

Life-Threatening Anomalies of the Thoracic Duct: Anatomic Delineation Dictates Management

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Congenital anomalies of the thoracic duct are rare, poorly characterized, and difficult to manage. The spectrum of pathophysiologic perturbations, presenting symptoms, radiographic findings, and interventions performed in 4 patients are shown. Accurate anatomic delineation of the malformation was only possible by direct injection contrast lymphangiography. Therapies tailored to address the anatomic aberrations included intralesional sclerotherapy, surgical excision and ligation, lymphovenous anastomosis, and omental interposition to interrupt dysfunctional collateral

lymphatics to the lung. Accurate anatomic diagnosis of central lymphatic channel anomalies by contrast lymphangiography facilitates an individualized multidisciplinary approach to repair.

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CONGENITAL ANOMALIES of the thoracic duct are extremely rare, poorly characterized, and difficult to manage. The functional, rheologic, and hemodynamic properties of the central lymphatics have not been well elucidated. Some clinical manifestations of central lymphatic dysfunction such as chylous pleural effusions and chylous ascites often are considered difficult or impossible to eradicate. Imprecise nonanatomic therapies such as pleurodesis, prosthetic pleuro-peritoneal or peritoneovenous shunts, or even radiation therapy often are used.

The lymphatics of the lower extremities and lower torso join with those of the bowel mesentery in the cisterna chyli, which lies in the lumbar prevertebral plane behind the aorta and inferior vena cava. Confluent with the cisterna chyli, the thoracic duct ascends in the thoracic prevertebral plane, behind the esophagus and aorta. The lymphatic drainage of the mediastinum, upper torso, arms, head, and neck join the central lymphatic channels via thoracic tributaries. The thoracic duct proper empties into the central venous system near the junction of the left internal jugular vein and subclavian vein. The accessory thoracic duct empties in a similar fashion on the right side.

Because of their extremely narrow caliber, the central lymphatic channels are difficult to anatomically delineate either by radiographic studies or surgical dissection. Although lymphangiography was performed commonly in earlier eras for identifying nodal metastases from malignancies, modern cross-sectional imaging techniques such as computed axial tomography and magnetic resonance imaging (MRI) have relegated contrast lymphangiography to a nearly forgotten technique. Nuclear scintigraphic lymphangiography, although of some util-

ity, provides far less anatomic detail than contrast injection lymphangiography. By example of 4 unique patients, we show the utility of contrast lymphangiography in conjunction with more modern imaging techniques in delineating anatomic malformations of the central lymphatics.

CASE REPORTS

Case 1

A previously healthy 8-year-old boy had a spontaneous right chylothorax that failed to resolve with 1 month of bowel rest and total parenteral nutrition. He underwent right thoracotomy and pleurodesis in Argentina, which successfully abated his effusion. However, 9 months later, severe chylous ascites developed. Explorations by both laparotomy and lumbotomy failed to identify and control the chyle leak. A peritoneo-venous shunt successfully controlled the ascites for 6 months until mechanical failure occurred. Subsequent shunts were nonfunctional. Repeated paracenteses then were performed at approximately 10-day intervals.

On arrival for consultation, now at age 11, he was profoundly cachectic with massive ascites. Computed tomography (CT) scan showed a right retroperitoneal mass consistent with lymphatic malformation. Contrast lymphangiography with injection via inguinal lymphatic cannulation showed reflux of contrast from the cisterna chyli via pericaval and segmental lymphatics through the pararenal mass and into the free peritoneal cavity (Fig 1). Laparotomy with resection of the mass and surrounding lymphatics, including stripping of the renal

