Minimal Lymphatic Leakage in an Infant With Chylothorax Detected by Lymphoscintigraphy SPECT/CT

abstract

A 7-month-old girl with history of persistent left chylous pleural effusion was referred for lymphoscintigraphy. A previous chest computed tomography (CT) scan demonstrated a small to moderate-sized left pleural effusion but could not identify the lymphatic leakage site. Lymphoscintigraphy using filtered ^{99m}Tc sulfur colloid showed minimal focal activity in the lower chest. A correlative single-photon emission computed tomography (SPECT)/CT localized this activity to distal paraesophageal region, being highly suggestive of the site of lymphatic leakage. Subsequent lymphangiography confirmed these findings, revealing an abnormal lymphatic branch at the level of T10 and T11 vertebrae with retrocrural extravasation toward the left hemithorax. Thoracic duct embolization was accomplished at and proximal to the site of chyle leak using a platinum coil and *n*-Butyl cyanoacrylate glue. The patient was followed up for >24 months and demonstrated no recurrence of pleural effusion. No ascites or other complications related to the procedure were noted. The case demonstrates that ^{99m}Tc sulfur colloid lymphoscintigraphy SPECT/CT can be a useful modality for detecting the chyle leakage site in children with chylothorax even when the amount of leakage is minimal. *Pediatrics* 2014;134:e606–e610

AUTHORS: Jigang Yang, MD, PhD,[a,b] Ion Codreanu, MD, PhD,[b,c] and Hongming Zhuang, MD, PhD[b]

[a]Department of Nuclear Medicine, Beijing Friendship Hospital of Capital Medical University, Beijing, China; [b]Department of Radiology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania; and [c]Department of Radiology, Medpark International Hospital, State University of Medicine and Pharmacy Nicolae Testemitanu, Chisinau, Moldova

KEY WORDS

lymphoscintigraphy, chylothorax, SPECT/CT

ABBREVIATIONS

CT—computed tomography
SPECT—single-photon emission computed tomography
TD—thoracic duct
TDE—thoracic duct embolization

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Address correspondence to Ion Codreanu, MD, PhD, Department of Radiology, The Children's Hospital of Philadelphia, 34th and Civic Center Boulevard, Philadelphia, PA 19104. E-mail: codrion@yahoo.com

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Lymphoscintigraphy is an imaging technique used to identify the pathways of lymphatic flow after injection of a radiopharmaceutical that is absorbed by the lymphatics. The technique is nontraumatic with no known adverse effects and is often used for evaluating patients with suspected lymphedema or lymphatic leakage. The method has been refined in recent decades, and advances in SPECT/CT imaging have significantly increased its diagnostic potential.

Chylothorax is a type of pleural effusion resulting from accumulation of lymphatic fluid (chyle) in the pleural cavity. Most often, it is caused by disruption or obstruction of the thoracic duct (TD) or 1 of the main lymphatic vessels that drain to it. The diagnosis is commonly confirmed by the presence of chylomicrons and triglycerides in the aspirated pleural fluid.[1–4] In children, pleural effusion is usually defined as chyle when it contains >1.1 mmol/L triglycerides (with oral fat intake) and has a total cell count ≥1000 cells/μL with a lymphocyte fraction >80%.[1] The condition should be occasionally differentiated from pseudochylothorax (cholesterol pleurisy), which represents long-standing fluid in a fibrotic pleura with a high content of cholesterol but no triglycerides or chylomicrons.[3] Chylothorax is commonly associated with significant morbidity and mortality. If the leaking vessel cannot be identified, this may limit the treatment options to such procedures as drainage of the fluid out of the pleural space and omitting fat from the diet, pneumoperitoneal shunting, or surgical or chemical pleurodesis. Loss of chyle through excessive pleural drainage may result in electrolyte, nutritional, and immunologic complications. In some patients, the effusion may also persist despite optimal medical therapy.[5] Therefore, localizing the leaking vessel becomes of paramount importance, especially in pediatric patients. Herein we report a case of minimal lymphatic leakage in a pediatric patient with persistent chylothorax detected by filtered ^{99m}Tc sulfur colloid lymphoscintigraphy SPECT/CT.

