An Infant with Bilateral Congenital Idiopathic Chylothorax that Required Thoracic Duct Ligation

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Abstract
Chylothorax is a relatively uncommon condition defined as an abnormal collection of lymphatic fluid within the pleural space and may be encountered at any age. It may be either congenital or acquired. It is very rarely seen bilateral. Here, we present a patient with bilateral congenital idiopathic chylothorax who was treated with operative intervention.

Keywords: Congenital chylothorax, medical treatment, surgery.

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INTRODUCTION

Chylothorax is the most common cause of pleural effusion causing respiratory distress in the neonate (1). It may be either acquired (usually from trauma to the thoracic duct during surgery) or congenital and very rarely seen bilateral (2,3). The incidence has increased as cardiac surgery is performed on more complex congenital abnormalities and half of the cases are now operative complications resulting from rupture of the thoracic duct. Primary or metastatic intrathoracic malignancies, restrictive pulmonary diseases, thrombosis of the duct or the subclavian vein are the less common causes of the chylothorax in childhood (4). Congenital chylothorax may be associated with abnormalities of the lymphatic system like lymphangiomatosis and lymphangiectasia (5,6), chromosomal abnormalities (7) and H-type tracheoesophageal fistula (8). Congenital heart disease, such as isolated congenital hypoplastic superior caval vein and large atrial septal defect can also present with congenital chylothorax (9-11). The treatment options of congenital chylothorax is still controversial. We herein present a case of idiopathic bilateral congenital chylothorax.

CASE REPORT

A nine-month-old boy who has had respiratory distress since birth was admitted. The patient's history revealed that unilateral pleural effusion was detected in the sixth month of the prenatal period. Physical examination revealed tachypnea with a respiratory rate of 80/min and bilaterally decreased respiratory sounds. Chest radiograph (Fig 1) and computerized tomography (CT) of thorax (Fig 2) showed massive bilateral pleural effusion. Thus bilateral chest tubes were inserted and 300 ml of chyle drained. Pleural fluid analysis showed 360 white blood cells/mm^3 with 96% lymphocytes; 6.1 g/dl of protein and 600 mg/dl of triglyceride. Echocardiography was normal. Chromosome analysis was also normal. An enteral diet including medium-chain triglycerides and proteins had been administered for two weeks. Because of continuing cylous drainage despite this regimen oral intake was ceased and total parenteral nutrition was started. Additionally, octreotide infusion was started (0.5 µg/kg per hour) a week later. This dose of octreotide did not reduce chylous drainage. Octreotide dose was increased to 7 µg/kg per hour after seven days for five days. Patient did not benefit from this intervention also. Albumin and intravenous immunoglobulin infusions were administered to restore serum albumin and immunoglobulin levels and their levels were restored to normal. Despite all the conservative efforts for four weeks, chylous drainage was still more than 200 mL/day. Operative intervention including ligation of the thoracic duct was planned. Before the surgery, the patient was fed with cream mixed Sudan III in order to help the visualization of the thoracic duct during the operation. Under the general anesthesia the patient was placed in a left lateral position. Right thoracotomy was performed and parietal pleura was incised between the azygous vein and the vertebral bodies. The esophagus was dissected and retracted medially. The descending aorta was dissected behind the esophagus. The thoracic duct was visualized between the esophagus and the aorta. Thoracic duct was ligated together with all the fibro-fatty tissue on it's around. Postoperative period was uneventful. Thoracic drainage was slowly decreased; cylous drainage ceased and turned to serous pattern. Then, the thorax tubes of the patient were removed. In the third and

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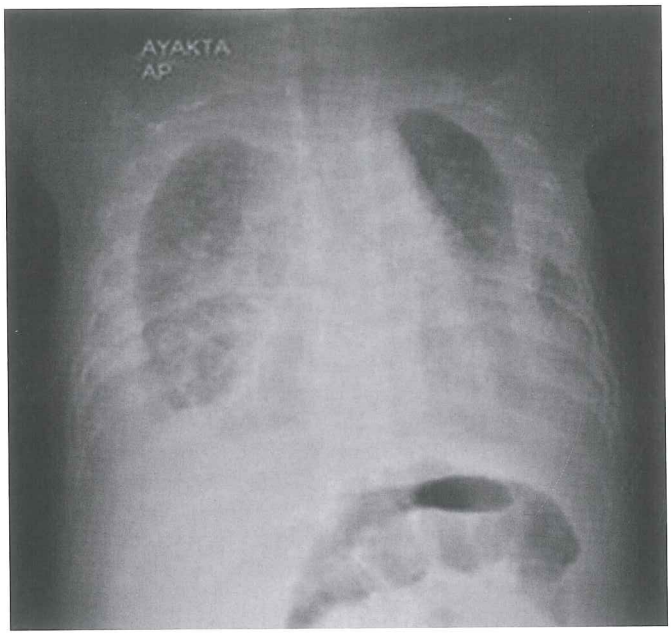

Figure 1. Chest radiograph. Note bilateral massive pleural effusions.

