Chylothorax

37

Dakshesh Parikh

Abstract

Congenital, idiopathic, pathological and iatrogenic chylothorax is encountered across all age groups. The aetiology and pathogenesis of the congenital chylothorax is unknown. Some congenital malformations and pathologies are known to be associated with chylothorax. Congenital diaphragmatic hernia after its correction can present with chylothorax as persisting and challenging problem. Thoracic chylous lymphatics can be accidentally injured during surgery or can leak after a blockage from an obstructing tumour or venous thrombosis of superior vena cava. Specific investigations to identify its causative aetiology are indicated in adult chylothorax. In infants and children, imaging is carried out after initial supportive measures have failed to resolve chylothorax. Respiratory compromise as a result of pleural collection of chyle may require drainage and ventilatory support. Chyle production can be reduced by withholding enteral nutrition and providing total parenteral nutrition. Surgical approach is considered in prolonged and intractable cases. Outcome can be variable depending on its aetiology, associated genetic and congenital malformations.

Keywords

Chylothorax · Thoracic duct · Congenital chylothorax · Iatrogenic chylothorax · Superior vena cava thrombosis · Congenital diaphragmatic hernia · Congenital lymphatic malformations · Antenatal pleural effusions

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37.1 Introduction

Chylothorax is collection of chyle within the pleural cavity. Chyle is a milky lymphatic fluid containing chylomicrons composed of small fat globules and protein and lymphocytes. The intestinal chyle containing long-chain essential fatty acid and lower limb lymphatics drains into cisterna chyli situated on the second lumber vertebra and is then carried through thorax by mostly a single thoracic duct. Iatrogenic accidental thoracic duct injury may occur in cardiac, spinal and oesophageal surgery. Chylothorax is not uncommon after mediastinal tumour resection. Radiological investigations and sometimes tissue diagnosis are required to identify the pathological cause in spontaneous chylothorax. The pathogenesis and aetiology of the congenital chylothorax remains unclear while its management mostly conservative [1]. The management of congenital chylothorax can be challenging, as it is associated with prematurity, lung hypoplasia and pulmonary hypertension [1]. In addition, associated genetic and congenital malformation complicates multisystem management. The knowledge of the anatomical relationships of thoracic duct is essential for the surgeons wanting to ligate the duct for the management of chylothorax.

37.2 Typical Thoracic Duct Anatomy [2]

Thoracic duct originates from cisterna chyli at around 2nd lumber vertebral body in midline adjacent to the abdominal aorta. It then enters the thoracic cavity through the aortic crus of diaphragm between the aorta and vena azygous on the right. Typically the thoracic duct can be found on the right thoracic cavity lying on the vertebral bodies between the azygous vein laterally, aorta medially and behind the oesophagus. It then crosses the midline at the level of 4th thoracic vertebra. Thoracic duct then ascends towards the neck where it comes out from behind the internal jugular vein and drains into the left subclavian vein at its junction with left internal jugular vein. Thoracic duct carries lymph from the lower half of the body, left chest and the lymphatic system of the left arm and left side of head and neck. The right side of thoracic cavity along with right arm and head and neck drains either independently or by a right-sided thoracic duct into the right brachiocephalic vein. It is important to note that the anatomy of thoracic duct is variable and typical anatomical description of thoracic duct is seen in 65% of populations [3]. Between 9 and 12 different anatomical variations are described in literature [3, 4].

37.3 Technical Tips and Tricks

37.3.1 Establish the Diagnosis

The presence of milky fluid in a pleural aspirate raises the suspicion, and confirmation can be ascertained by its biochemistry and presence of chylomicrons in enterally fed patients. In antenatally diagnosed hydrothorax, a pleural aspirate sent for

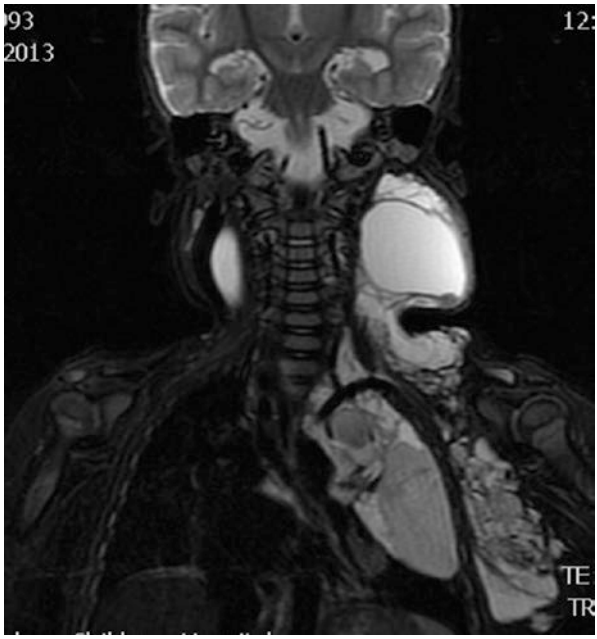

Fig. 37.1 MRI scan showing an extensive lymphatic malformation in the left axilla and neck and extending into mediastinum. He required tracheostomy as it caused significant respiratory compromise and required resection of neck cystic lymphatic malformation as injection of sclerosant caused significant respiratory distress.

