

LYMPHOGENOUS CYST-VEIN SHUNT IN THE MANAGEMENT OF CHYLOTHORAX AND CHYLORRHEA

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ABSTRACT

A 36 year-old woman developed marked lymphedema and chylous cysts of the lower abdominal wall, groin, labia, accompanied by chylorrhea. After cyst excision and transplantation of the greater omentum, a left chylothorax occurred. After thoracic duct ligation and left pleurodesis, pleural effusion recurred and worsened. Lymphangioscintigraphy and conventional lymphography suggested that undrained enlarged retroperitoneal lymphatics in the right iliac fossa had disrupted and lymph had leaked into the left chest from the right iliac fossa. Treatment by a lymphatic cyst-vein anastomosis redirected excess chylous lymph into the blood circulation and chylothorax initially remitted. Several years later with recurrence of chylorrhea, the anastomosis was found to be occluded. After a second operative connection between a lymphogenous cyst and the greater saphenous vein, chylorrhea subsided and chylothorax has remitted for more than 4 years.

CASE REPORT

A 36-year old woman noted that the skin of the lower abdominal wall was becoming gradually thicker with development of a chylous fistula that leaked lymph (approximately 100 ml daily) over 7 to 40 days and would periodically cease spontaneously. At another hospital, the abdominal wall cyst was

excised but post-operatively she developed fulminant chylorrhea (500-1000 ml per 24 hours) and 12 days later the greater omentum was transplanted into the subcutaneous tissue. Chylous drainage initially decreased but then she developed respiratory distress and 1000 ml of chyle was aspirated from the left chest. Thoracentesis for the next week removed approximately 800-1000 ml daily.

At the time of arrival at our hospital, she was in respiratory distress with tachycardia. The skin of the lower abdomen and labia was rubber-like. Chest x-ray confirmed a left pleural effusion with displacement of the mediastinum to the right. Aspiration of the fluid yielded a cell count of $0.64 \times 10^9/L$ (WBC was $0.5 \times 10^9/L$), monocytes were 90%, and the albumin concentration was 19 g/L. No tubercle bacilli or tumor cells were isolated in the pleural effusion. Lymphoscintigraphy showed a lymph-fistula in the right iliac fossa, which drained to the left thorax along retroperitoneal loose connective tissue (*Fig. 1*). Ultrasound showed there was no ascites. Conventional lymphography showed a lymph-cyst (3 cm) in the right iliac fossa.

After thoracotomy and removal of 5400 ml of chyle, many chylous vesicles were seen on the left posterior aspect of the pleura. The thoracic duct was ligated and a left pleurodesis performed. After operation, pleural fluid gradually recurred and thoracotomy drainage averaged 1200 ml/day. After removal of the chest tube, thoracentesis was continued with approximately 850 ml

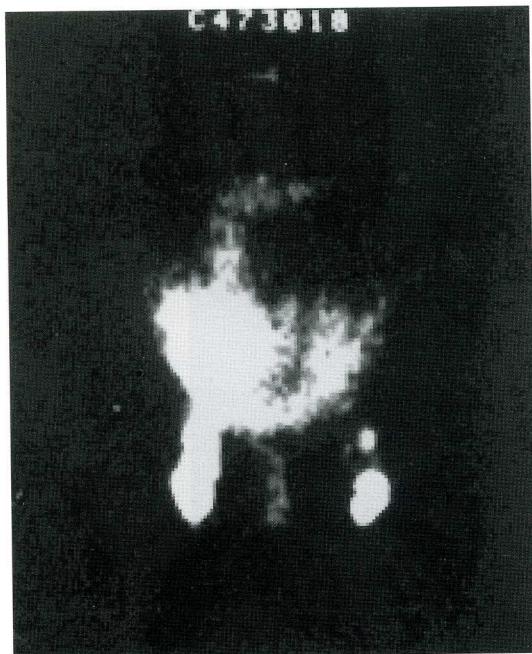


Fig. 1. Thirty minutes after technetium-99 sulfur colloid was injected into the skin of the foot, radiotracer can be seen leaking into the right iliac fossa.