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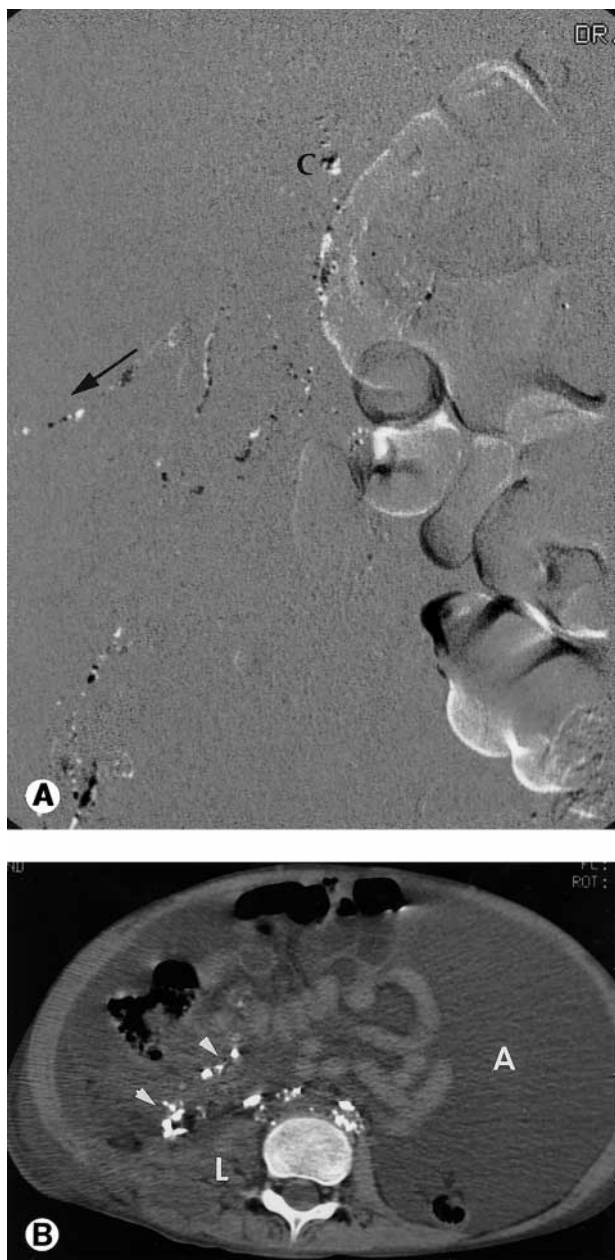


Fig 1. Eight-year-old boy with chylous ascites after thoracic duct ligation. (A) Digital subtraction image from a lymphangiogram shows contrast medium from the cisterna chyli (C) refluxing through pericaval and peri-iliac channels into the peritoneal cavity (arrow). (B) Axial CT image of the abdomen after lymphangiography shows the chylous ascites (A) with a lymphatic malformation involving the right psoas area (L) and the contrast leak through a lymphatic malformation out of the retroperitoneum (arrowheads).

capsule, as well as ligation of pericaval lymphatics and application of fibrin glue was successful at completely eradicating his ascites.

Case 2

A previously healthy 15-year-old boy had a massive spontaneous right chylothorax. Despite 2 thoracotomies for thoracic duct ligation, multiple thoracostomy tubes, pleurodesis, and 3 months of total par-

enteral nutrition, his chylothorax persisted. Drainage ceased after radiation therapy to the chest, mediastinum, left subclavian area, and central retroperitoneum down to the lower lumbar level. He remained well for the ensuing 6 years until cough, fever, and hemoptysis developed. Chest radiograph showed bilateral pleural infiltrates. Trans-bronchial biopsy confirmed cryptococcal pneumonia. Despite clearance of the cryptococcus, he had persistent and progressive respiratory decompensation. Persistent lymphopenia also was seen. At age 22, he experienced an episode of pneumoperitoneum. His abdominal examination was otherwise unremarkable. Rather, the air under his diaphragm was presumed to have tracked down from his lungs, because he could not accumulate a pneumothorax due to his prior pleurodesis. He also had chylous ascites at this time.

He persisted in a chronically debilitated state with respiratory insufficiency, increased work of breathing, oxygen dependence, and cachexia. Most interestingly, he had a persistent cough productive of several ounces of white sputum daily. Cross-sectional imaging showed no mass lesions in the abdomen or chest but did show ascites and significant pulmonary consolidation. Contrast lymphangiography followed by sequential CT scans showed contrast rising in the central retroperitoneal lymphatics to the clips on the thoracic duct at the level of the diaphragm. Contrast then diverted laterally within the diaphragmatic lymphatics and then directly upward into the pulmonary parenchymal lymphatics and the alveoli and bronchi (Fig 2). Thus, as a result



Fig 2. Twenty-year-old man with deteriorating pulmonary function secondary to chylobronchial fistula. (A) Axial CT scan at the level of the diaphragm after lymphangiography shows collateral lymphatic channels extending from the pericaval and periaortic lymphatics into the right and left hemidiaphragm. (B) Axial CT scan above the lung shows extension of the lymphangiographic contrast medium into the central part of the right lung (arrow).

of his prior thoracic duct ligation, pleurodesis and radiation therapy, he formed aberrant collateral lymphatic vessels which directed chyle from his abdominal lymphatics directly into his pulmonary lymphatics and airways, resulting in chylobronchorrhea. Flexible fiberoptic bronchoscopy showed white fluid, proven biochemically to be chyle emanating from his lower lobe bronchi.

