Introduction

Lymphangiomas are a heterogeneous group of benign vascular malformations of the lymphatic system, composed of cystically dilated lymphatics.1) Most cystic lymphangiomas are found in the cervical region, and isolated mediastinal cystic lymphangiomas are rare, accounting for less than 1% of all cystic lymphangiomas. Chylothorax is an uncommon condition due to leakage of chylous lymph into the thorax.

There are various therapeutic options including restriction of fat intake and surgical intervention. However, the control of chylothorax is sometimes difficult, and continuous drainage results in nutritional crisis. We present a case of refractory chylothorax accompanied with a mediastinal lymphangioma, successfully treated with...
Pleuroperitoneal Shunt for Refractory Chylothorax

Case Report

A 21-month-old Japanese boy was admitted presenting with fever and cough continuing for 17 days. The symptoms especially worsened since 9 days before admission. He had been well until then, and had no family history. On admission, he was clear and general condition was not bad. He was 81.5 cm tall (−0.5 SD) and weighed 8488 g (−2.3 SD). Body temperature was 37.6°C. Dyspnea was not seen, but his percutaneous oxygen saturation showed 93% and respiratory sound was weak on the right side. Blood test showed abnormal finding in the blood count (white blood cell count; 15800/μl, hemoglobin; 9.4 g/dl, platelet count; 96000/μl) and coagulability (fibrin/fibrinogen degradation products; 22.1 μg/ml, D-dimer; 7.3 μg/ml). Serum level of C-reactive protein was negative. Chest X-ray showed massive pleural effusion on the right thorax (Fig. 1A). Non-enhanced computed tomography (CT) showed pulmonary atelectases of the right upper and lower lobe associated with pleural effusion. Collected pleural effusion was cloudy and bloody, and had positive Rivalta reaction and positive chyle. The level of triglyceride in pleural effusion was 296 mg/dl (serum triglyceride; 112 mg/dl). Cytological examination showed no malignancy. Cultivation survey was negative in normal and acid-fast bacteria. To search for the cause of chylothorax, further imaging studies were added. Contrast enhanced CT showed poor contrast area at superior and anterior mediastinum on the delayed phase (Fig. 2A). Magnetic resonance imaging (MRI) showed granular T2-low area at the same area showed by CT, extending through the great vessels (Fig. 2B). These findings suggested mediastinal cavernous lymphangioma, and then lymphangioscintigraphy was conducted (Fig. 2C). Technetium-99m-diethylenetriamine pentaacetic acid human serum albumin (99mTc-HSAD) was injected into the first and second interdigital spaces of both feet subcutaneously. This test revealed that (1) the lymph vessels from bilateral dorsums of foot to conjunction at abdomen were clearly visualized, (2) the thoracic duct and the cisterna chilii were not detected, (3) a hot spot was detected at superior mediastinum, (4) and a leakage point into the intrathoracic space was not clear.

We diagnosed him with mediastinal cavernous lymphangioma accompanied with chylothorax. Under continuous drainage of the pleural effusion, we tried several means to decrease pleural effusion. The transition of the amount of pleural effusion is shown in Fig. 3. Initial treatment was nutritional management by using medium-chain triglycerides (MCT) formula (Meiji Co., Ltd., Tokyo, Japan), but it was unsuccessful. Therefore, total parenteral nutrition (TPN) was used since the 15th hospital day. Ocreotide (3 μg/kg/h) was administrated for 6 days since the 21st hospital day. Sclerotherapy by using minocycline (2 mg/kg/day each) through thoracic drainage tube was performed 6 times since 27th hospital day. Despite these therapies, pleural effusion continued to be drained between the amount of 400 and 1000 ml/day. Due to the loss of a large amount of body fluid, frequent transfusions of fresh frozen plasma, red cell concentrates, albumin and immunoglobulin were needed. Adjustment of electrolytes was also needed frequently.

On the 48th hospital day, we finally chose surgical pleuroperitoneal shunt.

