Lymphogenic Enteropathy from Thoracic Duct Obstruction

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Summary

A twelve year-old girl was studied with protein-losing enteropathy, confirmed by $^{51}$Cr-labeled albumin test. Pedal lymphangiography demonstrated an obstructed thoracic duct at the level of the cisterna chyli with mesenteric reflux and lymphatic collateralization through the liver.

A large proportion of patients with protein-losing enteropathy have insufficiency of the intestinal lymph circulation. Foldi (1) calls this form of protein-losing enteropathy lymphogenic enteropathy, thus distinguishing it from exudative enteropathy where pathologic changes of the mucosa are primary and lymphatics have only a secondary role.

Enteric loss of protein is caused by a variety of abnormalities of the intestinal lymph circulation, but in some instances, the primary cause must be looked for outside the lymphatic system (e.g. increased central venous pressure or nephrotic syndrome). Intestinal lymphatic insufficiency may be congenital or acquired. Congenital disturbances are usually manifest at infancy or early childhood (2-4), and frequently the peripheral lymphatic system is also affected (5-7). In some patients, a strong hereditary trait is demonstrable (8, 9). Acquired disease often results from tumor invasion of lymphatics (10-12), but inflammation too may impair flow of intestinal lymph (13). In rare instances, fistulae may develop between the lymphatic system and the bowel lumen producing considerable protein loss (14-16), occasionally in conjunction with chylous ascites (17). In this report, a young girl developed obstruction to the lower part of the thoracic duct with protein-losing enteropathy.

An unusual compensatory network of lymphatic drainage developed through the liver and then towards the anterior mediastinum thereby circumventing the obstructed thoracic duct.
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Fig. 1 Microscopy of small bowel. a) Jejunal villi. Cross-sections of extremely dilated lymphatics in the stroma of the villi, H.E. x 133.

Case Report

A twelve year-old girl with a past history of epidemic hepatitis and scarlet fever at six years developed blisters on the legs, followed soon thereafter by diarrhea, fever, pain in the major joints accompanied by frequent hypocalcaemic tetany, and edema of the face and legs two months before admission. On admission she manifested generalized anasarca and was gravely ill. Beneath the right scapula, a moderate size haemangioma was present. Both legs showed partly scarred, erythemic papules. The liver edge was barely palpable. Laboratory tests indicated an erythrocyte sedimentation rate of 5 mm in 1 hour; leucocyte count 2000 mm³, with normal differential. Serum protein level was 2.7-3.1 gm/100 ml. Calcium 5.1-7 mg/100 ml. Magnesium 0.4-1.8 meq/liter. Serum level of cholesterol was 170 mg/100 ml, prothrombin activity was 100%, prothrombin consumption was 29%. 131-I-labeled triolein test for fat excreted in the stool, absorption of glucose, xylose and lactose, enzymatic studies of stool, blood, gastric and duodenal juices, bacteriologic and parasitologic examinations of stool, blood and duodenal fluid and urinalysis were all unremarkable. 51Cr-albumin test indicated 47.4% of total value excreted in the stool for four days (normal < 1%) (19). Contrast X-ray examination of the small bowel was consistent with slight "enteritis". Peroral biopsy of the jejunum demonstrated on microscopy bulky villi and dilated thin-walled lymph vessels visible in the stroma occupying a major portion of the tunica propria (fig. 1a). Dilated lymph vessels were also found in the submucosa (Fig. 1b), suggestive of lymph stasis and blockage of drainage.

Bipedal lymphangiography (Lipiodol Ultra-fluide, Guerbet, France) showed normal subcutaneous lymphatics in the medial side of the thigh. In contrast, the medial iliac lymph channels were absent and paraortic lymphatics were dilated. The cisterna chyli was visualized at L1, but the thoracic duct was not seen. Instead, contrast medium regurgitated into the subhepatic-hepatic and mesenteric lymphatics (Fig. 2a-b). The periportal lymphatics were dilated and tortuous and primarily those draining toward the right lobe of the liver were opacified including hepatic capsular lymph vessels (Fig. 3). From the liver, contrast medium flowed into a parasternal lymph node above the diaphragm. Only the upper 1/3 of the thoracic duct was opacified. X-rays taken 24 hours later demonstrated several enlarged granular lymph nodes in the inguinal and iliac regions bilaterally. The paraortic nodes were small with little contrast medium. Mesenteric reflux was demonstrated by opacified mesenteric lymph nodes in-
Fig. 1 Microscopy of small bowel. b) Basal mucous membrane and submucosa. The lymphatic vessels are significantly dilated in the loose connective tissue of the submucosa, H.E. x 100.

cluding two paraceliac nodes (Fig. 4a-b). Some contrast medium was still visible in the lymphatic network of the liver. Diaphragmatic and parasternal lymph nodes were also evident on plain chest X-rays (Fig. 5a-b), and the right parasternal lymphatic chain and lymph nodes along the upper part of the anterior mediastinum were easily seen. Lymph nodes in the posterior mediastinum did not visualize. Even five days after lymphangiography, a small quantity of contrast medium was still seen in the hepatic lymphatic network.

