
Photoclinic



Figure 1. Small bowel series.

A 17-year-old boy was referred for evaluation of chronic watery diarrhea. He had a history of large-volume non-bloody diarrhea for four months, about three times a day, not associated with pain or weight loss. There was a history of perioral paresthesia with hand and foot cramps for three months, which was treated by three grams of daily elemental calcium and 8000 IU of calcitriol (Rocaltrol®). On physical examination, the vital signs were normal without fever or any peripheral lymphadenopathy. The body mass index was 20 kg/m². Mild anemia and

bilateral pedal edema were detected, along with iron deficiency anemia, hypokalemia, hypocalcemia, hypoalbuminemia, and pan-hypogammaglobulinemia. Direct stool examination and culture showed no pathogen and it was negative for occult blood. Fat droplets were detected in stool by Sudan III staining. HIV antibody was negative. Abdominal ultrasonography, chest radiography, echocardiography, and upper gastrointestinal endoscopy were normal. Abdominal and pelvic computed tomography with intravenous and oral contrast showed no ascites or lymphadenopathy. Images from his small bowel series are shown in Figure 1.

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What is Your Diagnosis?

See pages 475 – 476 for the diagnosis

Photoclinic Diagnosis:

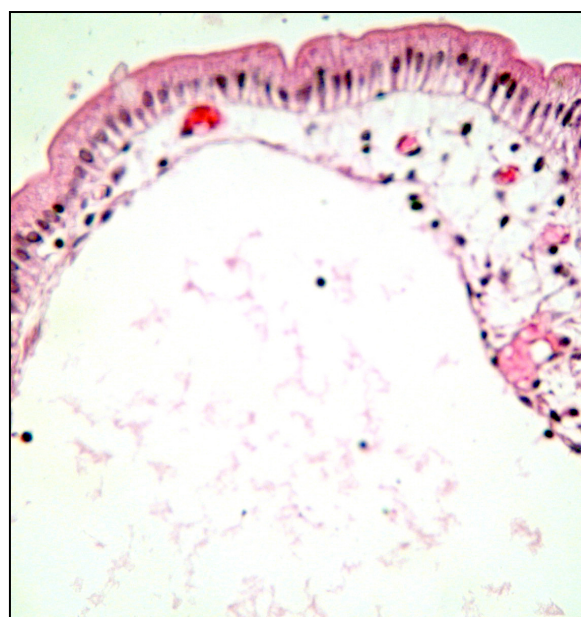
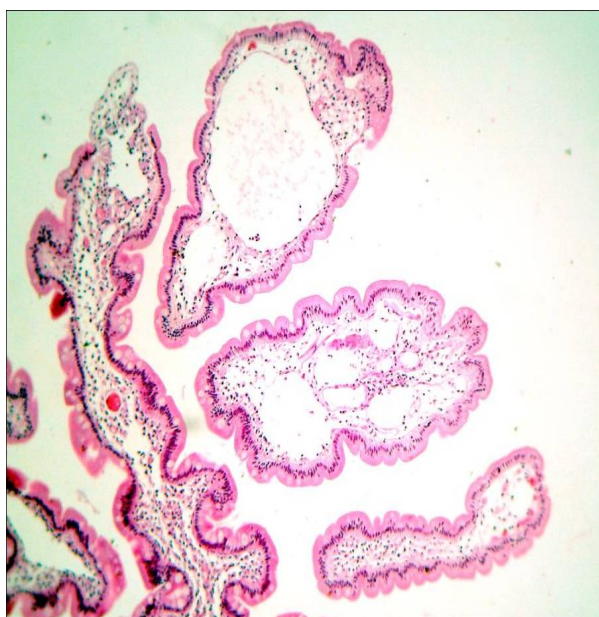
Primary Intestinal Lymphangiectasia

Figure 2. Histopathology of the biopsy specimen from the second part of the duodenum (H&E staining).

Protein-losing enteropathy is characterized by excessive loss of serum proteins into the gut lumen. It should be considered in patients with hypoproteinemia in whom malnutrition, proteinuria, and liver diseases have been excluded. Once plasma proteins pass into the gut lumen, they are degraded to amino acids and reabsorbed into the portal circulation. Iron, lipids, and trace elements also may be lost in the gut. Intestinal mucosal injury with or without erosions or ulcerations as what is seen in inflammatory bowel disease and celiac disease, cause leakage of plasma proteins into the gut.

Increased lymphatic pressure in the gut due to granulomatous and neoplastic involvement of the lymphatic system or dilated lymphatic vessels of the gut (intestinal lymphangiectasia) and venous stasis in diseases such as congestive heart failure or constrictive pericarditis can cause the leakage of protein via the surface epithelium into the gut.

Intestinal lymphangiectasia is abnormal dilatation of lymphatic channels of gut mucosa leading to loss of lymph with immunoglobulins and lymphocytes into the gut. The disorder may be congenital (primary), or secondary to processes that obstruct lymphatic drainage of the gut or raise the central venous pressure.

Primary intestinal lymphangiectasia is characterized by diffuse or localized ectasia of gut lymphatics, which may be associated with other lymphatic abnormalities in the body.

Clinical manifestations of primary intestinal lymphangiectasia are intermittent diarrhea, steatorrhea (in some cases), edema, and sometimes pleural and pericardial effusion.

The diagnosis of primary intestinal lymphangiectasia is established based on the clinical manifestations, laboratory, and histologic findings.

Laboratory findings are hypoproteinemia with decreased serum levels of albumin, IgG, IgM, IgA, transferrin, and ceruloplasmin. Loss of lymphocytes into the gut can result in significant lymphocytopenia with alteration in cellular immunity. Patients who have steatorrhea may develop fat-soluble vitamin deficiencies. Small bowel contrast studies may show thickened, nodular mucosal folds that simulate stacked coins.¹ In endoscopic evaluation, scattered white spots, which have been described as having a snowflake-like appearance, may be seen in the small intestinal mucosa.^{2,3} Histologic examination reveals dilated lymphatics (Figure 2).⁴

The mainstay of treatment is a low-fat, high-protein, and medium-chain triglyceride diet.^{5, 6} Hypogammaglobulinemia and lymphopenia are not usually severe, but sometimes are associated with increased rate of infections. Patients with recurrent infections and low serum IgG may benefit from gammaglobulin infusions.

Some patients require additional supplementation with calcium salts and water soluble forms of fat soluble vitamins.

The need for diet therapy is often permanent, although occasional spontaneous remissions do occur. Diarrhea and electrolyte disturbances were cured by low-fat diet in this patient.

References

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