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Short Report

Congenital Chylothorax

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Although chylothorax is an uncommon disorder, it is the most common cause of pleural effusion in the newborn infant. The majority of spontaneously occurring cases lack any known cause, while others occur secondary to thoracic and cardiovascular surgery and some are associated with lymphangiomatosis (1). It occurs twice as often in males, and the incidence has been reported as 1/10,000-15,000 (2). In this report, a male infant with congenital chylothorax who had ultrasonographically established left pleural and pericardial effusion in the prenatal period, is presented.

A 21-year-old primipara was referred to the perinatal unit of the Uludağ University Faculty of Medicine for evaluation of nonimmune hydrops that was detected during ultrasound examination at 38 weeks. The fetus was noted to have left pleural and pericardial effusion and subcutaneous edema. Moderate hydramnios was also present. Immune causes of fetal hydrops were excluded.

The infant boy was born by spontaneous vaginal delivery with a birth weight of 3000 g and Apgar scores of three and five at one and five minutes, respectively. Because of respiratory insufficiency he was intubated and mechanical ventilation was instituted. Physical findings revealed a depressed baby with respiratory difficulty and absent breath sounds in the left hemithorax. A chest roentgenogram showed a large left-sided pleural effusion which was confirmed by ultrasonography. One hundred eighteen milliliters of yellow-brown fluid was removed from the left chest by thoracentesis. The pleural fluid contained about 3860 white cells/mm^3 with 90% lymphocytes, red blood cells 1.180/mm^3. Total protein was 5.3 g/dl, glucose 179 mg/dl. The specific gravity was 1015, and the pH of the fluid was 8. Gram stain showed no bacteria, and the culture was sterile after 7 days. Echocardiography showed no cardiac defect except pericardial effusion (4 mm) and a computed tomograpy of the chest showed pleural effusion only. Complete blood count showed a hemoglobin of 13.5 g/dl, hematocrit of 41%, white blood cell count of 13,780/mm^3 with 64% segmented neutrophils, 32% lymphocytes and 4% eosinophils. Other laboratory investigations revealed normal urinalysis, blood urea nitrogen, creatinine, liver function tests, electrolytes, total protein (5.7 gr /dl), albumin (3.5 gr/dl), triglyceride (83 mg/dl), cholesterol (154 mg/dl), and glucose (122 mg/dl) levels. Blood gases demonstrated respiratory acidosis. Serologic tests for toxoplasma, rubella, cytomegalovirus and parvovirus (B-19) were negative. The karyotype was 46 XY.

In the first 24 hours, the infant required multiple thoracentesis and subsequently the placement of a chest tube into the left pleural space, which drained 180 ml of pleural fluid. Analysis of repeated samples of pleural fluid was similar to the original specimen. The infant was given prophylactic antibiotic treatment. He had a right focal seizure that was treated with phenobarbital on the 2nd day. Ultrasonography and cranial tomography showed intraventricular hemorrhage. Subsequently, the pleural fluid had a milky appearance with a high triglyceride level (1266 mg/dl) and the feedings were stopped. Total parenteral nutrition was initiated followed by a formula containing medium-chain triglyceride. Losses through the chest tube were replaced with fresh frozen plasma and

albumin. After persistent effusion for six weeks of medical treatment, the thoracic duct was surgically ligated. No signs of distal edema or respiratory problems appeared after the operation. Progressive ventricular dilation ensued after intraventricular hemorrhage which required a ventriculoperitoneal shunt.

Pisek (3), and Stewart and Linner (4) were the first to recognize spontaneous chylotorax in newborns, in 1917 and 1926, respectively. Other observations followed. Congenital chylothorax is the most common cause of pleural effusion in the newborn infant (1). There is controversy about the etiology of congenital chylothorax. A possible explanation is the vulnerable path of the thoracic duct. A chylothorax may be the result of a rupture of the thoracic duct by hyperextension of the spinal column and increased venous pressure during birth. This has, however, rarely been proven by autopsy. Diffuse leakage of chyle points more to a congenital malformation of the thoracic duct, or to a failing connection of one of the multiple segmental components of the embryonal lymphatic network (5).

