

Congenital Chylothorax

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Abstract

Congenital chylothorax should be considered in neonates with symptoms and imaging consistent with pleural effusion, but analysis of the pleural effusion is necessary to confirm chylothorax. Lymphangiogram can identify a leak or highlight a structural lymphatic anomaly and is useful in guiding therapy. However, this case adds to the recent evidence in the literature that lymphangiography alone can resolve a lymphatic leak. Further research should be done on the efficacy of therapeutic lymphangiography to further elucidate its role in congenital chylothorax management.

Keywords: lymphatic, congenital

Case Summary

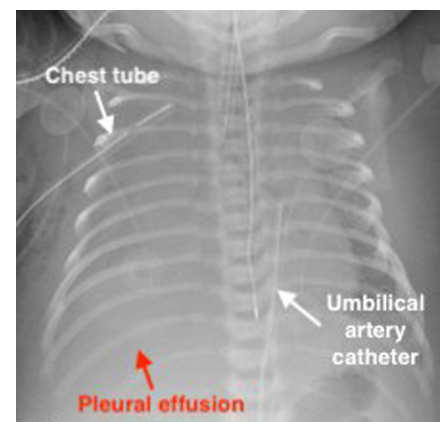
An infant with horseshoe kidney and hydrops fetalis was born at 37 weeks' gestation and required intubation and chest compressions at birth. Chest x-ray confirmed the presence of a pleural effusion. A right chest tube was placed and drained a chylous effusion with a triglyceride level of 165 mg/dL and a lymphocyte count of 88%. Total parenteral nutrition therapy was initiated, and chest tube output increased with each feeding. Sirolimus therapy failed to improve the effusion. At 44 days of age, intranodal lymphangiography was performed with an access in the left inguinal lymph node using 0.9 mL oily contrast (Lipiodol, Guerbet, Princeton, New Jersey) and revealed contrast leaking into the bilateral peribronchovascular lymphatic channels consistent with a lymphatic channel anomaly. Following lymphangiography, serial chest x-rays did not demonstrate reaccumulation of pleural effusion. The patient's chest tube was removed 4 days

later, and the infant was weaned off diuretic medication. Genetic testing was performed, but no genetic mutations associated with congenital chylothorax were identified.

Imaging Findings

Chest x-ray performed at 41 days old revealed opacities primarily in the right lung, consistent with right-sided pleural effusion (Figure 1). Intranodal lymphangiography done at 44 days of age revealed normal iliac and lumbar lymphatics, cisterna chyli, and thoracic duct. Congestion was found at the left supraclavicular lymphatic channel with no contrast passage into the subclavian vein as well as reflux of contrast into the peribronchovascular lymphatics bilaterally. These findings are consistent with a central conducting lymphatic anomaly (CCLA) and chylothorax due to leakage of chyle into the pleural spaces (Figure 2). Chest x-ray performed 60 days after lymphangiography revealed

Figure 1. Frontal chest x-ray when the patient was 2 days old demonstrates near complete opacification of the right hemithorax. This finding is consistent with pleural effusion.



normal pleural spaces and resolution of chylothorax (Figure 3).

Diagnosis

Chylothorax caused by CCLA.

Discussion

Chylothorax is a collection of lymphatic fluid in the pleural spaces and is the most common cause of congenital pleural effusion in neonates. Congenital chylothorax may present with lung hypoplasia and pulmonary hypertension, complicating cardiorespiratory function as well as the development of hydrops fetalis.

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Figure 2. Lymphangiography when the patient was 44 days old demonstrates lymphatic congestion at the left supraclavicular space (short red arrow), as well as bilateral peribronchovascular leakage (long red arrow). This is consistent with congenital conducting lymphatic anomaly. Filling of the lumbar (short white arrow) and inguinal lymphatics (long white arrow) is also noted.

