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ORIGINAL ARTICLE

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Drainage of subcutaneous lymphatic fluid for the management of respiratory distress in a case of generalized lymphangiectasia in an infant

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Abstract A 10-month-old girl was referred to our hospital because of congenital and persistent bilateral chylothorax and generalized lymphedema as well as long-standing respiratory disturbance. Radiological studies showed a diffuse network of superficial lymphatic vessels without major trunks throughout her entire body as well as the lung. She was diagnosed with systemic lymphangiomatosis complicated with pulmonary lymphangiectasia. Percutaneous puncture in the lower leg was performed to discharge the lymphatic fluid and proved to be effective for the respiratory disturbance. This procedure is safe and easy and effectively improves the quality of life of the patient and the family in case of such a persistent disease.

Keywords Generalized lymphangiectasia · Pulmonary lymphangiectasia · Pleural effusion · Chylothorax

Introduction

Congenital generalized lymphangiectasia involving the lung represents one group of Wagenaar's classification modified from Noonan's classification of pulmonary lymphangiectasia (PL) [1]. Its prognosis is reportedly better than that of isolated PL because of a less severe pulmonary involvement. Even in this type, however, management of respiratory problems in the neonatal and infantile periods is crucial for both life-saving and maintenance of a good quality of life. This article deals with a sporadic case of generalized lymphangiectasia in a young child with special emphasis on the management of respiratory disturbance.

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Case report

A 10-month-old girl was referred to our hospital because of intractable bilateral chylothorax. Hydrops fetalis and bilateral pleural effusion had been detected by means of transplacental ultrasonography in the 24th gestational week. Transplacental thoracentesis was performed three times at a local hospital until her birth at 31 weeks and 4 days of gestational age with a birth weight of 3086 g. Although she could be weaned from the ventilator assistance after 3 weeks, slight systemic edema and bilateral pleural effusion persisted. The pleural effusion changed to chyle after the start of breast feeding, so that the nutritional formula was changed to medium-chain triglyceride(MCT)-enriched milk. Moreover, minocyclin was infused into the pleural cavity twice, but both times it failed to reduce the effusion. Finally, the patient was referred to our hospital for possible surgical intervention for chylothorax. Physical examination revealed growth retardation (body weight 4400 g, SD 4.5 g; height 60 cm, SD 6 cm), puffiness of the whole body, and facial anomalies comprising hypertelorism, low-set ears, and a flat nasal bridge. Her chromosomal complement was 46XX. Similar abnormalities were not observed among her family members. A chest X-ray showed massive bilateral pleural effusion and marked interstitial images (Fig. 1A). Echocardiography indicated no significant abnormalities. Arterial blood gas characteristics could be kept almost normal (PH 7.399, $PO_2$105.6, PCO_2 42.6, HCO_3 25.8, BE 1.4) during continuous drainage of the pleural effusion consisting of a daily volume of 300–500 ml pinkish turbid fluid. Total parenteral nutrition did not reduce the chylothorax. Lymphoscintigraphy when the patient was 1 year old showed RI diffusely distributed throughout the entire body and no localized leaking of the lymph into the pleural cavity. Conventional lymphangiography using a lipid-soluble contrast medium (Lipiodol 10 ml) injected from the dorsum of the right foot showed a diffuse and fine network in both legs, the retroperitoneum, and the thoracic cage (Fig. 1B). A chest X-ray obtained 3 h after the injection of the contrast medium showed a reticular pattern of dilated lymphatic vessels. A CT scan of the chest performed 24 h after lymphangiography also showed dilated lymphatic vessels enhanced in both lower lungs. These studies indicated that the respiratory problem was due not only to pleural effusion, but also to lymphangiectasia of the lung itself. After the lymphangiography, the patient's respiratory condition deteriorated probably due to inflammation caused by the Lipiodol. Three weeks later, as her condition improved, the volume of the drained pleural effusion decreased. When the patient was 13 months old, the chest drain

