

LYMPHOGRAPHIA

NON-TRAUMATIC CHYLOTHORAX ASSOCIATED WITH DIFFUSE LYMPHATIC DYSPLASIA

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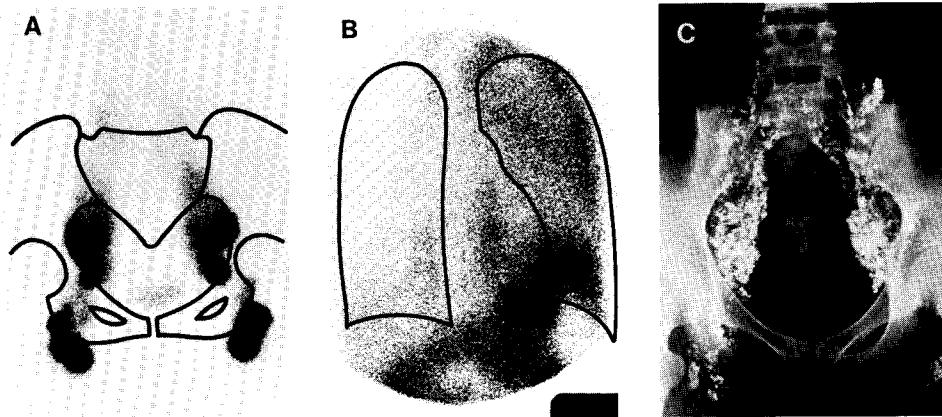


Fig. 1: ^{99m}Tc -rhenium colloid lymphoscintigraphy of the pelvis 15 minutes after injection (A) and of the left hemithorax (anterior view) 3 hours later (B). Note intense bilateral radioactivity in inguinal and lower iliac nodes (A) and diffuse radioactivity in the left chest (B) indicative of lymph leakage into the pleural space. Radioactivity in the upper abdomen (B) represents free technetium in the digestive tract. C represents bipedal lymphograms at 24 hours. Iliac nodes are cauliflower-like in appearance and lumbar nodes are barely visualized.

CASE HISTORY

An 18-year old female developed a massive left pleural effusion. Thoracentesis yielded milky fluid with a specific gravity of 1.045, total protein 6.6g/dl, triglycerides 805mg/dl, and cholesterol 107mg/dl, findings consistent with chylothorax. Smears and subsequent cultures of sputum and pleural effusion were negative for bacteria including acid-fast bacilli. Steatorrhea, chyluria, and peripheral edema were absent. Lymphoscintigraphy was performed after subcutaneous injection of 5mCi of ^{99m}Tc -rhenium colloid into the first web space of each foot. Scin-

tigrams taken 15 minutes after injection showed radioactivity bilaterally in the inguinal and iliac regions (Fig. 1A). Sequential follow-up over the chest was done at hourly intervals, and after two hours radioactivity was clearly seen in the left hemithorax (Fig. 1B) verifying leakage of lymph into the pleural cavity. Bipedal lymphography showed normal leg lymphatics but the architecture of iliac nodes bilaterally was distinctly abnormal with diffuse puddling of contrast. Lower inguinal and femoral nodes were unremarkable. After 24 hours, numerous droplet-like particles were visible in the iliac region but lumbar nodes

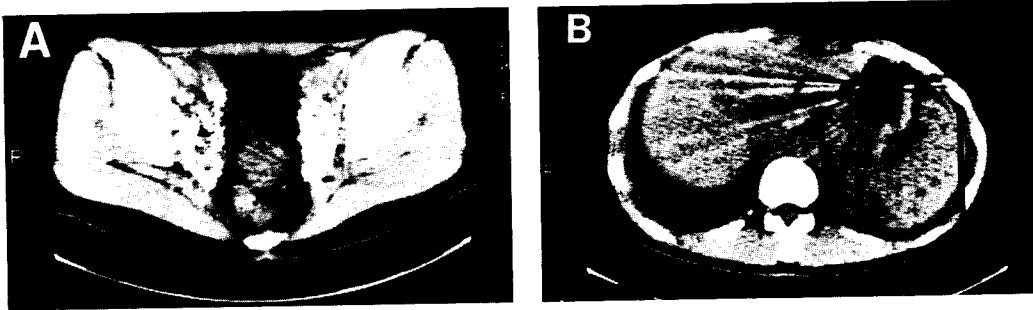


Fig. 2: Computed tomography of the pelvis (A) and upper abdomen (B) showing abnormal parailiac puddling of contrast (A) along with multiple filling defects in the spleen (B) (brackets).

were sparse suggesting diffuse hypoplasia of the retroperitoneal lymphatic system. Some particles in the lumbar area probably represented contrast medium in dilated lymphatics (Fig. 1C). Neither the cisterna chyli and thoracic duct nor leakage of lymph into the left chest was visualized. Thoracotomy failed to clarify the specific site of leakage, and chylous effusion persisted. Abdominal computed tomography postoperatively revealed parailiac dispersion of contrast medium with poorly-defined architecture of adjacent lymph nodes (Fig. 2A). Multiple low-density filling defects were also detected in the spleen (Fig. 2B) consistent with a generalized abnormality of the reticuloendothelial system. No mass obstructing lymphatic channels was found. After placement of the patient on a low-fat diet with medium chain triglycerides and after multiple chemical pleurodeses, chylous effusion gradually decreased. Despite persistence of moderate pleural effusion, she is now asymptomatic on a regular diet.

COMMENT

In infants, chylothorax usually develops in conjunction with congenital abnormalities of the lymphatic system (1-3). In adults, chylothorax typically follows chest trauma (including thoracotomy) or is associated with mediastinal malignancy or infection (4-7). In our adult patient, however, chylous leakage with effusion probably developed from an underlying congenital lymphatic dysplasia. Although con-

trast lymphography failed to detect leakage or the site of extravasation (probably because of clearance by dysplastic lymph nodes and sequestration in ectactic retroperitoneal lymphatics), it did demonstrate structural lymphatic abnormalities. On the other hand, ^{99m}Tc -rhenium colloid lymphoscintigraphy delineated chylous leakage but without further clarifying the etiology. In conjunction with computed tomography these imaging procedures delineated an uncommon cause for adult chylothorax—congenital lymphatic dysplasia.

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