PATIENT PRESENTATION

A 7-month-old girl with history of persistent left chylous pleural effusion and clinical concern for lymphatic leak was referred to the nuclear medicine department for a lymphoscintigraphy study. A previous chest CT scan revealed a small to moderate-size left pleural effusion but was unable to localize the site of lymphatic leakage.

A total of 438 μCi filtered ^{99m}Tc sulfur colloid was injected subcutaneously in divided aliquots into the interdigital web spaces between the great and second toes of each foot. The injection site was covered with a cotton ball and bandage to prevent leakage of tracer through the needle puncture site. Both feet were gently massaged for 2 minutes at the injection sites to promote uptake of the tracer into lymphatic channels and lymphatic flow. Multiple static images of the body taken in multiple projections ≤90 minutes after injection using a Forte Nuclear Medicine (Royal Philips, Amsterdam, Netherlands) γ camera proved unremarkable. Subsequent delayed images were obtained ~7.5 hours after injection, supplemented by SPECT/CT images of the upper abdomen and thorax. Both subclavian veins and cardiac silhouette could be clearly identified on these planar delayed images, confirming flow of the radiotracer from the TD into the vasculature and cardiac blood pool. A suspicious focus of minimal tracer activity in the left lower chest was also noted on delayed planar images (Fig 1), localizing to distal paraesophageal region on SPECT/CT images (Fig 2). The SPECT/CT findings were particularly suggestive of the site of lymphatic leakage.

After consultation with interventional radiology, a team decision was made to perform an ultrasound-guided intranodal lymphangiography followed by thoracic duct embolization (TDE). The procedure was performed under general anesthesia. The lymph nodes in each groin were punctured using 25-gauge needles under ultrasound guidance. When test injections of contrast showed satisfactory position and opacification of efferent cephalad lymphatic vessels,

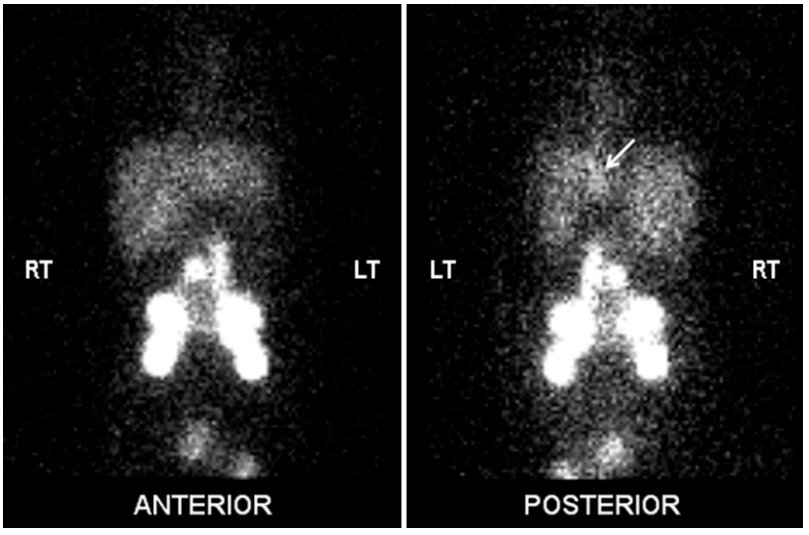

FIGURE 1
Planar lymphoscintigraphy revealing minimal focal activity in the lower chest, slightly more prominent on posterior view (arrow). RT, right; LT, left.