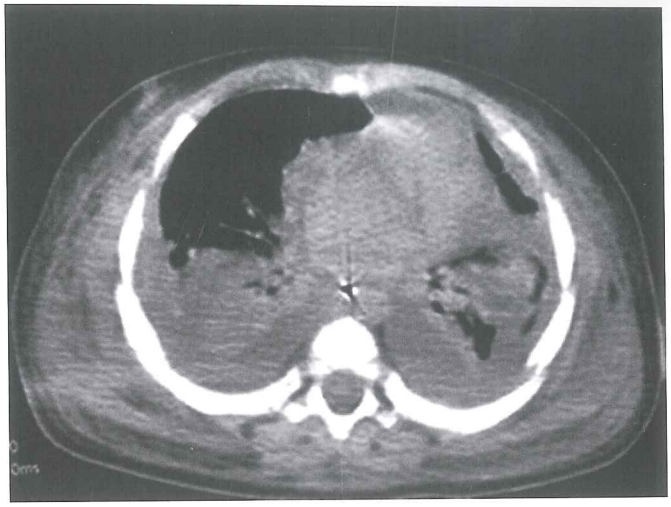

Figure 2. Computerized tomography. Note bilateral partially collapsed lower lung segments besides massive pleural effusions.

the sixth months of the follow up period, pleural effusion was not detected but pleural thickining was seen in the CT of the thorax of the patient.

DISCUSSION

Conservative treatment modalities employed in congenital chylothorax include thorasynthesis, tube thoracostomy drainage, protein rich enteral nutrition containing only medium-chain triglycerides and total parenteral nutrition (12). Additionally there are a number of reports regarding the successful usage of somatostatin (or the synthetic analogous octreotide) in the treatment of congenital chylothorax. Somatostatin reduces portal blood flow, intestinal secretion and absorption. This results in diminished chylomicron synthesis and thoracic channel lymph flow. Rasiah et al reported usage of octreotide for 10 days in a case. They started at a dose of 0.5μg/kg per hour and increased the dose daily by 1μg/kg per hour to 10μg/kg per hour which resulted in resolution of chylothorax (13). Gonzalez Santacruz et al. treated two newborn infants with congenital chylothorax by using somatostatin (14). Sahin Y et al from Turkey also used octreotide successfully in a premature infant with bilateral congenital chylothorax which remains a promising alternative to surgery (15). Our case did not benefit from octreotide administered besides total parenteral nutrition.

The duration of conservative treatment in congenital chylothorax cases is an issue that is still debated on. While some authors (16,17) recommend surgery if the pleural effusion persists for longer than two weeks; others (16-18) advocate operative intervention if daily thoracic drainage exceeds 100 mL per year of age. However; most authors recommend a longer period of conservative management and prefer to wait for four weeks for the resolution of pleural effusion (12, 19). İn our case, operative intervention was delayed for four weeks in order to wait for the resolution of the chylous drainage, but our patient did not benefit from conservative treatments. Operative interventions may include pleurectomy, talc pleurodesis, pleuroperitoneal shunting and repair or ligation of the thoracic duct via thoracoscopy or thoracotomy (20).

Prolonged chest tube drainage, total parenteral nutrition, malnutrition, lymphopenia and low immunglobulin levels in cases with congenital chylothorax have been shown to increase nosocomial infections and this consequently increases morbidity (12). Intravenous immunoglobulin has been shown to provide both antibody coverage and exert wide range of immunomodulatory effects (21). Four weeks of conservative treatment using the aforementioned modalities resulted in hypoalbuminemia and low immunoglobulin levels that we had to administer intravenous immunoglobulin which restored immunoglobulin level.

Thoracic duct ligation may be the suitable operation for massive bilateral idiopathic congenital chylothorax resistant to conservative treatment which provides prompt resolution.

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