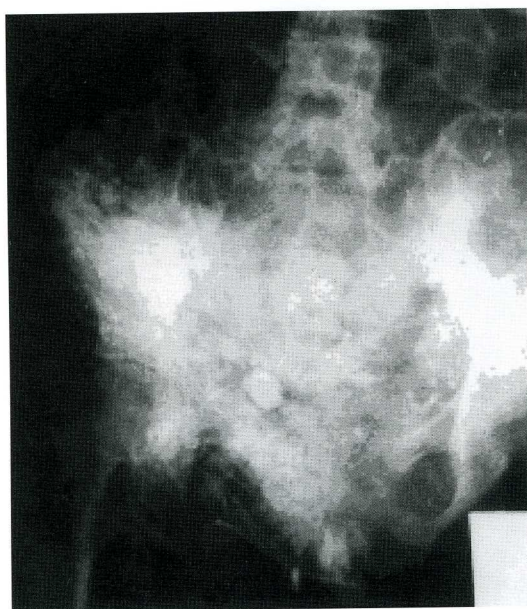


Fig. 2. After 60 ml of 60% meglumine diatrizoate was injected into the chylous cyst, it promptly drained to the abdominal wall and labia. Several smaller cyst collectors also are highlighted.

removed each day. A conventional lymphogram through the right groin was attempted but when the skin was incised, chylous leakage was seen and a 30 ml cyst encountered. Injection of 60 ml of 60% meglumine diatrizoate into the cyst drained to the abdominal wall and labia with display of several cysts (15-25 mm) and enlarged lymphatics (3-4 mm) (Fig. 2). We opted to anastomose the right groin cyst to an adjacent collateral superficial vein (2 mm) and to lymphatics with a nearby vein (1 mm). One month after operation, the pleural effusion remitted. We reinjected 60 ml contrast media into the cyst and 30 minutes later the cyst image disappeared and the urinary bladder was highlighted (Figs. 3, 4). Repeat lymphangioscintigraphy showed radiotracer flowed from the right inguinal fossa to beneath the aortic arch but did not leak into the chest.

Five years later, the right groin cyst suddenly enlarged (11 cm) and chyle began to again drain from the right lower abdominal wall. The cyst-vein anastomosis was occluded. Most of the cyst was reexcised but the residual was anastomosed to the greater saphenous vein. For the last four years, there has been no chylothorax or recurrence of the chylothorax.

DISCUSSION

Primary chylothorax is rare (1). Secondary chylothorax may be caused by transection of the thoracic duct at operation and may require thoracic duct ligation and/or pleurodesis (2). The clinical manifestations of chylous reflux syndrome varies and presenting signs and symptoms may not necessarily reflect the extent of the lymphangiodysplasia. The tenuous nature of lymphatic vessels also increases the difficulty of diagnosis and treatment. Direct lymphography and lymphoscintigraphy are the most useful diagnostic imaging techniques.

Initially we thought that multiple lymph fistulae contributed to the chylothorax and,