biochemical analysis showing triglyceride content more than 1.2 mmol/L, protein levels greater than 2.5 g/dL, lactic-dehydrogenase above 110 IU/L and cell count predominantly of lymphocytes greater than 1000 cells/ml confirms the diagnosis of congenital chylothorax.

The radiological imaging is required to investigate its aetiology. Chest x-ray can only delineate the presence of pleural effusion and sometimes widened mediastinum, and indicates the presence of a pathological cause. In acquired spontaneous cases, in the absence of mediastinal or neck pathology and in resistant congenital cases, specific investigations are performed to identify and rule out any pathological cause before labelling as idiopathic chylothorax. Magnetic resonance imaging (MRI) is the investigation of choice for any soft tissue hamartomatous malformations (cystic hygroma, lymphangioma and lymphovenous malformation) (Fig. 37.1), lymph nodal disease (infective or malignant) and superior vena cava obstruction (primary or secondary). The lymphangiography with radionucleotide can demonstrate area of leak, lymphatic malformation. Lymphography is indicated in persistent leak that may require surgical intervention. Tissue diagnosis may become imperative in suspected malignant conditions.

37.3.2 Medical Management

- Prenatal intervention: Persistent hydrothorax detected antenatally may require either serial foetal thoracocentasis or pleuro-amniotic shunt. This prenatal intervention improves the pulmonary hypoplasia and pulmonary hypertension.

- Postnatal management: Initial diagnosis in newborn, which is not fed enterally is based on the biochemical analysis and cell content of pleural aspirate. Once the neonates are fed, the pleural fluid will contain chylomicrons and pleural fluid typically becomes milky. It is important to note that almost all congenital chylothorax cases are resolved with conservative measures. The associated mortality recorded in many series is generally related to associated congenital, genetic and cardiac defects, while some has been attributed to sepsis [5].
- Most antenatally diagnosed and postnatal early chylothoraxes generally have a degree of respiratory compromise, and many of them additionally have persistent pulmonary hypertension (PPH) that will require careful management along with the management of chylothorax. Most require insertion of intercostal tube drainage and ventilator support [5]. Supporting their nutrition initially with total parenteral nutrition and withholding the enteral nutrition have been found to be useful [5, 6]. Once there is stoppage of chyle production usually within 3–6 weeks, enteral nutrition with medium chain triglycerides can be commenced and continued up to approximately 3 months. Other supportive measures include administration of albumin, immunoglobulin, prophylactic antibiotics and fresh frozen plasma/blood products [5]. Some infants may benefit from administration of intravenous octreotide/somatostatin [5, 6]. Published regimens for administration of octreotide are variable, but the general agreement is to start with small dose and gradually increase the dose as necessary [7, 8].
- Monitoring: It is essential to monitor these infants regularly for blood counts, liver functions, serum albumin and immunoglobulins. They are prone to overwhelming sepsis, and therefore in addition to prophylactic antibiotics, regular blood inflammatory marker trends are helpful to detect sepsis in blood culture and intervene with appropriate antibiotics before fulminant sepsis.
- Most cases are successfully managed medically with supportive measures in congenital chylothorax and cases following the repair of congenital diaphragmatic hernia repair.

37.3.3 Operative Interventions

Pleuroperitoneal shunt: Some authors have claimed good success with diversion of chyle into peritoneal cavity in as high as 75% of cases along with dietary restriction of long-chain fatty acids on MCT diet [9]. This is a useful management option in intractable cases and can be palliative for incurable malignant conditions. In infants loss of chyle can cause overwhelming sepsis, and therefore its diversion into peritoneal cavity may be considered a viable option. Tube is passed both in pleural and peritoneal cavities with a pumping chamber containing one-way valve in the middle located on the thoracic wall in the subcutaneous space. The pumping chamber is pressed so as to divert the chyle into the abdominal cavity [10].

Ligation of thoracic duct: Continued significant drainage resistant to medical management despite total parenteral nutrition and bowel rest for a reasonable period

would be an indication for thoracic duct ligation. Ligation of the duct should be undertaken as it emerges from diaphragm into the chest for it to be effective [6, 11, 12]. The reported success following thoracic duct ligation is equivocal which may be related to patient selection and the duct ligation performed as a last resort in sicker patients. Patients with poor outcome following surgery could have anatomical variations and anomalies of thoracic duct.