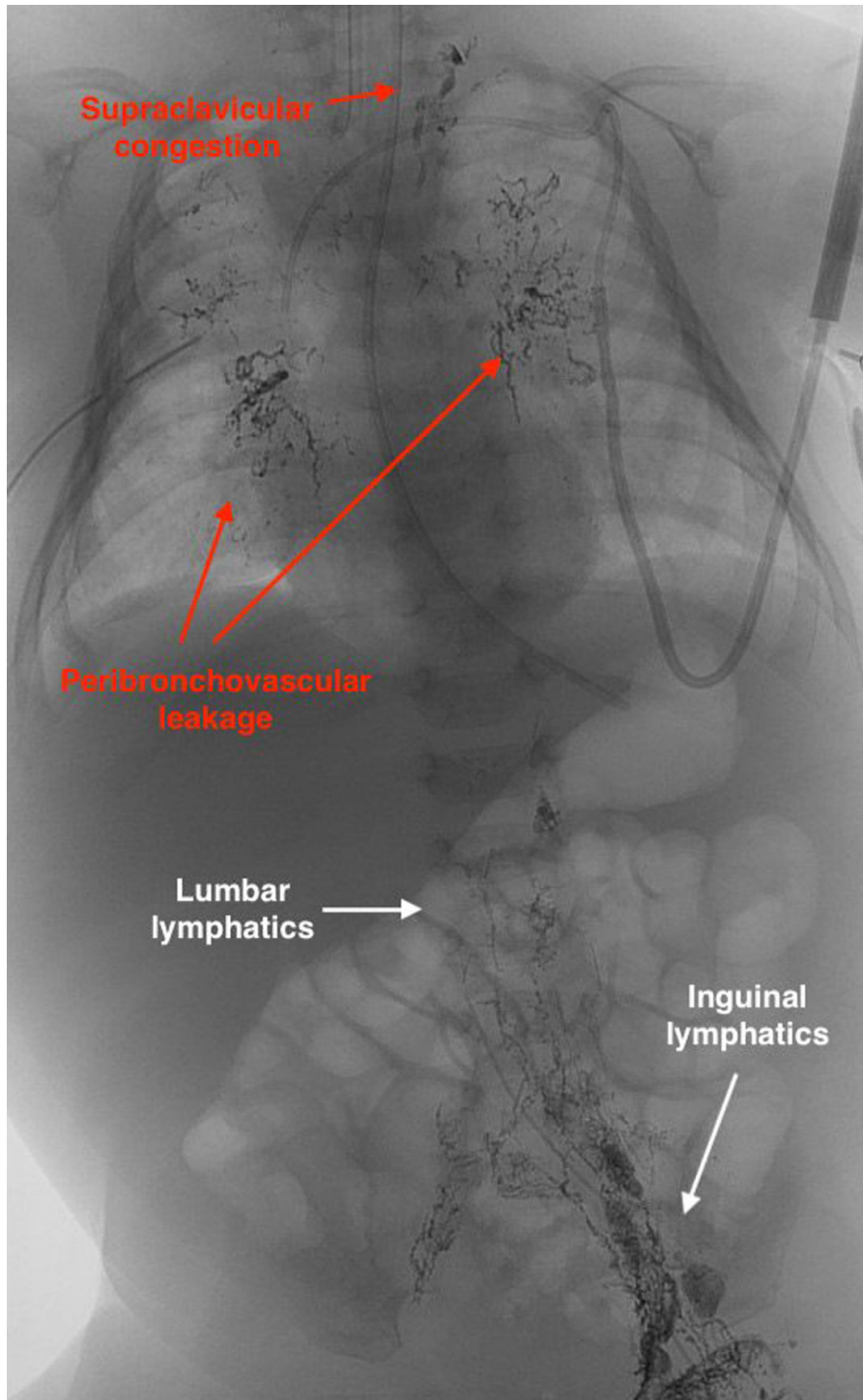
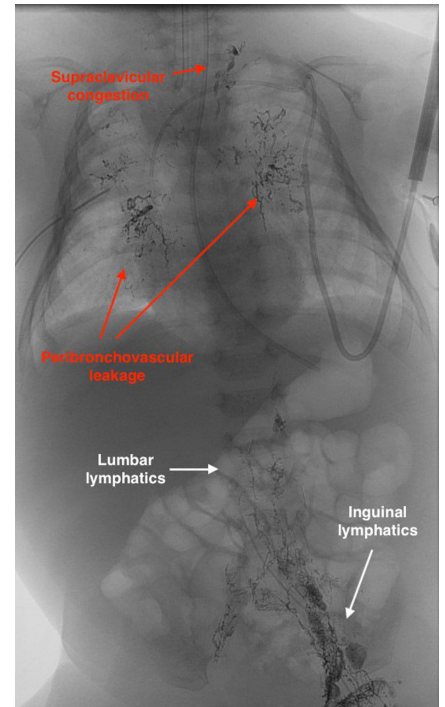


Figure 3. Normal chest x-ray done post-lymphangiography when the patient was 3 months old, representing resolution of the chylothorax. Residual lipiodol is seen in the mediastinum and cervical lymph nodes bilaterally.