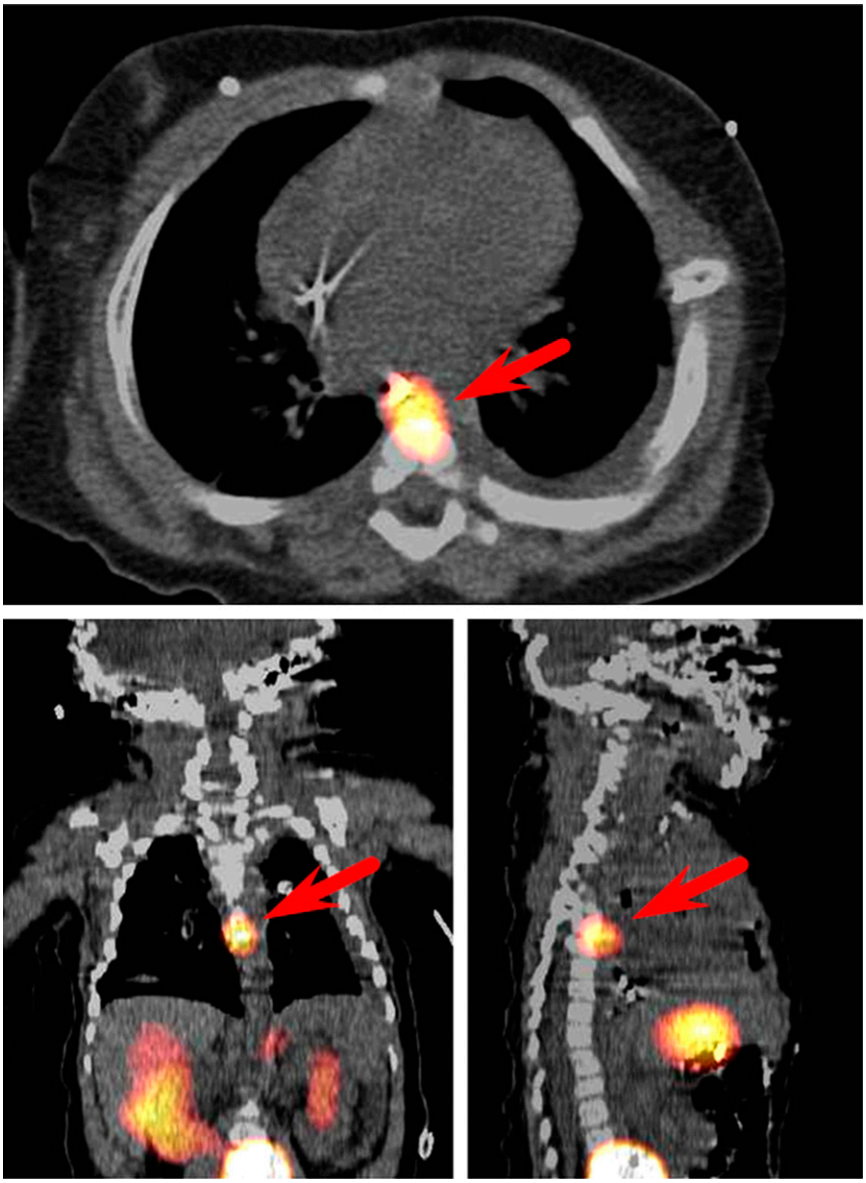

FIGURE 2
Associated SPECT/CT images (axial, coronal, and sagittal views) localizing the abnormal focal activity to the distal paraesophageal region (arrows).

