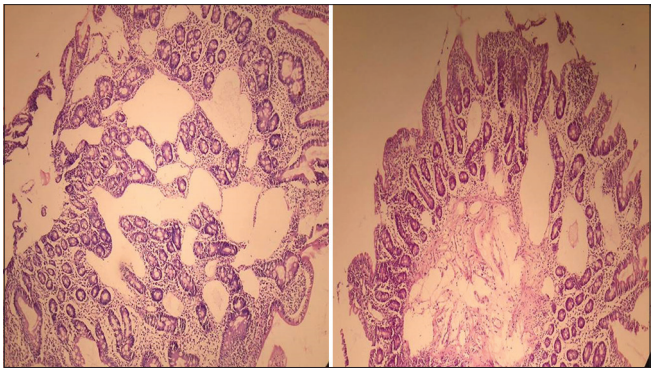

Figure 2: Duodenal biopsy showing dilated lacteals in mucosa and submucosa

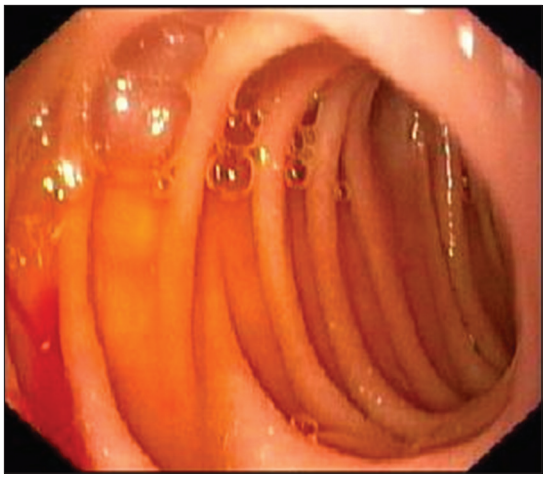

Figure 3: Normal upper gastrointestinal endoscopy after 6 months of dietary modification

PIL is a congenital malformation that presents with chronic diarrhea and malabsorption and is diagnosed after ruling

out secondary causes. Its late onset can be explained by the patchy involvement of the small bowel. Secondary intestinal lymphangiectasia can occur due to various causes such as constrictive pericarditis, intestinal lymphoma, intestinal tuberculosis, sarcoidosis, scleroderma, Whipple's disease, Crohn's disease, autoimmune polyglandular syndrome and radiation or chemotherapy induced retroperitoneal fibrosis.[3] Intestinal lymphangiectasia in adults has few or no features of malabsorption, as in our case. Its diagnosis is done by specific endoscopic features and corresponding histopathological findings. Endoscopic features of intestinal lymphangiectasia are scattered white punctate snowflake lesions in small intestine due to dilated lacteals that cannot be washed away. Upper gastrointestinal endoscopy can diagnose 86% of intestinal lymphangiectasia cases.[4] Histopathology of small bowel will show characteristic dilated lacteals in the mucosa and submucosa. The cornerstone of its treatment is diet modification. Diet should be low in fat with substitution of long chain fatty acid with MCT, as it directly enters the portal venous system bypassing the intestinal lacteal system, thereby preventing lacteal engorgement and rupture.[5] Most of the patient respond with this treatment in few weeks with reversal of clinical and biochemical signs.

In conclusion, PIL is a rare cause of chronic diarrhea, especially in adulthood, that requires meticulous examination of duodenum, as early diagnosis and treatment can reduce the morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms from the patient. The patient has given her consent for images and other clinical information to be reported in the Journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal the identity.

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Conflicts of interest

There are no conflicts of interest.

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