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CONGENITAL CHYLOTHORAX*

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Congenital chylothorax is a rare condition in which chyle accumulates in the pleural space because of an intrauterine obstruction or anomalies of the thoracic duct. This paper presents a case of congenital chylothorax diagnosed antepartum echographically. The patient's history revealed a previous sibling with a similar diagnosis. The baby developed respiratory distress after delivery and the diagnosis was established by thoracentesis. Computed tomography of the chest and nuclear lymphangiography were obtained to evaluate the origin of the pleural effusion, but a congenital fistula or other pathology of the thoracic duct could not be demonstrated. Management of the baby consisted of ventilatory support in the delivery room, repeated thoracentesis and thoracostomy tube drainage, total parenteral nutrition and formula containing medium-chain triglycerides. The infant was discharged six weeks after birth in good condition.
Key word: congenital chylothorax.

Chylothorax is a rare cause of pleural effusion, although it is the most common cause of pleural effusion in the neonatal period[1]. Chylothorax most commonly occurs following thoracic surgery along with mediastinal masses and rarely in the neonatal period, idiopathically[2]. Chylothorax results from the escape of chyle from multiple fistulas of the malformed thoracic duct, but the anatomical defect can be demonstrated in few cases[1]. Though chylothorax is seen more commonly on the right side due to the duct's greater length on this side, it can also occur bilaterally or on the left side[3,4]. It occurs twice as often in males, and the incidence has been reported as 1/10.000-15.000[5,6]. In this report, a male infant with congenital chylothorax who had ultrasonographically established "isolated right pleural effusion" in the 36th week of gestation and a history of a sibling death with similar complaints, is presented.

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Case Report

A 27-year-old woman, para 1, was referred to the perinatal unit of Dokuz Eylül University School of Medicine for evaluation of a right pleural effusion in the fetus, which was noted during a routine ultrasound examination at 36 weeks. The first baby of the family had died at six days of life due to respiratory distress with an autopsy finding consistent with the diagnosis of left lung hypoplasia. This clinical picture was believed to be most consistent with left congenital chylothorax. The infant was delivered by cesarean section at 37 weeks.

The infant weighed 3000 g and had Apgar scores of four and six at one and five minutes, respectively. The baby was resuscitated in the delivery room, and mechanical ventilation was begun. Physical findings revealed a depressed baby with respiratory difficulty and absent breath sounds in the right hemithorax. Right-sided thoracentesis was performed in the delivery room, producing approximately 170 ml of straw-colored fluid. The fluid contained 10.000 leukocytes/ml with a differential count of 96% lymphocytes and 4% granulocytes. The protein and glucose contents were 3.6 g/dl and 137 mg/dl, respectively. The density was 1015, and the pH of the fluid was 8. Triglyceride and cholesterol contents were 22 mg/dl and 24 mg/dl, respectively. Cultures of fluid were negative. Other laboratory investigations revealed normal urinalysis, complete blood count, blood urea nitrogen, creatinine, liver function tests, electrolytes, total protein (5.9 g/dl), albumin (3.2 g/dl), triglyceride (100 mg/dl), cholesterol, glucose (138 mg/dl), amylase and lactic dehydrogenase levels. Blood gases demonstrated respiratory acidosis. Serologic tests for toxoplasma, rubella and cytomegalovirus were negative. X-ray, ultrasonography and computerized tomography of the chest demonstrated a right pleural effusion with normal parenchyme and mediastinum (Figs. 1, 2, 3). Nuclear lymphangiography was

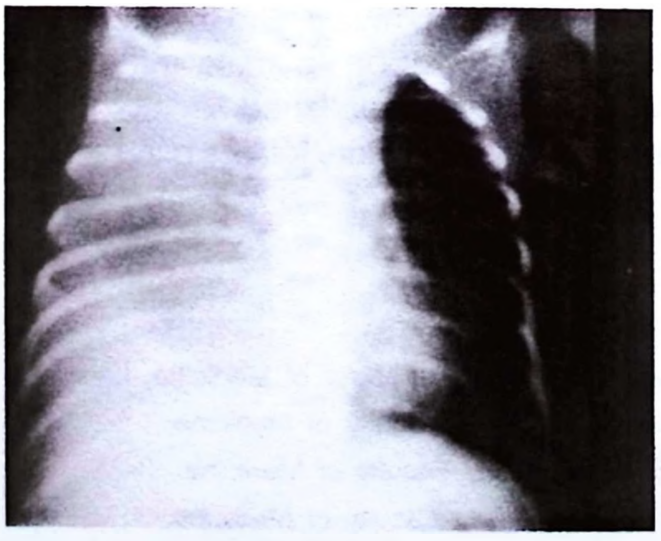

Fig. 1: Chest x-ray of the baby before thoracentesis.