To reverse some of his chronic pulmonary failure by diverting the chyle from entering his right lung, where the majority of aberrant vessels were directed, these collaterals were divided. This was accomplished by a reoperative right thoracotomy through the irradiated, postpleurodesis field. With great difficulty and significant bleeding, the lower lobe was mobilized from its dense union with the diaphragm and pericardium. To prevent reformation of collateral lymphatics from the diaphragm into the lung, the omentum was mobilized and transposed on a right gastroepiploic pedicle up through the diaphragm and interposed between the lung and the diaphragm. Despite several months of preoperative preparation with bowel rest and total parenteral nutrition to both minimize chyle flow into the lungs and maximize nutritional status, this operative procedure resulted in a prolonged postoperative critical care phase. Six months of mechanical ventilation via tracheostomy was necessary. However, his pulmonary function eventually recovered resulting in improved pulmonary function over his preoperative status. His cough and chylobronchorrhea was eliminated. Nutritional and immunologic parameters also improved toward normal. Not surprisingly, his chylous ascites increased requiring treatment with paracenteses and peritoneovenous shunting. Subsequent lymphangiography has shown successful fusion of his diaphragmatic lymphatics to his omental lymphatics with absence of any contrast in the lung parenchyma.

Case 3

A boy in early infancy had a progressive cutaneous purple lesion on his back and neck. Biopsy result was inconclusive but suggested a vascular etiology. As the lesion expanded to encompass the entire back, a ballotable fluid collection in the subcutaneous tissues of the back and neck developed. He eventually manifested a large left pleural effusion. Pleurocentesis yielded pure white chyle. The fluid in the subcutaneous tissues also was chyle. Nuclear lymphangiography and cross-sectional imaging studies failed to explain these clinical findings. A contrast lymphangiogram showed a normal caliber and course of the thoracic duct. However, no contrast was seen to pass from the thoracic duct into the vascular system. On the contrary, the contrast was seen to drop from the end of the thoracic duct behind the left clavicle and down into the left pleural cavity (Fig 3).

At 13 months of age surgical exploration of the left supraclavicular fossa while infusing cream into the duodenum showed a disjointed thoracic duct pouring chyle into the soft tissues. A primary end-to-end lymphovenous anastomosis was performed from the thoracic duct to a valved periesophageal vein. This resulted in dramatic diminution of the chylothorax and subcutaneous chyle accumulation. A repeat lymphangiogram 4 months postoperatively showed the lymphovenous anastomosis to be patent and functioning. Nonoperated anomalous torso lymphatics remained. Thirty-five months postoperatively, the child continues to do well from a respiratory standpoint. He has had several episodes of sclerotherapy of his subcutaneous tissues to treat slowly accumulating chyle. He also had undergone a partial tenth rib resection and ligation of aberrant lymphatics emanating from the region of the cisterna chyli into the paraspinal soft tissues to diminish the subcutaneous chyle accumulation. He has required no further pleurocenteses.

Case 4

A 15-year-old boy with severe Klippel-Trenaunay syndrome, with lifelong difficulties caused by combined capillary lymphaticovenous



Fig 3. Digital subtraction image of the thorax during lymphangiography shows oily contrast medium moving up the thoracic duct to the left supraclavicular region (arrow). After this, droplets of contrast medium could be seen within the left chest (arrowheads).