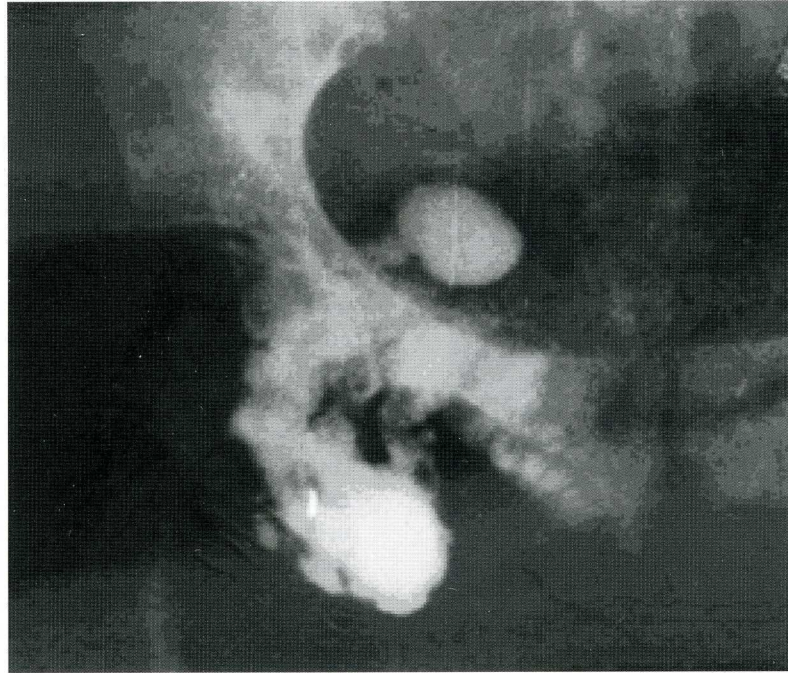


Fig. 3. Five minutes after contrast media was injected into the chylous cyst, the intensity of its image was considerably diminished (see Fig. 4).

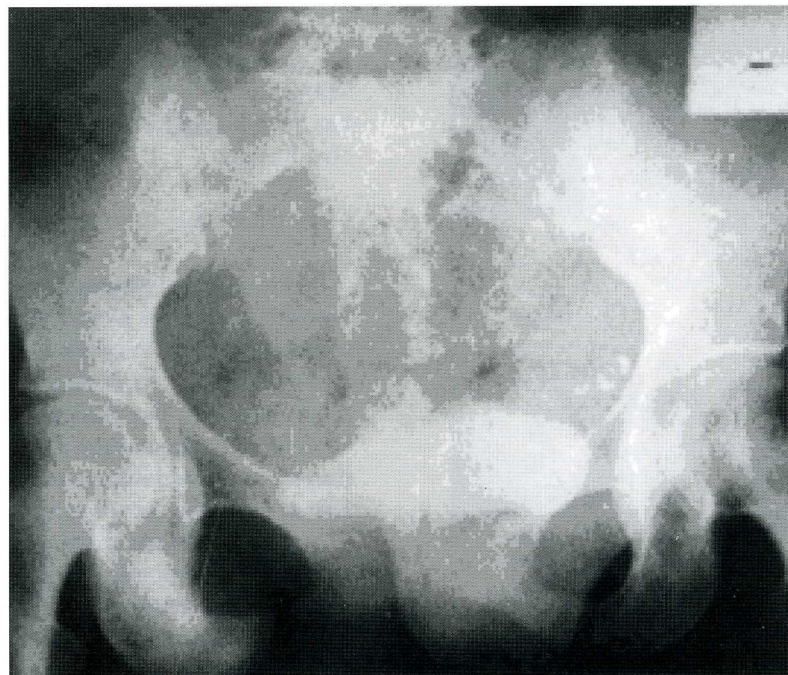


Fig. 4. Thirty minutes after contrast material was injected into the cyst, the cyst image totally disappeared and now the urinary bladder was seen.

accordingly, thoracotomy and thoracic duct ligation with pleurodesis was undertaken. In retrospect, this decision was probably unwise and aggravated ongoing chylous reflux and lymphedema of the lower abdominal wall, groin, and labia with cyst formation and chyloorrhea. After the initial chylous cyst excision, disruption of retroperitoneal lymphangiectasia likely occurred and with prolonged bed rest and negative intrathoracic pressure with respiration, chyle drained from the right iliac fossa to the thorax along loose retroperitoneal connective tissue and spilled into the left chest. The thoracic duct ligation aggravated ongoing chylous reflux and chylothorax. After anastomosis of the right groin lymphogenous cyst to a nearby vein with shunting of chyle into the bloodstream, the chylothorax disappeared. When lymph efflux from the lymphatic fistula was great, it is difficult to image the thoracic duct even by direct lymphography. Conventional lymphography showed retroperitoneal lymphangiectasia and likely valvular insufficiency of retroperitoneal lymphatics.

Lymphangiodyplasia (lymphangiectasis and valve dysfunction) generated chylous reflux, lymphedema, chylous cyst formation, and chyloorrhea. Chylous cyst-to-vein anastomosis initially alleviated the chylothorax but chylous reflux likely persisted. The shunt operation had the same disadvantage as lymphatic-venous anastomosis, namely, occlusion. The second successful cyst to vein anastomosis in this patient showed again the importance of facilitating drainage of stagnant lacteal lymph into the blood vascular compartment.

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