bilateral injections of approximately 2 mL ethiodized poppyseed oil (Ethiodol; Savage Laboratories, Melville, NY) were made under fluoroscopic guidance, followed by 2 mL saline. As the lumbar trunks began to opacify the cisterna chyli, a 22-gauge Chiba needle (Cook Medical, Bloomington, IN) was used under fluoroscopic guidance to puncture the lower cisterna percutaneously from an anterior transabdominal approach. A stiff guidewire (V18 Control; Boston Scientific, Natick, MA) was advanced into the cisterna chyli and manipulated cephalad into the TD. Over the wire, a 2.3F rapid transit microcatheter was placed farther into the TD. Contrast injection of the TD demonstrated a leftward abnormal lymphatic branch at the level of T10 to T11, with retrocrural extravasation of contrast toward the left basal hemithorax (Fig 3A). Several unsuccessful attempts were made to selectively enter this channel using a variety of guidewires and a rapid transit microcatheter. Accordingly, a decision was made to embolize the TD at and below this point to stop the leak. One 0.5-cm straight platinum coil was placed (Fig 3B), followed by injection of 0.5 mL *n*-Butyl cyanoacrylate (Trufill; Cordis Corporation, Warren, NJ) diluted 1:1 in Ethiodol. The microcatheter was immediately removed. The lower TD, the leaking branch, and the cisterna chyli down to the entrances of the right and left lumbar trunks were occluded. The patient was followed up for 24 months and demonstrated no recurrence of pleural effusion. No ascites or other complications related to the procedure were noted.

DISCUSSION

Chylothorax has been reported as the most common form of pleural effusion in the first few days of life, and understanding its pathophysiology may assist in clinical decision-making.[4,6] Potential causes of chylothorax in children can vary and have been previously grouped into such categories as congenital, traumatic, related to high central venous pressure, and malignancies as well as miscellaneous causes such as benign tumors, granulomatous disease, and transdiaphragmatic movement of chylous ascites.[4,7–13] Tumors have been reported to account for >50% of all cases of chylothorax, and traumas are the second major cause.[7] Therefore, patients presenting with nontraumatic or idiopathic chylothorax should undergo an appropriate workup to exclude a neoplastic etiology, especially lymphomas. Traumatic events resulting in chylothorax are often obvious, even though minor traumas affecting intrathoracic pressure such as severe coughing or vomiting, sudden stretching, and hyperextension injuries have also been reported as precipitating factors.[7] Because no other obvious causes for chylothorax were revealed in our patient apart from an abnormal lymphatic branch at the level of T10 to T11

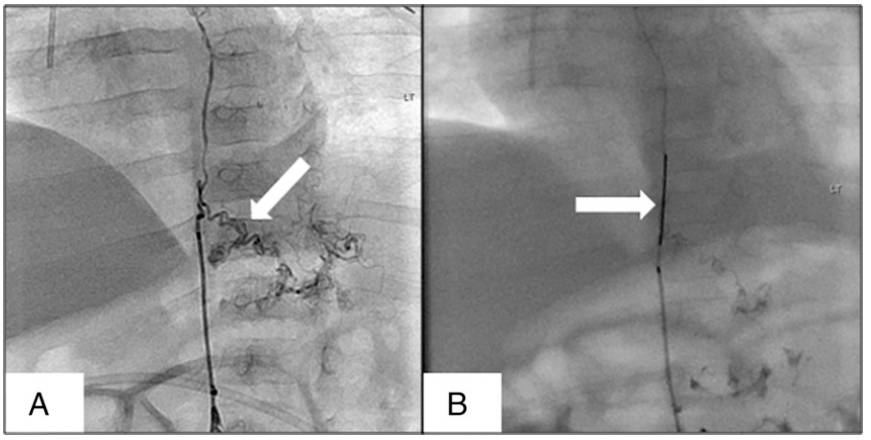

FIGURE 3

A, Follow-up lymphangiography showing a leftward abnormal lymphatic branch at the level of T10 to T11 vertebrae with retrocrural extravasation of contrast in that region (arrow). B, Embolization of the TD was undertaken using a platinum coil (arrow) with subsequent administration of 0.5 mL of radiopaque cyanoacrylate glue.

vertebrae, such a minor injury accompanied by variations of intrathoracic pressure may have precipitated the vessel rupture in this case.

