Successful treatment of familial congenital chylothorax by ligation of the thoracic duct: A case report

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A full term boy was admitted with respiratory distress in the fourth week of his life due to spontaneous chylothorax in his right hemithorax. Spontaneous chylothorax occurred previously in a first cousin of the neonate establishing that way the final diagnosis of familial idiopathic congenital pneumothorax. Failure of the conservative treatment consisting of chest tube drainage, discontinuation of oral diet and administration of total parenteral nutrition in combination with octreotide for one month was followed by the successful ligation of the thoracic duct through a right thoracotomy. The boy still remains free of symptoms and without recurrence of the chylothorax two years later.

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1. Introduction

Chylothorax is a very rare condition of pleural infusion in young children, occurring in approximately 1 in 10,000–15,000 pregnancies which is associated with a high overall mortality rate of 25–50% [1,2]. It is not presented commonly however; it is a very common cause of pleural effusion in the fetus and neonatal age [1,2]. It has been observed that males are most commonly affected than females. Moreover; most of the cases involve the right side of the chest [3]. Chylothorax is usually induced spontaneously due to lymphatic malformations or may be caused during birth, due to a trauma in the thoracic duct following difficult labor [4]. It may cause nutritional failure, respiratory compromise or even immune system deficiency, which subsequently can cause septic complications [1]. Herein, we present a case of familial congenital chylothorax which was definitely treated with an open thoracic duct ligation after failure of the conservative treatment.

2. Case report

A 3010 g term boy was born. Congenital chylothorax previously occurred to a first cousin of the neonate and was treated unsuccessfully elsewhere. The full term neonate was symptom-free during the first 3 weeks of his life followed by the progressive development of respiratory distress on the fourth week. Upon admission, respiratory distress and decreased breath sounds were observed on the right side of the thorax. Plain chest radiograph revealed a very large right-sided pleural effusion with a medias-tinum shift to the left and diffuse alveolar lung disease (Fig. 1). We performed a diagnostic and at the same time therapeutic thoracocentesis and we removed 150 ml of milky fluid. Biochemistry of the pleural fluid confirmed the diagnosis of chylothorax (protein 1.6 g/dl, lactate dehydrogenase (LDH) 643 U/L, triglycerides 295 mg/dl, glucose 80 mg/dl). Chest tube drainage of the right hemithorax with an Argyle No 8 tube, discontinuation of enteral feeding and
administration of total parenteral nutrition (TPN) were instituted since the establishment of the diagnosis. Intravenous infusion of octreotide started at 1 μg/kg/hr rate and progressively increased to 10 μg/kg/hr without the occurrence of any side effect. Paradoxically, chyle flow was averaging 300 cc per day with a consequent fall in serum albumin from 3.6 to 2.0 g/100 ml and a loss of 1.9 pounds (861g) of body weight. Physical deterioration was evident due to the respiratory distress. The clinical course after four weeks of conservative management did not improve and lymph drainage was more than 10 ml/kg/day. MRI lymphangiography was performed to detect any possible lymphatic pathology. The MRI results did not confirm any leak and congenital anomalies were also excluded. Surgical treatment was decided. The patient underwent right posterolateral thoracotomy, adhesiolysis and complete evacuation of the residual loculated milky pleural effusions. Chyle leak was detected in the area between esophagus, descending aorta and azygos vein and the thoracic duct underwent mass ligation with non-absorbable pledged stitches close to the hemi diaphragm. Additional pledged stitches were put on the area between azygos vein and esophagus up to the level of the carina to secure ligation of any tributaries to the thoracic duct. The infant was discharged on standard formula within 2 weeks following oral nutrition with medium-chain triglyceride containing diet.

