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Intestinal Lymphangiectasia: Evaluation by CT and Scintigraphy

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Abstract. Intestinal lymphangiectasia caused severe diarrhea and generalized edema in a 40-year-old man. The diagnosis was established by clinical, laboratory, and duodenal biopsy findings. The abnormalities detected on computed tomography (CT) and scintigraphy using ^{99m}Tc human serum albumin are herein described and pertinent literature is briefly reviewed.

Key words: Intestinal lymphangiectasia, CT diagnosis—Protein-losing enteropathy, radionuclide studies.

Intestinal lymphangiectasia is an uncommon disease characterized by dilated intestinal lymphatics, enteric protein loss, edema, hypoalbuminemia, and lymphopenia [1, 2]. This entity is usually diagnosed on the basis of a characteristic small bowel mucosal histology along with demonstration of enteric protein loss. Enteric protein loss is usually demonstrated using ^{51}Cr- or ^{131}I-labeled human albumin and timed measurement of fecal excretion of radioactivity or by measuring fecal clearance of α_1-antitrypsin [2, 3]. All these techniques require fecal collection and are not available in the majority of centers, especially in developing countries. We report a patient with intestinal lymphangiectasia in whom ^{99m}Tc-labeled albumin scintigraphy, a recently described technique for demonstration of enteric protein loss [4], and computed tomography (CT) played a role in determining the diagnosis. CT also showed a streaky appearance of the small bowel not previously described, but apparently caused by markedly dilated lymphatic channels.

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Case Report

A 40-year-old man presented with diarrhea and generalized swelling of the body, including the feet, abdominal wall, and the periorbital region, for 1 year. The diarrhea consisted of 8–10 semiformed to watery large-volume stools per day without mucus or blood. He also complained of generalized weakness, anorexia, and a feeling of lethargy. There were no abdominal pain, fever, or night sweats. He had received antituberculous chemotherapy for a period of 2 months prior to presentation to us without relief in symptoms. Physical examination revealed symmetrical bilateral pedal edema, ascites, and left-sided pleural effusion. There was no evidence of neck vein engorgement, lymph node enlargement at any site, or hepatosplenomegaly.

Investigations revealed a hemoglobin concentration of 15 g/dl and a total peripheral leukocyte count of 7200/μl (7% lymphocytes). Renal and liver function tests were normal. Serum cholesterol concentration was 100 mg/dl. Total serum protein and albumin concentration were 2.3 and 1.2 g/dl, respectively. Serum IgG, IgA, and IgM levels were 853, 282, and 159 mg/dl, respectively, and 24-h urinary protein excretion was 150 mg.

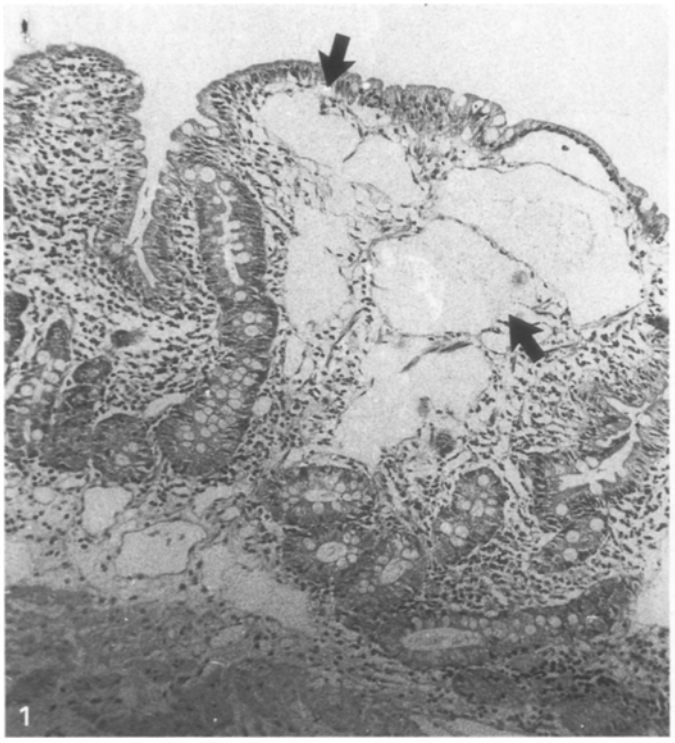
Chest x-ray revealed a left-sided pleural effusion. Pleural fluid had a protein concentration of 0.4 g/dl and 156 leukocytes/μl. Ascitic fluid examination showed protein concentration of 0.6 g/dl, triglycerides 7 mg/dl, and 72 leukocytes/μl.

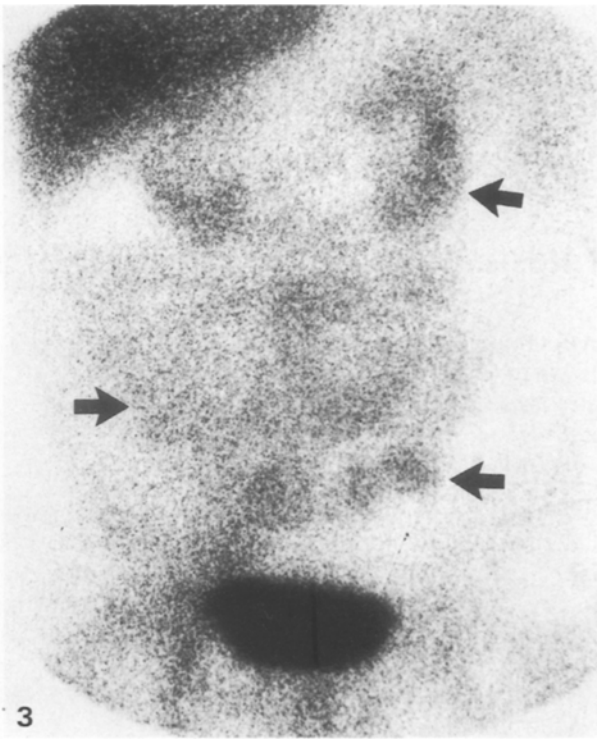
Fecal fat content while receiving a 50-g fat diet was 6 g/24 h (patient had intolerable diarrhea with 100 g/day fat diet) and D-xylose excretion was 0.9 g/5 g/5 h. Upper gastrointestinal endoscopy revealed absence of esophageal varices but presence of numerous elevated yellowish blebs in the second part of the duodenum. Biopsy from these areas showed edematous and blunted villi with ectatic lymphatics within them and in the lamina propria (Fig. 1).