Discussion

In this patient, obstruction to the thoracic duct above the cisterna chyli restricted lymph flow and led to enteric loss of lymph with associated symptoms of edema. Lymphography demonstrated retrograde filling of mesenteric lymphatics although no contrast material was seen in the small bowel lumen. \(^{51}\)Cr-albumin test documented the loss of serum proteins, while microscopy after biopsy of the small bowel demonstrated dilatation of small intestinal lymph vessels.

Mesenteric reflux previously had been observed with (12, 14) or without (20) obstruction of the thoracic duct. In the patients reported by Desprez-Curely et al. (14) and Wenzel et al. (16) enteric loss of protein was induced by a lymph-intestinal fistula and
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Fig. 2  a) Anteroposterior. b) Lateral abdominal lymphangiogram. Dilated, varicose paraaortic lymphatics. Increased hepatic and mesenteric reflux with collateral network draining towards lymph nodes of the diaphragm.

Fig. 3  Frontal abdominal tomogram in the median plan of the body. Subhepatic and periportal lymphatics are extremely dilated, similarly to mesenteric lymph vessels. Contrast medium in the deep lymphatic system of the liver has flowed retrograde into the liver’s surface.

during lymphography contrast medium was seen in the small bowel.

In the present patient, as a result of increased intralymphatic pressure behind the obstructed thoracic duct, deep and capsular lymphatics of the liver filled retrograde. Hence, periportal lymph flow that normally drains away from the liver (21, 22) reversed and drained into superficial and deep lymphatic system of the liver. Thus a collateral lymphatic network developed through the liver and passed via the diaphragm towards parasternal lymphatics. Such reversal of flow presumes high pressure to overcome normal lymphatic valvular resistance. Although radioisotopic examination of Magnenat and Delaloye (24) suggested an absence of drainage between the hepatic capsular lymphatic system toward a deep lymphatic network, in pathologic conditions there may be such lymphatic pathways including antegrade transhepatic lymph flow. During lymphography, the upper portion of the thoracic duct was visualized which is unusual as right parasternal lymph
Fig. 4 a) Anteroposterior. b) Lateral lymphadenogram of the abdomen. Even after 24 hours, a large quantity of contrast medium is found in the lymphatic system of the liver. Pancreaticocolic, mesenteric and two paracolic lymph nodes are clearly visible.

Flow normally drains toward the right lymphatic duct (21, 25), not the thoracic duct. However, Küttn er (26) observed that material injected into the liver progressed as far as the lymph nodes in the supraclavicular fossa on both sides. Magnenat and Delaloye (24) demonstrated with scintigraphy that radioactive colloid instilled into Glisson's capsule of the right lobe of

Fig. 5 a) Anteroposterior. b) Lateral chest X-ray picture. Diaphragmatic, parastrernal and upper anterior mediastinal lymph nodes are opacified.
the liver not only reached parasternal lymphatics, but also the left supraclavicular and upper mediastinal lymph nodes. The findings suggest that potentially, communications exist between the right parasternal chain and the cervical section of the thoracic duct, which under pathologic conditions may enlarge.

The cause of obstruction of the thoracic duct in this patient was not clarified. No findings were uncovered suggesting tumor or inflammation, known causes of secondary lymphangiectasia. Although the patient may develop fibrosis in the future there was no evidence of functional damage at the time of radiologic examination.

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The Flow Rate and Macromolecule Content of Hilar Lymph from the Rabbit's Kidney under Conditions of Renal Venous Pressure Elevation and Restriction of Renal Function — Studies on the Origin of Renal Lymph.*

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Summary

The aim of this investigation was to ascertain whether renal lymph can be correctly thought of as a mixed product consisting of protein-free fluid reabsorbed from the renal tubules together with plasma. By progressively compressing the terminal segment of the renal vein the hydrostatic pressure within it was raised to over 40 mmHg. The glomerular filtration rate (GFR) and the absolute amount of fluid reabsorbed fell simultaneously to 20% of their initial values. Lymph flow in a renal hilar lymphatic rose proportionately to renal vein pressure by a factor of up to 10. The concentrations of protein and polyvinylpyrrolidone (PVP, MW 110,000) in the renal lymph remained unaffected at levels of 76 ± 13% and 56 ± 21% of their respective plasma concentrations. The intrarenal 131I-albumin pool also remained unchanged at 19.7 ± 4.3 ml plasma/100 g kidney. It is concluded that renal lymph is formed mainly by a filtration process from plasma. The results provide evidence against any admixture of protein-free reabsorbed fluid with the renal lymph.

Introduction

As long ago as 1931 Drinker et al. (5) postulated that renal lymph was derived from two sources: the protein-free reabsorbate from the renal tubules and a protein-containing plasma filtrate originating from the peritubular capillaries. If this mixture hypothesis is correct it would be expected that experimental modification directed at one or other of these sources would cause changes in the protein concentration of the lymph. The aim of the present study was to put this to the test. Depending on species and technique, the protein concentration of renal hilar lymph ranges from 30-70% of the plasma protein.

* Part of this work was presented at the Spring Meeting of the German Physiological Society, Erlangen, 1970