3. Discussion

Congenital chylothorax is a relatively rare disease. It has been recognized as a clinical entity since the first report by Aselli in 1627–1628. Previously Pisek, Stewart and Linner, had firstly recognized spontaneous chylothorax in newborns from 1917 to 1926 [4,5]. In other cases, symptoms can become evident - as in the reported herein case - during the first weeks of life [4,5]. It has been previously observed that congenital chylothorax is associated with Turner syndrome, Noonan syndrome, down syndrome and hydrops fetalis [6]. Moreover; several other conditions have been associated with this condition such as; congenital lymphangiectasis, congenital goiter, lung tumors, congenital cytomegalovirus pulmonary sequestration and adenoviral infections, right diaphragmatic hernia, and group B streptococcal infections [5]. In our case, no specific etiology was found. It is remarkable to point out that the nephew of the father was diagnosed with chylothorax at the age of 5 years and multiple chest tube drains were placed until the age of 13 years.

Our case belongs to the category of idiopathic congenital chylothorax with familial character, estimated as an occasional disease with a low recurrence risk. There are rare familial cases mentioned in literature [7]. Based on these reports and knowing the 2:1 ratio male to female, it was initially believed that a possible X-linked form of inheritance existed. A more recent report by King et al. two consecutive affected female siblings were born [7]. It was suggested that autosomal recessive inheritance was more likely than X-linked recessive form [7].

Initial management of spontaneous chylothorax includes conservative treatment with chest tube drainage and nutritional support including medium-chain-triglyceride formulas and/or TPN. The intravenous infusion of the somatostatin analogue octreotide can be used in addition of TPN for the treatment of persistent or high output chyle leak [8–10]. The efficacy of octreotide on chyle leak is related to the decrease of the gastrointestinal blood flow and secretions, consequently reducing the splanchnic lymphatic flow. The success rate of conservative treatment ranges from 16% to more than 75% depending mainly on the cause of chylothorax. Non-operative management has a high rate of success >80% of the cases, even in patients with chylothorax associated with thoracic surgery, in less than 4 weeks [11]. A small percentage of patients (<10%) will require surgery after failure of conservative therapy or because of the development of complications either of the prolonged chyle leak or of the treatment itself [11]. There is no accurate etiology of conservative medical treatment failure in the literature as well as controlled studies for the exact time of duration of the octreotide administration. It has been suggested that daily use of drainage can be used as a guide for clinical improvement or failure, in specific (<10 ml/kg per day of pleural drainage is considered to be an improvement; >10 ml/kg per day of pleural drainage is considered to be a failure after 4 weeks of nonsurgical management). Generally, operative intervention can be used when: (i) daily loss exceeds 1500 milliliters per year of age in young children during a five-day period; (ii) chyle flow has not stopped during the following 14 days; (iii) in case of complications of TPN or since the development of malnutrition and/or immunosuppression; or (iv) Since the development of multiple localizations of fluid within the hemithorax despite the appropriate chest tube drainage [12,13]. Duration of conservative treatment in congenital cases depends mainly on the co-existence of other congenital defects or anomalies. However, as a rule, conservative treatment is applied for longer time periods in children than in adults, even in postoperative cases [12,13]. In the presented case, conservative management was applied for 4 weeks and according to the existing evidence surgical intervention was decided to avoid the high risk of malnutrition and immune-suppression.

The first successful transthoracic ligation of the thoracic duct was performed by Lampson in 1946 and the specific surgical procedure has major impact on the treatment of chylothorax in children [14,15]. Success rate of surgical treatment can reach up to 90%, although in up to 11% of patients more than one attempt is demanded to achieve the desired outcome. Open thoracic duct ligation has a high complication rate of up to 38% and a 25% mortality rate depending mainly on the co-existence of other congenital defects/anomalies, malnutrition or immunodeficiency and not to the procedure itself [12]. Thoracoscopic surgery is the alternative to open surgery access to perform ligation of the thoracic duct. However, the important pre-requirement of one-lung ventilation in order to proceed with thoracoscopic surgery is not always possible in neonates.

Mass ligation of the thoracic duct is a challenging procedure for the treatment of chylothorax and especially of congenital chylothorax. Ligation of the thoracic duct offers a permanent solution in neonatal with spontaneous congenital chylothorax when the...
appropriate conservative treatment fails to heal the leak. The contribution of the thoracic surgeon seems to be important in the treatment of the resistant to conservative treatment cases with congenital chylothorax.

Conflict of interest

None to declare.

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All authors contributed equally.

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