malformations of his right lower extremity, pelvis, genitalia, and retroperitoneum presented with progressive respiratory insufficiency. He had severe orthopnea, requiring him to sleep upright. He had missed an entire year of school because of his respiratory failure. Pulmonary function findings showed a severe obstructive pattern involving the small airways. He had previously undergone multiple surgical debulking procedures and percutaneous sclerotherapy sessions of various areas of his malformations. His respiratory failure had been unexplained, but he was noted to have extension of his retroperitoneal lymphatic malformation up into the posterior mediastinum and paraspinous regions. Direct right paraspinous percutaneous contrast injection into this component of the malformation showed a multitude of serpiginous lymphatic channels, similar to that seen in other areas of his body. However, on one contrast injection, a large pooling effect was noted. Further injection showed a long tubular structure extending the length of the thorax, which proved to be a massively enlarged thoracic duct (Fig 4). The duct was approximately 4 cm in diameter. It was shown to communicate with the left subclavian vein in the appropriate location. There was some reflux seen into segmental and paraspinous channels. The mass of serpiginous lymphatics adjacent to this thoracic duct were sclerosed with 3% tetracycline. No sclerotherapy was performed directly into the thoracic duct. After sclerotherapy, the patient experienced dramatic improvement of his respiratory symptoms and pulmonary function tests. His orthopnea was relieved. It was felt that his prior pulmonary symptoms likely resulted from interstitial pulmonary lymphatic hypertension caused by chylo-lymphatic reflux from a dysfunctional, poorly emptying thoracic duct. His respiratory improvement has persisted for 42 months as of this writing.

DISCUSSION

Vascular anomalies in general often are poorly understood and difficult to differentiate.^{1,2} More specifically, anomalies of the thoracic duct are particularly rare and difficult to manage. Traumatic thoracic duct injuries are not infrequent after cervical or thoracic surgery or



Fig 4. Fifteen-year-old boy with the diagnosis of Klippel-Trenaunay syndrome and severe orthopnea. CT imaging had shown posterior mediastinal mass. Direct contrast injection after percutaneous cannulation of the posterior mediastinal malformation confirms the presence of a markedly dilated thoracic duct (TD), with peristalsis and communication with the subclavian vein (V; arrow).

trauma and often will heal spontaneously or respond to simple ligation.³ In contrast, congenital anomalies of the central lymphatics often present with seemingly insoluble clinical scenarios. As shown by the patients in this series, their multifaceted manifestations can include respiratory compromise, chylothorax, chylous ascites, malnutrition and hypoproteinemia, lymphopenia and immunosuppression, and bony erosion. Although patients presenting clinically with combinations of these clinical findings often are recognized to have abnormalities of

lymphatic function, the specific etiology usually is not determined. Chyle normally is only present in the mesenteric lymphatics, cisterna chyli, and thoracic duct. Chyle in any other body cavity or tissue implies leakage from the central lymphatic channels. Chronic management with diuretics, pleurocenteses, peritoneocenteses, shunting procedures, pleurodeses, and parenteral nutrition may provide short-term palliation, but rarely result in long-term improvement in quality of life and survival. These therapies often are temporizing and nonspecific.

We believe that anatomic delineation of each patient's underlying anomaly is crucial to designing directed and more definitive therapy. As shown by the cases presented, this may require direct contrast lymphangiography. Lymphangiography is not a new technique. It was used through the 1970s to study nonmalignant lymphatic disorders⁴⁻⁶; however, its more prevalent indication was for the identification of malignant nodal metastases. With the advent of sophisticated cross-sectional imaging techniques such as CT and MRI scanning, lymphangiography fell out of favor. Thus, experience with this technique in recent years is extremely limited. Although nearly a lost art, lymphangiography remains the best technique to delineate the anatomy of aberrant lymphatic channels. More technologically advanced imaging techniques have not been successful in this regard. The disappearance of lymphangiography from the routine arsenal of diagnostic imaging studies over the last several decades has resulted in a lack of attention to the understanding of lymphatic channel disorders. Although the performance of a contrast lymphangiogram requires tedious and meticulous dissection and a large time commitment, unfortunately it appears to remain the only method by which some central lymphatic anomalies can be seen. In each of the cases presented in this series, anatomic explanations for a patient's symptoms were identified that otherwise would not have been predicted. The surgical and radiologic interventions performed successfully would not have been possible in the absence of the lymphangiographic road maps. Until a method is devised by which radiopaque contrast can be taken up by lymphatics, direct cut-down contrast lymphangiography remains an essential technique. Patients with symptoms suggestive of thoracic duct or cisterna chyli anomalies should be considered for lymphangiography before progression to debilitating disease states.

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