A contrast-enhanced CT scan showed large amounts of ascitic fluid and diffuse thickening of the small bowel loops with hypodense intramural streaks in the absence of any mesenteric or paraaortic lymph node enlargement. Liver and spleen were normal (Fig. 2).

For demonstration of gastrointestinal protein loss, 10 mCi of freshly prepared ^{99m}Tc-labeled human albumin was injected intravenously, and serial abdominal images obtained at 15-min intervals for the first hour and subsequently at hourly intervals for the next 6 h, as previously described [4]. It revealed a progressive collection of radiopharmaceutical in the small bowel from 15 min onward which moved distally with time confirming enteric protein loss (Fig. 3). A ^{99m}Tc-labeled sulfur colloid scan showed normal hepatic uptake.

A diagnosis of protein-losing enteropathy due to primary in-





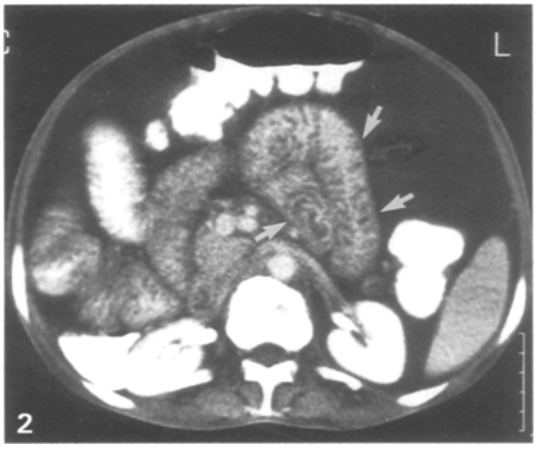


Fig. 1. Biopsy of the intestinal mucosa shows blunted villi and markedly ectatic lymphatic channels in the lamina propria (*arrows*). Hematoxylin & eosin stain; original magnification, ×200.

Fig. 2. Contrast-enhanced CT scan through the midabdomen demonstrates moderately dilated small intestine with hypodense linear streaks intramurally due to lymphangiectasia (*arrows*). The bowel loops are separated by ascitic fluid.

Fig. 3. Anterior view of the abdomen 45 min after injection of ^{99m}Tc-human serum albumin shows excretion of radionuclide into the small bowel loops (*arrows*). The liver and urinary bladder are also visualized.

testinal lymphangiectasia was made. The patient was treated with a diet rich in proteins and medium-chain triglycerides, intravenous albumin infusions, and low dose diuretics. His diarrhea decreased and weakness and edema improved. Three months later, he had a sudden worsening of diarrhea lasting for 3–4 days and died at home.

Discussion

In this patient, hypoproteinemia and hypoalbuminemia were attributed to enteric protein loss in view of normal liver functions, normal ^{99m}Tc-sulfur colloid scan, absence of varices, and significant proteinuria. This was confirmed by rapid accumulation of ^{99m}Tc-labeled albumin in the small bowel after intravenous administration. Of various causes of gastrointestinal protein loss, intestinal lymphangiectasia was suggested by low serum immunoglobulin concentration and lymphopenia [1, 2]. The diagnosis was confirmed by visualization of mucosal blebs on upper gastrointestinal endoscopy and a characteristic duodenal mucosal biopsy appearance [1, 2]. Secondary intestinal lymphangiectasia was unlikely in view of absence of right heart failure and of lymphadenopathy both clinically and on investigations.

Detection of gastrointestinal plasma protein loss is usually done by measuring the fecal excretion of various intravenously administered radiolabeled

macromolecules, such as ^{51}Cr- and ^{131}I-labeled albumin or by measuring the fecal clearance of α_1-antitrypsin [2, 3]. These methods are cumbersome as they involve prolonged stool collection and fecal handling [3]. In addition, the above radiopharmaceuticals are not readily available. Recently, it has been shown that serial imaging of the abdomen following intravenous administration of ^{99m}Tc-labeled human serum albumin can help in the diagnosis of enteric protein loss by showing accumulation of radioactivity in the intestinal system [4]. This technique has the following advantages: convenience, no need for fecal collection, easy availability of the isotope and radiopharmaceutical (as it is frequently used for blood pool studies), and the ability to provide results in 1 day. This technique has been previously used with success in a few patients with enteric protein loss [5, 6]; it also provided a clue to the site of protein loss in our case (Fig. 3).

There has been only one report of CT findings in this disease [7]. In that report, diffuse nodular thickening of the small bowel without lymphadenopathy or hepatosplenomegaly was reported in two patients with this disease. In our patient, this finding was associated with a linear hypodense streaking pattern of the small bowel which has not been previously reported. These hypodense lesions are probably due to dilated lymphatic channels which are seen on histology (Figs. 1 and 2). Though this sign appears to be specific, further observations will be needed to assess its exact nature.

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