Fig. 1 Chest X-ray showed massive pleural effusion on the right thorax on admission (A), after excision, the left pleural effusion in addition to right one and pulmonary edema emerged (B), pleuroperitoneal shunt was placed from the right thorax to the peritoneal cavity, the double-valved pumping chamber (arrow head) (C), there is no pleural effusion on the both sides in a distant time (D).
procedure because all the noninvasive therapies had seemed to be ineffective. The mediastinal tumor and the thymus were excised through a median sternotomy. The thymus had to be excised because it could not be separated from the tumor. Although 100 ml of milk with dye (1 g of Sudan Black) was administrated 2 hours prior to and during the operation, a leakage point of lymph into the intrathoracic space was not found. The thoracic duct ligation could not be performed because the thoracic duct was not detected despite thoracosscopic surveillance. Histological investigation confirmed the diagnosis of cavernous lymphangioma. Postoperative course was not easy because of symptoms induced by drastic change of lymphatic pathway; left pleural effusion in addition to right one, pulmonary edema, increased pericardial effusion and hepatosplenomegaly (Fig. 1B). Palliative therapies including tracheal intubation followed by face mask with continuous positive airway pressure, and cardiotonic agent were needed. And, the right pleural effusion had continued to be drained, although it decreased to some degree after the operation.

On the 90th hospital day, pleuroperitoneal shunt was placed by using Denver pleuroperitoneal shunt system (Denver Biomaterials Inc., Evergreen, Colorado, USA). The shunt is 15.5 Fr, 84 cm long (can be trimmed to the appropriate lengths), flexible, has no cuffs, a double-valved pumping chamber, fenestrated afferent and efferent catheter limbs. Subcutaneous tunnels were made at both the thoracic and peritoneal sides, and the pumping chamber was exposed. The chamber has a pumping volume of 1.5 ml. Therefore, the daily amount of shunt

Fig. 2  Computed tomography (CT) showed poor contrast area (arrow head) at superior and anterior mediastinum on the delayed phase (A). Magnetic resonance imaging (MRI) revealed granular T2-low area at the same site of involvement, extending around the great vessels (B). Lymphangiocintigraphy showed that 1) the lymph vessels from bilateral feet to conjunction at abdomen were clearly visualized, 2) the cisterna chilii and the thoracic duct were not detected clearly, and 3) a hot spot was visualized at superior mediastinum (arrow head) (C).

Fig. 3  A large amount of pleural effusion had continued to be drained until a placement of pleuroperitoneal shunt, although the tumor resection reduced it to some degree.
was estimated by counting the frequency of pumping (Fig. 1C). After the placement of pleuroperitoneal shunt, the amount of pleural effusion continued to decrease despite taking general diet orally. There was no adverse event including abdominal ascites, shunt closure and breakage. On the 126th hospital day, he was discharged with the shunting amount under 100 ml, which had been under 10 ml since the 142nd day. When the shunt was removed on the 148th day, subcutaneous abscess was found in the shunt tunnel of the abdominal wall. The following course was uneventful, and there has been no reaccumulation of pleural effusion and no recurrence of mediastinal tumor for 1 year of observation (Fig. 1D).

**Discussion**

Lymphangiomas are a heterogeneous group of benign vascular malformations of the lymphatic system, composed of cystically dilated lymphatics. Most cystic lymphangiomas are found in the cervical region. Isolated mediastinal cystic lymphangiomas are very uncommon, accounting for less than 1% of all cystic lymphangioma. Meanwhile, cystic lymphangioma make up 5%–6% of mediastinal masses in children. Mediastinal cystic lesions are mostly asymptomatic, and three quarters of these lesions are diagnosed in adults. Especially children under 2 years of age with mediastinal cystic lymphangioma, however, often have respiratory symptoms, including dyspnea, cough, chest pain, acute respiratory distress, and respiratory failure.

Ultrasonography, CT scan, and MRI were all used to establish a preoperative diagnosis. Common descriptions on CT include well circumscribed cystic mass with compression or deviation of the trachea, superior vena cava, etc. Contrast enhancement, visualized of septa or calcification is rare. Typical MR appearance is cystic component with the multilocular septums with low signal intensity on T1W1 and high signal intensity on T2W1 and focal inhomogeneities.

Standard treatment for mediastinal lymphangioma is surgical excision. Nanson advocated immediate surgical excision because of the potential for this entity to grow and thereby complicate future interventions. Alternative treatments include laser therapy, radiation therapy, chemotherapy, sclerotherapy, or observation. In the largest case series of mediastinal lymphangioma, all the asymptomatic cases finally had several symptoms during observation.