Malnutrition and immune compromise can also manifest because of protein and cell leakage into the pleural space.^{1,2} Postnatal development may be more insidious, with gradual worsening of dyspnea, cough, and chest discomfort.² Spontaneous regression of congenital chylothorax has been reported in 22% of cases, while severe conditions associated with hydrops may lead to high morbidity and mortality.¹ Prenatally, US can diagnose pleural effusion while chest imaging is used postnatally.^{1,2} Thoracentesis or chest tube placement can extract the pleural fluid for analysis and identification of the effusion. Chylous effusion is milky white, and fluid analysis reveals triglyceride levels > 110 mg/dL, a pleural fluid to serum cholesterol ratio of <1.0, and evidence of chylomicrons.²

Chylothorax can be caused by traumatic or iatrogenic injury of the thoracic duct, be a complication of malignancies such as lymphoma, and be associated with genetic syndromes and other congenital causes.² CCLA is defined by abnormalities in the thoracic duct that impede lymphatic flow into the subclavian vein, resulting in retrograde reflux and leakage of lymphatic fluid into the pleural space.³ The causes of CCLA are poorly understood, though some genetic conditions like the RASopathies have been associated with congenital chylothorax.^{2,4} Genetic syndromes are suspected when multiple embryological defects exist such as in this case of horseshoe kidney and CCLA, but no characteristic genetic mutations were identified in this patient.

Treatment of chylothorax often relies on conservative management. Octreotide reduces lymphatic flow via vasoconstriction and reduced fat absorption and has been shown to be effective in 47% of cases.^{1,5} Nutritional management consists of a fat-free diet with medium-chain triglyceride supplementation (MCT) because long-chain triglycerides (LCTs) are absorbed into the lymphatic system while MCTs get directly absorbed into the systemic circulation. Increasing the ratio of MCT to LCT therefore reduces the amount of lymph available for leakage into the pleural space and gives time for the leak to spontaneously close.^{1,2} Sirolimus has also been shown to be effective in treating various lymphatic malformations and one case of CCLA specifically, though its role in chylothorax management is not clearly defined.^{6,7} Cases refractory to these interventions may require surgery or lymphatic interventions such as thoracic duct embolization.⁸ In this patient, chylothorax returned when LCTs were reintroduced and sirolimus was not shown to mitigate the effusion.

Lymphangiography is commonly performed to visualize the lymphatic system to determine the etiology of the effusion and guide treatment. A contrast agent, such as Lipiodol, is injected into an inguinal lymph node and imaged via fluoroscopy, tracking the passage of Lipiodol through the thoracic duct and into the left subclavian vein.⁸ While lymphangiography is most often done for visualization prior to a therapeutic procedure, several cases of resolution of chylothorax after lymphangiography alone have been reported.⁹ Similarly, lymphangiography alone has been found to resolve other lymphatic leaks such as chylous ascites, where, similarly to chylothorax, lymphatic fluid leaks into the abdominal cavity.¹⁰

In congenital chylothorax specifically, one case report describes resolution of congenital chylothorax after lymphangiography in 2 cases of RASopathies in an 8-year-old with cardiofaciocutaneous syndrome and a 2-month-old patient with Noonan syndrome.⁴ The case presented here represents additional evidence of lymphangiography resolving congenital chylothorax and supports therapeutic lymphangiography as a potential treatment modality for medically refractory congenital chylothorax.

Conclusion

Congenital chylothorax should be considered in neonates with symptoms and imaging consistent with pleural effusion, but analysis of the pleural effusion is necessary to confirm chylothorax. Lymphangiogram can identify a leak or highlight a structural lymphatic anomaly and is useful in guiding therapy. However, this case adds to the recent evidence in the

literature that lymphangiography alone can resolve a lymphatic leak. Further research should be done on the efficacy of therapeutic lymphangiography to further elucidate its role in congenital chylothorax management.

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