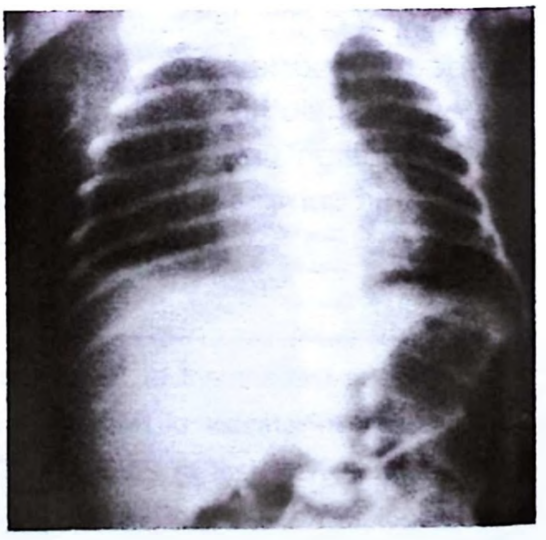

Fig. 2: x-ray of the baby after thoracentesis.

normal (Fig. 4). Repeated thoracentesis was performed because of reaccumulation. On the fifth day of life, the baby was extubated and oral feedings were begun. After the infant began feeding, the pleural fluid had a milky appearence with a high triglyceride level (140 mg/dl).

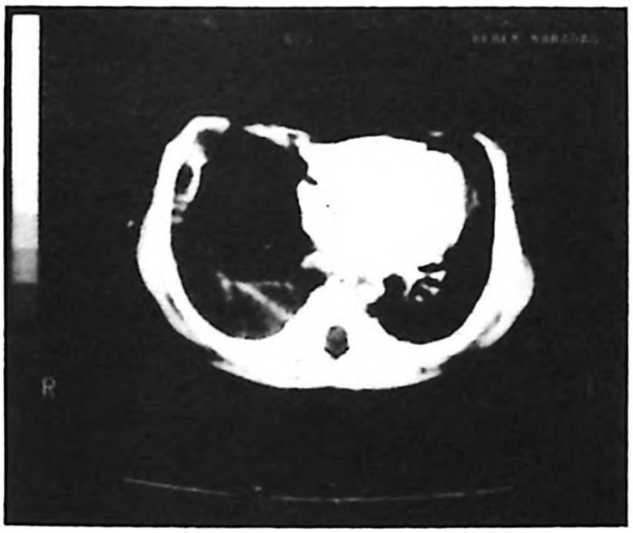

Fig. 3: Thoracal CT before thoracentesis.

Fig. 4: Scintigraphic lymphangiography.

Recovery occurred after treatment with repeated thoracentesis for four weeks and thoracostomy tube drainage for two weeks. During this period, the baby received total parenteral nutrition and then a formula containing medium chain triglyceride. The infant was discharged six weeks after birth in good condition, receiving breast milk.

Discussion

Chylothorax results from the leakage of chyle from the thoracic duct into the thoracic cavity. The fluid is milky, odorless and alkaline with a density of 1012, is biochemically composed of protein, glucose, urea and electrolyte contents identical with plasma, and is rich in triglycerides and lymphocytes[7-9]. The definitive diagnosis is established when the thoracentesis demonstrates a fluid with a chylomicron concentration of 110 mg/dl. However, the pleural fluid usually demonstrates this property after enteral feeding is started[7]. Though congenital chylothorax results from congenital anomalies and obstructions of the thoracic duct, the etiology usually remains unclear. It is also associated with congenital goiter[4], lung tumors[10], congenital lymphangiectasis[3], pulmonary sequestration[11], congenital cytomegalovirus[6] and adenoviral infections[12], right diaphragmatic hernia and group B streptococcal infections[13]. In our case, we could not identify any specific etiology. Hagay et al.[14] have reported that congenital chylothorax

might be associated with Down, Turner or Noonan Syndromes or congenital heart disease. Our case had atrial and ventricular septal defects. The most prominent finding in these patients is respiratory distress in the postnatal period due to fluid collection.

In cases of chylothorax there is increased dullness on precussion and the breath sounds are suppressed on the affected side[15]. The ultrasonograph can detect fluid collection in the gestational period which may be associated with polyhydramnios[15, 16]. Treatment is mainly conservative. Most of the cases without respiratory distress recover spontaneously. Repeated aspirations may be needed in cases with respiratory distress. Chest tube drainage may be performed in persistent cases, and if the region of leakage cannot be detected, pleurodesis may be indicated[17, 18]. Fluid, electrolyte, lipid and lymphocyte loss, pneumothorax and infection may be associated with repeated thoracenteses. The patient should be fed with total parenteral nutrition, while medium-chain triglycerides, which pass directly into the venous circulation, should be started for enteral feeding[19]. In our patient, ventilatory support was done for respiratory distress and repeated thoracenteses and chest tube drainage were carried out. He was fed with TPN, and formula containing medium chain triglycerides was started for enteral feeding.

Management after intrauterine diagnosis is not certain, as a limited number of studies and cases have been reported. Repeated thoracenteses or pleuro-amniotic shunt procedures are suggested in intrauterine-diagnosed cases, if an increase in fluid collection is observed[14]. The pleuro-amniotic shunt procedure is reported to increase survival[15]. Perinatal mortality rates range between 15 and 30 percent. Prematurity, accompanying pulmonary hypoplasia and development of non-immune hydrops increase mortality.

Mortality increases to 55 percent in cases with established pleural effusion earlier than the 32nd week of gestation[14]. In our case, absence of pulmonary hypoplasia development suggests that chylothorax had developed in the late gestational period, leading to a good prognosis. The clinical improvement of our case suggests the importance of antenatal diagnosis, early intervention and appropriate follow-up.

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