A Newborn with a Congenital Chylothorax

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ABSTRACT

Congenital chylothorax is an uncommon cause of respiratory distress and a life-threatening condition in newborns. It is identified by abnormal accumulation of chyle in the pleural space. Herein, we report a male infant who was born by a successful vaginal delivery. After the first three days of life, he had feeding difficulties and cyanotic episodes during breastfeeding. On admission, physical examination showed decreased breath sounds on the right side of the chest and the plain chest X-ray indicated right-sided pleural effusion. This patient was diagnosed with congenital chylothorax, based on the evaluation of pleural fluid via diagnostic and therapeutic thoracentesis and postnatal diagnostic X-rays. Since the effusion was persistent after drainage of pleural space, total parenteral nutrition (TPN) and surgical pleurodesis (on the right side of lung) were performed on the patient.

Due to the fact that congenital chylothorax is a life-threatening condition, reporting these cases could be a great assistance to physicians for managing the disease; it can also help with reducing the resultant morbidity and mortality.

Keywords: Congenital chylothorax, Enteral nutrition, Newborn, Pleurodesis

Introduction

Congenital chylothorax, a life-threatening condition in newborns, is identified by abnormal accumulation of chyle in the pleural space, and is considered an uncommon cause of respiratory distress (1). The diagnosis is based on the evaluation of pleural fluid, however, it is firstly diagnosed by postnatal X-rays or prenatal ultrasound (2). This disease can be treated by conservative management and surgery is only necessary in resistant cases (3).

Herein, we report a case with congenital chylothorax who was necessitated to pleurodesis on the right side of the lung.

Case presentation

The study case is a male neonate, who was vaginally delivered at 37 weeks of gestation from a 34-year-old mother (gravid 3). The infant was born in a good condition with Apgar score of 9 at the first minute. His birth weight, head circumference and height were 3500 gr, 36 cm and 50 cm, respectively.

Clinical examinations revealed none of the following symptoms: high palate, ptosis, triangular-shaped face, epicanthal folds, low-set ears, upward slanting eyes, single crease, small mouth, undescended testicles, and short neck.

The neonate had no symptoms in the first three days of life. After this period, the newborn experienced cyanotic episodes during breastfeeding and had feeding difficulties; also, respiratory distress was observed during breastfeeding later on. On admission, physical examination showed respiratory difficulties and decreased breath sounds on the right side of the chest. Vital signs were reported as follows: respiratory rate, pulse rate and body temperature were 64 per min, 140 per min and 370 °C, respectively. Saturation of oxygen was 80% and right-sided pleural effusion was present in chest X-ray (Figure 1).

Sixty milliliter of milky-appearing fluid was removed by diagnostic and therapeutic thoracentesis. The fluid consisted of triglycerides 632 mg/dl, cholesterol 68 mg/dl, glucose 105 mg/dl, lactate dehydrogenase (LDH) 519 U/L, protein 6.3 g/dl and WBC (white blood cell) 15,200 with 95 % lymphocytes.
The gram stain showed no microorganisms, therefore the culture was considered sterile. Rubella IgG antibody result was positive, unlike IgM; therefore, the neonate had no acute TORCH Infections. Also, serological and biochemistry evaluation and blood cultures showed negative results.

After diagnosing chylothorax and thoracentesis, respiratory distress significantly improved. Once the enteral feeding was stopped, total parenteral nutrition (TPN) as well as vancomycin and ampicillin antibiotherapy were started. When the amount of pleural drainage (60 ml on day 1) gradually decreased and the effusion fluid became clear on the 2nd day, enteral feeding was restarted with human milk. After this process, the symptoms reappeared within 3 days, and pleural effusion reemerged on the same side. Enteral feeding was discontinued again, and 60 ml milky-appearing fluid was re-drained. Pleural effusion reappeared on the right side for the third and fourth times; whenever the infant was unfed, the color of pleural fluid became yellowish (Figure 2).

Electrolyte and volume hemostasis was maintained and immunoglobulins and protein loss was compensated. Due to persistent effusion, the infant was transferred to referral Hospital for performing surgical pleurodesis on the right side of the lung.

**Discussion**

Having an atypical non-syndromic face, this case was diagnosed with congenital chylothorax, based on the evaluation of pleural fluid and postnatal X-rays.

The disease was first diagnosed by Asellius in 1627–1628 (2), and is identified by abnormal accumulation of chyle in the pleural space (1). It is a rare condition (1-2% of pleural effusions) with prevalence between 1:8600 and 1:15,000 live births (4). Its etiology is unknown in most newborns (5), but is mostly associated with Turner syndrome, birth trauma, Down syndrome, hydrops fetalis or Noonan’s syndrome as well as right diaphragmatic hernia, congenital cytomegalovirus, congenital lymphangectasia, adenoviral infections, lung tumors, congenital goiter and group-B streptococcal infections (2).

Chylothorax is an uncommon cause of respiratory distress (1, 6) and is a potentially life-threatening disorder in newborns, which can lead to serious metabolic, immunological, and nutritional complications (1). Right-side chylothorax is more frequently observed (55% of cases), and only 35% occur on the left side. It is usually unilateral; mediastinal may shift toward the contralateral lung and breath sounds can decrease over the site of the effusion. In the present case, right-sided pleural effusion was shown in chest X-ray, and caused decreased breath sounds in the patient. Despite the fasting of the patient, the pleural fluid became typically milky (7).

The characteristics of chylothorax are as follows: triglyceride concentration >100 mg/dl, pleural fluid protein concentration >20 g/L, number of cells per milliliter >100 with lymphocyte predominance, and sterile culture (2). Other characteristics of the chyle include glucose 2.7-11 mmol/L, and cholesterol 65-220 mg/dl (7).

Researchers have not still found the best management method for chylothorax (3,8), however, pleural space drainage, use of medium-chain triglyceride oil, fat-free baby formula, or enteric rest with total parenteral nutrition (TPN) (2) have always been the first treatment option for post-operative chylothorax.

Surgical operation should be performed in resistant cases with either excessively prolonged (>3 to 4 weeks) drainage or >10 ml/kg/day or...
>100 ml/year of age, since continued medical therapy is associated with high morbidity and mortality (9, 10). In our case, the effusion was persistent to medical treatment, therefore, surgical pleurodesis on the right side of lung was considered as the appropriate method of therapy. Due to the fact that congenital chylothorax is a life-threatening condition, reporting such cases can be a great assistance to physicians for managing the disease; it can also help with reducing the resultant morbidity and mortality.

References