Other potential causes of chylothorax in children include congenital lymphatic dysplasias, which may be encountered as separate entities or be part of associated conditions such as hydrops fetalis or Down, Noonan, or Turner syndrome.[4,11] Of note is that peripheral lymphedema may not be always clinically evident at birth or during the early years of life, despite the presence of severe visceral lymphatic impairment.[14] When such patients present with chylothorax, chylous pericardial effusion, or chylous ascites, lymphoscintigraphy may show various degrees of lymphatic impairment, providing valuable diagnostic clues.[14] Venous thrombosis of the superior vena cava or adjacent portions of subclavian veins may also precipitate TD rupture caused by significantly elevated central venous pressure and should be included in the differential diagnosis in patients with central venous catheters, recent thoracic surgery, or hypercoagulable states.[4,8,12,13] When investigating pediatric patients, one should also remember that chylothorax in a child may be a manifestation of child abuse.[15–17] A radionuclide bone scan to detect potential skeletal injuries may prove essential for reaching the diagnosis in such patients and should be considered among the requested investigations when child abuse is suspected.

The initial diagnosis of chylothorax is usually made by pleural fluid analysis, regardless of its etiology. Subsequent investigations are directed at identifying the site of chyle leakage and the underlying cause, which may become essential for guiding the therapy in patients with persistent or recurrent disease. Lymphangiography and lymphoscintigraphy are 2 specific methods for imaging lymphatic system that may aid in finding the chyle leakage site. Lymphangiography enables radiographic visualization of lymph vessels and nodes after injection of a contrast medium. Although its utility in the diagnosis of lymph node pathology has decreased with recent advances in CT and MRI, the modality still has an important role in the diagnosis of various lymphatic pathology, including identifying the site of chyle leak or obstruction. Its use in children is also limited by the ability to cannulate the tiny lymphatic vessels in these patients. Lymphoscintigraphy, on the other hand, is a faster and less traumatic procedure requiring just an intradermal or subcutaneous injection. Its main disadvantage, the lack of anatomic correlation, has been overcome by recent advances in SPECT/CT imaging. Recent studies have already reported the usefulness of lymphoscintigraphy SPECT/CT for identifying the site of lymphatic leakage in adult patients with TD injuries.[18,19] Our case suggests that lymphoscintigraphy SPECT/CT is also suitable in pediatric patients presenting with chylothorax even when the amount of chyle leakage is minimal. In this neonate, the technique detected the site of lymphatic leakage, guiding the intranodal lymphangiography followed by TDE. It is believed that after the TD is occluded, new lymphovenous communications develop and "dormant" anastomoses reopen.[20] Thus, TDEs performed at higher levels allow a greater number of lymphovenous communications and better collateral circulation. In our patient, the site of the lymphatic leak was low, and every attempt was made to selectively embolize only the leaking branch. However, repeated attempts to enter this tiny vessel proved unsuccessful, and a decision was made to embolize the TD at and below this point to stop the leak. Despite a higher risk of subsequent complications, low-level embolizations including cisterna chyli have been described in the literature.[21] Furthermore, successful TDEs at this level have been previously performed in our center in neonates.[22] In this neonate, however, a lymphoscintigraphy SPECT/CT was requested for the first time. We assume this might have been related to the tiny caliber of the leaking vessel, making it undetectable by other modalities and its subsequent cannulation impossible. The case also demonstrates that ^{99m}Tc sulfur colloid lymphoscintigraphy SPECT/CT can be a useful modality for detecting the chyle leakage site in children with chylothorax even when the amount of leakage is minimal.

MR lymphography with intracutaneous and subcutaneous administration of various lymphotropic paramagnetic contrast agents is another emerging modality that may open new perspectives for imaging lymphatic system, including in patients with chylothorax.[23,24] However, most of these lymphotropic contrast agents are still in the preclinical phase or under development, and validation in larger studies is needed before their routine use in clinical practice.[24]

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