Thoracic duct ligation can be performed thoracoscopically by identifying the duct between azygous vein and aorta on the right side. This can be aided by giving the child or infant or adult a full fat milk or double cream approximately 6–8 h prior to surgery to make the duct more identifiable. The effectiveness of surgical thoracic duct ligation in management of congenital chylothorax is uncertain. Thoracoscopic duct ligation near its entrance through the right diaphragm is very effective in containing excessive chyle leakage after head neck surgery when it is difficult to identify and ligate the leak point in the neck.

Pleurodesis: We do not recommend this approach in infants and children. However many have attempted pleurodesis in the management of chylothorax with variable success. Various chemical agents have been used. The difficulty and disadvantage is that it is a blind procedure. Therefore continued chyle leakage results in loculated and medial collections that inevitably become inaccessible to percutaneous drainage. Recently some authors have used dilute Betadine for pleurodesis in persistent chylothorax through the intercostal tube.

Thoracic duct embolization: This minimally invasive interventional radiological technique has been recently employed successfully in adults to manage chylothorax. The technique involves lymphographic identification of cisterna chyli and percutaneous catheterization and embolization of thoracic duct [13, 14]. This technically demanding technique has also showed success in young infants [15].

37.4 Chylothorax Post Congenital Diaphragmatic Hernia (CDH) Repair

The occurrence of chylothorax is well recognized following repair of CDH in approximately 10–15% of cases. The postoperative chylothorax significantly increases the morbidity associated with CDH and inevitably increases the dependence of ventilation, need for oxygen, length of stay and the risk of infection [16]. The pathogenesis of its occurrence in CDH is unclear. It is believed to be associated with PPH as other cases in premature infants and cardiac surgical cases are also associated with PPH. The management of chylothorax in patients with CDH responds generally to conservative management, providing the infant with adequate respiratory support, draining the chylous pleural effusion and keeping the infant nil orally supported with parenteral nutrition. Some studies and our experience in intractable cases suggest a period of octreotide administration is useful in addition to other supportive and conservative measures.

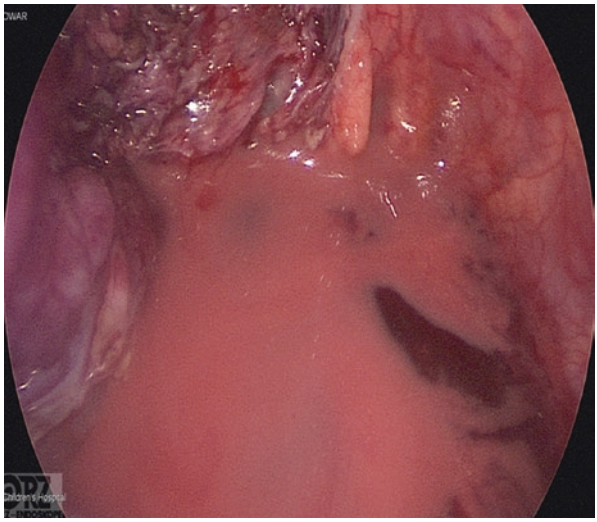

Fig. 37.2 Thoracoscopic view of thoracic cavity containing chyle prior to resection of lymphatic mediastinal malformation.

37.5 Chylothorax in Congenital Lymphatic Malformation and Mediastinal Cysts

Congenital lymphatic malformation with its anomalous and ectatic lymph channels results in leakage of lymphatic fluid and chyle into pleural or peritoneal cavity. The extensive cystic hygroma extending in the mediastinum obstructs the chyle drainage and may result in preoperative chylothorax even before surgery (Fig. 37.2). The leakage of chyle can happen after a resection surgery. The surgical accidental injury could be a result of the thoracic duct involvement in the lymphatic malformation or associated thoracic duct anomalies. Lymphangiectasia of the lung is a morbid condition and is not amenable to surgery. Chylothorax management in these cases has better chance of success with conservative approach. Surgery with thoracic duct ligation at its origin may be of value in selected cases.

Conclusions

Chylothorax can pause a management challenge. Most cases will require investigations for the diagnosis of its causative pathology. The management strategy can be tailored accordingly, both in adults and children. Conservative measures are mostly effective in infants and young children. The continuous loss of chyle results in loss of immunoglobulins and lymphocytes that may predispose infants and children to an overwhelming sepsis. Urgency to contain the chyle loss is indicated in cases that are intractable to conservative measures. Future in the management of refractory chylothorax lies in the advances in minimally invasive radiological embolization of thoracic duct techniques.

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