Pleuroperitoneal Shunt for Refractory Chylothorax

Chylothorax is an uncommon condition resulting from accumulation of chylous lymph in one or both hemithoraces. Chylothorax can be classified as congenital (idiopathic) and acquired one. Congenital chylothorax results from congenital lymphatic malformations (lymphangiomatosis, lymphangiectasia), absence or atresia of the thoracic duct, or associated with various syndromes. Although the mechanism of chylous effusion in lymphatic malformations is still unclear, leakage of the chyle into the pleural space may occur as a result of rupture in abnormal lymphatic systems, induced by pneumonia, infection, severe coughing, vomiting or trauma. Acquired one can be secondary to mediastinal malignancies, trauma, thoracic operations or vena caval thrombosis.

The initial therapy for these effusions involves complete drainage of the pleural space with thoracentesis or tube thoracostomy. Attempts to diminish thoracic lymphatic flow with total parenteral nutrition and enteral feedings with medium-chain triglycerides are also employed. Radiotherapy is reported to be effective, but it is not easy to accommodate for children because of its side effect. Surgical interventions including pleurodesis and thoracic duct ligation are important therapeutic options to resolve refractory chylothorax. Milsom, et al. reported that only 1 patient of 20 survived without an operation. In our case, the ligation of the thoracic duct was the first choice of surgical intervention, but could not be accomplished because of absence of the thoracic duct itself. Preoperative lymphangioscintigraphy revealed that the lymphatic duct from the lower limb to abdominal conjunction was clearly visualized, but the thoracic duct and the cisterna chilie were not detected. Thoracoscopic survey also could not find the thoracic duct during the operation with the administration of milk with dye. The reason why the thoracic duct was absent might be because the patient had congenital lymphatic disorder in the entire chest. Recently, video-assisted thoracoscopic surgical (VATS) approach has been recommended as it has a lower rate of complications and better cost-effectiveness. In our case, however, VATS was not applied because it was technically difficult to remove the entire lymphangionioma spreading at the superior and anterior mediastinum with involvement of the great vessels.

The timing of surgical intervention for chylothorax is still controversial. Selle, et al. have suggested that surgical intervention is indicated when the daily loss of chyle exceeds 1500 ml in adults or 100 ml per year of age in children for a five-day period, when chyle flow has not diminished over a period of 14 days, or when nutritional complications appear imminent.

In our case, thymectomy was needed because the tumor and thymus became as one part and could not be
separated. The thymus is a main place to produce activated T cells during the first years of life, and complete its roles afterward. Therefore, an immunological negative effect of thymectomy during infancy has always been of concern to surgeons. Actually, however, the partial or total thymectomy during cardiac surgery for neonates rarely induces major immunological or malignant problems, although there are effects on populations of peripheral T-cell. These lead us to think that the thymectomy for 12-month-old boy (our case) was acceptable.

Pleuroperitoneal shunt for pediatric patients was reported first by Azizkhan and colleagues in 1983 based on previous reports for adult patients with malignant pleural effusion. They used this technique successfully for 5 premature newborns who were ventilator-dependent by refractory chylothorax. Pediatric cases treated by pleuroperitoneal shunt were reported previously. Success rate was 79% overall, 83% in neonates (n = 12), 79% in infants (n = 28), 78% in child over 1 year old (n = 18), respectively. Underlying diseases were classified in 3; post-thoracic surgery (n = 27), lymphatic malformation (n = 18) and obstruction of superior vena cava (n = 13); success rate was 89%, 89% and 46%, respectively. Complications including shunt closure (10), ascerts (6), exteriorization of catheter (3), infection (3), breakage of catheter (1), lung emphysema (1) were reported. The advantages of the pleuroperitoneal shunt are less-invasive, and as effective as other surgical options. Furthermore, abnormal loss of nutritional and immunological elements through the chyle can be avoided during course of treatment. The pleuroperitoneal shunt, therefore, is worth trying, although the mechanism of this treatment has not been fully elucidated. Taking time to develop physiological drainage route without loss of body fluid may be one of the possible mechanisms of the treatment effect.

Conclusion

The pleuroperitoneal shunt for chylothorax due to lymphatic malformation could be a considerable option.

Disclosure Statement

All authors have no conflict of interest.

References