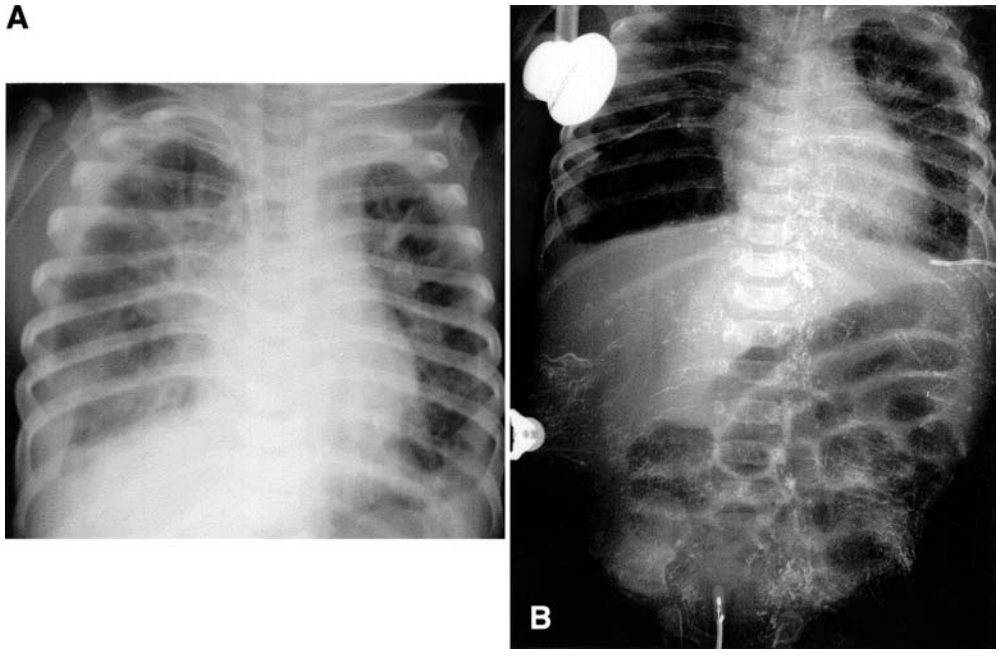

Fig. 1 **A** Chest X-ray on the patient's admission showed massive pleural effusion and interstitial images of lung fields. **B** Systemic lymphangiography taken 3 h after injection of Lipiodol showed a diffuse and fine network of lymphatics of both legs, retroperitoneum, thoracic cage, and lung

could be removed; however, after ordinary formula, milk was tried to supplement essential fatty acid, and generalized edema, especially of the lower limbs and face, increased. Her respiratory condition also became worse again despite no increase in the pleural effusion. Drainage of the subcutaneous lymphatic fluid by skin puncture was then attempted to reduce generalized lymphedema. A 24-G intravenous cannula was inserted and kept in the ante-tibial region, which was the most edematous (Fig. 2). Chylous fluid (500–600 ml) was discharged in several hours, followed by a dramatic reduction in the edema of the whole body and improvement in the respiratory condition. Blood gas characteristics also improved: before drainage, PH was 7.5, PO_2 74.4 mmHg, PCO_2 61.4 mmHg, HCO_3 32.8 mEq/l, BE 6.8 mEq/l, and SO_2 94.7%; after drainage, PH was 7.47, PO_2 128.2 mmHg, PCO_2 40.4 mmHg, HCO_3 29.1 mEq/l, BE 5.3 mEq/l, and SO_2 99.2%. After confirmation of the positive effect of this procedure, the patient could be discharged from hospital. Intermittent drainage of the subcutaneous lymph has been continued on a weekly basis by a local pediatrician. The patient is now 2 years old, and is still fed with MCT milk supplemented with parenteral free fatty acid. She is gaining weight and is gradually attaining normalcy.

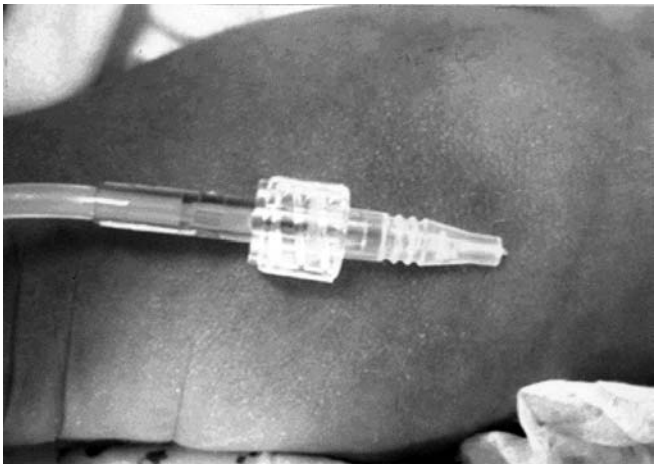

Fig. 2 Drainage of the subcutaneous lymph by a 24-G cannula. The drained fluid was chyle

Discussion

According to Wagenaar's classification, our case was type 1 (primary)–type 3 (generalized), which reportedly has a better prognosis because of a less severe form of pulmonary involvement [1]. Despite this better prognosis, intensive care is needed because of many problems due to systemic lymphangiectasia, such as respiratory, nutritional, and growth problems. Among those, the respiratory problem is the most critical and significant in the early years of life. Similar cases have been reported previously [2, 3]. Jacquemont et al. described four cases (three siblings, one sporadic) [2]. Three of the four cases showed good outcomes (survival until 3–15 years old), although two of them have occasional respiratory problems. In this congenital anomaly, respiratory function is compromised by many factors, such as massive pleural effusion, edematous airway, and poor gas exchange. Several radiological studies indicated that the lymphatic channels of our case showed a prominently abnormal configuration. Pulmonary lymphangiectasia was the major cause of respiratory distress, resulting in the treatment or management being targeted at this condition. Retrospectively, contrast dye (Lipiodol) used in the lymphangiography is thought to have caused micro-inflammation of the peripheral lymphatic vessels [4]. Inadvertently, this procedure proved to be effective in reducing the edema of the lung and pleural effusion after causing a transient deterioration in the respiratory condition; however, it did not result in a cure for PL.

In our case, local drainage of the lymph also drained the lymph of the whole body. It was surprising that chyle could be aspirated via a cutaneous puncture of the lower limb. This confirmed the open communication of the lymphatic network of this patient. Since the subcutaneous lymphatic vessels were dilated, it was not difficult to find a suitable point to obtain sufficient drainage. With this easy and safe procedure, we could reduce the edema of the whole body including the lungs and airways, and it proved to be effective for maintaining a good respiratory condition. This resulted in the eventual discharge of the patient, improved her mental development, and enhanced the relationship with her parents.

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