(Figure 1). She underwent subtotal resection of the small intestine. The pathology of the resection was consistent with PAN (Figure 2); therefore, treatment protocol with intravenous pulse steroid and cyclophosphamide was started. However, she developed severe leukopenia on the 8th day of immunosuppressive treatment and granulocyte colony-stimulating factor (G-CSF) was initiated. Repeat bronchoscopy revealed widespread tissue destruction due to infection. Septic shock developed despite antibiotic and antifungal treatment, and she succumbed to death subsequently.

The diagnosis of PAN is established with the combination of clinical, serological, hematological, radiological, and histological findings. Histological evaluation of biopsy or resection material showing inflammation and fibrinoid necrosis of small- to medium-sized arteries is the gold standard for diagnosis. Immunosuppression with corticosteroid and cyclophosphamide is the mainstay of treatment particularly for the patients with gastrointestinal involvement. However, treatment should be customized according to the patient's general condition and comorbidities. Omitting or reducing the dose of cyclophosphamide in an effort to avoid severe bone marrow suppression in patients with a recent history of or tendency to infections would be a reasonable approach.

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A rare case of primary intestinal lymphangiectasia diagnosed by double balloon endoscopy

Çift balon endoskopi ile tanı konulan nadir bir primer intestinal lenfanjiektazi olgusu

To the Editor,

Protein-losing enteropathies (PLEs) are a wide variety of disorders characterized by excessive loss of serum proteins into the gastrointestinal tract. Intestinal lymphangiectasia (IL) is a rare cause of

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PLE, and its diagnosis depends on endoscopic and pathological examination of the small intestine (1). Herein, we describe a case of primary IL.

CASE REPORT

A 67-year-old woman was admitted to our hospital with a two-year history of leg edema. Her physical examination revealed only bilateral pitting pretibial edema. The laboratory findings demonstrated hypoproteinemia and hypoalbuminemia. The liver and kidney function tests, lactate dehydrogenase level, ferritin level, transferrin saturation, and vitamin B12 level were within normal limits. Antinuclear, anti-dsDNA, anti-gliadin, and anti-endomysium antibodies were negative. Urine protein excretion was normal. The stool α_1-antitrypsin level was high (8.91 mg/g dry stool, normal range: 0.5-3.7 mg/g dry stool). Her abdominopelvic ultrasonography, small intestine barium contrast graphy and thoracoabdominal computed tomography were normal. Double balloon endoscopy (DBE) revealed edematous jejunal mucosa with wide whitish spotty lesions (Figure 1). The jejunal biopsies showed lymphatic dilatation in the lamina propria and submucosa.

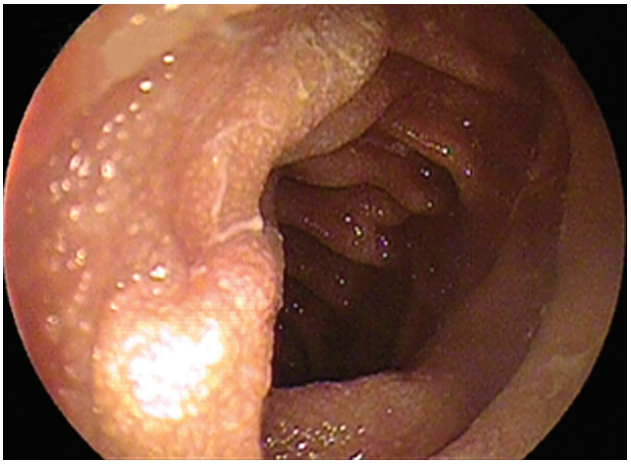

Figure 1. Double balloon endoscopy showing wide whitish spotty lesions located on the edematous mucosa of the jejunum.

DISCUSSION

Protein-losing enteropathies (PLEs) are a group of diseases that are characterized by excessive enteric protein loss. IL is a rare cause of PLE and it is associated with impaired intestinal lymphatics (1). Primary IL is a congenital disorder of the lymphatic system. Secondary IL is associated with diseases that impair lymph flow. Constrictive pericarditis, Fontan operation, congestive heart failure, intestinal lymphoma, lymph-enteric fistula, chronic pancreatitis and pseudocyst, Whipple's disease, Crohn's disease, intestinal tuberculosis, and sarcoidosis are the most common causes.

Limb edema, diarrhea, abdominal pain, nausea, and vomiting are common symptoms (2). Laboratory findings are hypoproteinemia with decreased serum levels of albumin, immunoglobulin (Ig)G, IgM, IgA, and transferrin. The stool α_1-antitrypsin level is a reliable indicator of the enteral protein loss (3). Diagnosis of the IL depends on the characteristic features defined during endoscopic and pathological examination. Macroscopic abnormalities are creamy yellow small intestinal villi corresponding to dilated lymphatics and mucosal edema. Pathologic examination reveals dilated mucosal and submucosal lymphatic vessels with normal polyclonal plasma cells (3, 4).

In conclusion, IL is a rare disorder that should be considered in a patient presenting with limb edema, hypoalbuminemia and lymphocytopenia. As the diagnosis depends on the endoscopic and histological examination, DBE is valuable in the evaluation of PLEs to identify the presence of IL and to obtain biopsies when upper and lower endoscopies do not define any abnormalities.

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