Usually this condition is associated with birth trauma. It is believed that birth trauma may result in a tear of the major duct leading to persistent pleural effusion (1). This infant showed signs of hydrops fetalis at birth and had asphyxia with severe respiratory insufficiency. In 50% of cases, symptoms of congenital chylothorax are present at birth, but they can also develop during the 1st week of life. In some cases, a congenital chylothorax is associated with Turner syndrome, Noonan syndrome, hydrops fetalis, or Down syndrome (5). It is also associated with congenital goiter, lung tumors, congenital lymphangiectasis, pulmonary sequestration, congenital cytomegalovirus and adenoviral infections, right diaphragmatic hernia and group B streptococcal infections (6). In the case described here, we could not identify any specific etiology.

The diagnosis is made after analysis of the pleural fluid drained by thoracentesis or chest tube placement, procedures done for diagnosis and treatment of the pleural effusion. Chyle, a sterile fluid that appears straw-colored or milky depending on the infant's feeding status, is characterized by elevated total protein and albumin levels, a specific gravity of greater than 1012, the presence of white blood cells with a predominance of lymphocytes, and elevated triglycerides, cholesterol, and total fat levels if the infant is milk-fed (7).

The optimal treatment of congenital chylothorax has not been defined. Treatment is mainly conservative. In cases of chylothorax, feeding with formulas containing long-chain fatty acids is believed to lead to persistence of the pleural effusions by enhancing intestinal lymph flow. The basal flow of lymph in the thoracic duct can increase tenfold following a fatty meal, although smaller increases occur after a meal of protein or carbohydrate (1). It has been reported in animals that even water ingestion leads to increased lymphatic flow. Medium- or short-chain fatty acids, unlike other lipids containing long-chain fatty acids, bypass the intestinal lymphatics and are absorbed directly into the portal vein system, leading to a marked diminution in both volume and lipid concentrations of the pleural effusion (8). In many cases resolution of the chylothorax occurs spontaneously with time. The presumption is that collateral lymphatic channels develop. Repeated thoracenteses and mechanical ventilation may be needed in cases with respiratory distress. Chest tube drainage may be performed in persistent cases. When drainage remains persistent and copious, surgery may be necessary (5). Successful ligation of the thoracic duct in a case of congenital chylothorax was reported in 1957 by Randolph and Gross who, however, also sutured several small leaks in the duct (9). No lymphedema developed in our patient after ligation of the duct. Loss of electrolytes and protein in the pleural drainage may lead to decreased blood serum levels and the resultant hyponatremia or metabolic acidosis must be corrected. Because of the high concentration of lymphocytes in the fluid, lymphopenia can also occur and infection may follow (1). If the pleural effusion has not been arrested with medical management at the age of 6 weeks, exploratory thoracotomy should be performed, as in the case described here.

The prognosis of congenital chylothorax is good. Perinatal mortality is between 15 and 30 percent. Prematurity, accompanying pulmonary hypoplasia and development of nonimmune hydrops increase mortality (6). Recently, drainage has been performed in the fetus to try to prevent compression of the lungs and pulmonary hypoplasia (10).

The workup of a fetus with hydrothorax should include fetal karyotype determination, maternal antibody screen and viral studies, fetal echocardiography, and a thorough ultrasound examination to look for associated anomalies. In cases of congenital chylothorax, knowledge of the disease at birth revealed by antenatal diagnostic procedures could be life-saving and prevent severe asphyxia. Supportive mechanical ventilation is usually necessary because of either insufficient lung expansion, persistent fetal circulation or lung hypoplasia.

Resuscitation immediately after birth in the congenital form, supportive mechanical ventilation, parenteral nutrition and supplementation of noted fluid losses, lead to improvement in the final prognosis of chylothorax in the newborn.

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