Case Report

Congenital neonatal chylothorax with hydrops fetalis treated with octreotide

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ABSTRACT

Congenital idiopathic chylothorax is the most common form of pleural effusion in neonates. It results from collection of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries. Chylothorax complicated by hydrops fetalis in a premature or term infant is an uncommon, serious clinical problem. We report on two cases with congenital chylothorax both treated with octreotide. One of the cases responded well to octreotide treatment, while the other did not. We conclude that octreotide should be considered in the treatment of neonatal chylothorax.

Key words

Chylothorax; Hydrops fetalis; Octreotide; Preterm; Thoracostomy.

INTRODUCTION

Chylothorax complicated by hydrops fetalis in a premature or term infant is an uncommon, serious clinical problem. It refers to collection of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries. The prevalence of congenital type is rare, while it reaches up to 1% in post-cardiothoracic surgery, with no racial differences and male to female ratio of 2:1. Congenital idiopathic chylothorax is the most common form of pleural effusion in neonates. Because of immunologic, metabolic and nutritional complications in neonates, significant long-term morbidity and mortality were reported. Many modalities of conservative medical therapy were found to be ineffective in treating idiopathic congenital chylothorax. Although the initial first-line therapy in the treatment of chylothorax is thoracostomy drainage, octreotide, a long-acting somatostatin analog that acts on somatostatin receptors in the splanchnic area to reduce lymph fluid production, has been used in chylothorax in neonates with acceptable results [1,2]. Herein, we report two cases with congenital chylothorax both treated with octreotide. One of the two cases responded well to octreotide treatment, while the other case did not. We conclude that octreotide should be considered in the treatment of neonatal chylothorax.

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How to cite this article:

Ibrahim A. Dammas AS. Congenital neonatal chylothorax hydrops fetalis treated with octreotide. Sudan J Paediatr 2013; 13(1):43-48.

CASE REPORTS

Case 1

A late preterm male newborn was delivered at 34 weeks of gestation by cesarean section due to polyhydramnios and decreased fetal movements since two days. The mother was booked with good antenatal care. She was 29-years-old. Her blood group is O and she is Rhesus (Rh) negative. She did not receive anti-D before. There was no consanguinity between parents. Her previous obstetric history included one spontaneous abortion and two healthy children. Antenatal fetal ultrasound showed hydropic baby, so fetal echocardiography was done and showed normal fetal heart. The baby was born flat, his Apgar scores were 3 and 5 at 1 and 5 minutes, respectively. He was resuscitated and intubated and shifted to the neonatal intensive care unit (NICU). He was severely hydropic, in severe respiratory distress requiring high ventilator settings. So, thoracostomy tubes were placed bilaterally, immediately and 400 ml of serous fluid was drained. Then paracentesis was done and drained further 300 ml of serous fluid, after which hypoxemia was corrected, ventilator settings dropped to minimum and the baby was stabilized. Clinical examination revealed a hydropic baby with no dysmorphism, diminished air entry bilaterally with distant heart sounds and no neurological deficits. The patient was given surfactant on admission because of respiratory distress syndrome. Umblical vein catheter (UVC) was inserted and he received inotropic support (dopamine and dobutamine). Septic screen was done and he was started on ampicillin and gentamycin which were discontinued when cultures came to be negative. The baby was extubated to nasal continuous airway pressure (NCPAP) and weaned off oxygen over one week. Feeding was started at the age of three days and the baby reached full feeding at the age of one week. Again, he developed respiratory distress and chest x-ray showed recollection of pleural effusion bilaterally. Thoracostomy tube drainage was performed because of persistent pleural effusion after repeated thoracocenteses. Large amount of whitish milky pleural fluid was drained, analyzed and showed: triglyceride 3.1 mmol/L, cholesterol 3.22 mmol/L, LDH 216 IU/L, protein 40gm/L, glucose 7.1mmol/L, RBCs 190 X 10^3/dl, WBCs 9.5 X 10^3/dl: lymphocytes 95%, and neutrophils 5%. No microorganisms were found in the culture. Computed tomography (CT) of the chest was normal. The oral feeding was replaced by total parentral nutrition (TPN) and intralipids as fat-restricted oral diet supplemented (Monogenic formula) with medium-chain triglycerides (MCT) is not available. He was started on octreotide infusion 1µg/kg/hour and titrated up to 10µg/kg/hour with blood sugar monitoring. There was no chylus collection in the pleural space for three weeks but when octreotide infusion was discontinued and the patient was given normal milk, chylus had collected again in the pleural space, so the patient was transferred to King Fahad Armed Forces Hospital, Jeddah for surgical intervention.

Case 2

Full term baby boy was born at 38 weeks of gestation. He was delivered by cesarean section due to obstructed labor. Mother was 32-year-old, gravida 5, para 3 + 1 abortion. She had brucellosis during pregnancy treated with antibiotics, and was not suffering from other chronic illnesses. She was not booked. On admission her fetal ultrasound showed hydropic baby. Her blood group was O positive.

The baby was born flat, severely edematous with Apgar score of 0 at 1 minute, so he was immediately intubated and required prolonged resuscitation because of persistent bradycardia. Umbilical catheter was inserted and he was given all resuscitative medications, then the faint heart beats increased to 120 beats/minute.

The baby was admitted to NICU and connected to mechanical ventilation. His birth weight was 4 kg, length 55 cm and head circumference 35cm. Oxygen saturation was 80%. Clinical examination revealed a limp hydropic baby, with no dysmorphic features, who was hypotonic and hyporeflexic (Figure 1). Air entry was markedly decreased bilaterally with distant faint heart sounds. Arterial blood gas at 30 minutes of age showed PH 6.85, PCO2 58 and HCO3 5. His blood group was A+ with a positive direct comb's test. Bilateral thoracostomy tube drainage was performed and the tube drained 480 ml of serous fluid. Paracentesis was also taken and the tube drained 350 ml. Ventilation improved with oxygen saturation >95%. Because of high index of suspicion of chylothorax, analysis of pleural fluid was taken and it showed predominant lymphocytes 96%.Complete blood count (CBC) showed WBC 12.7 X 10^3/dl , Hb 15g/L, platelets 153 x 10^3/dl, reticulocyte count 6.5% and no indicators of hemolysis. Electrolytes and liver function tests were normal except for high creatinine 140µmol/L and low albumin 18g/L. The patient developed generalized seizure in the first hour which was controlled by phenobarbitone.

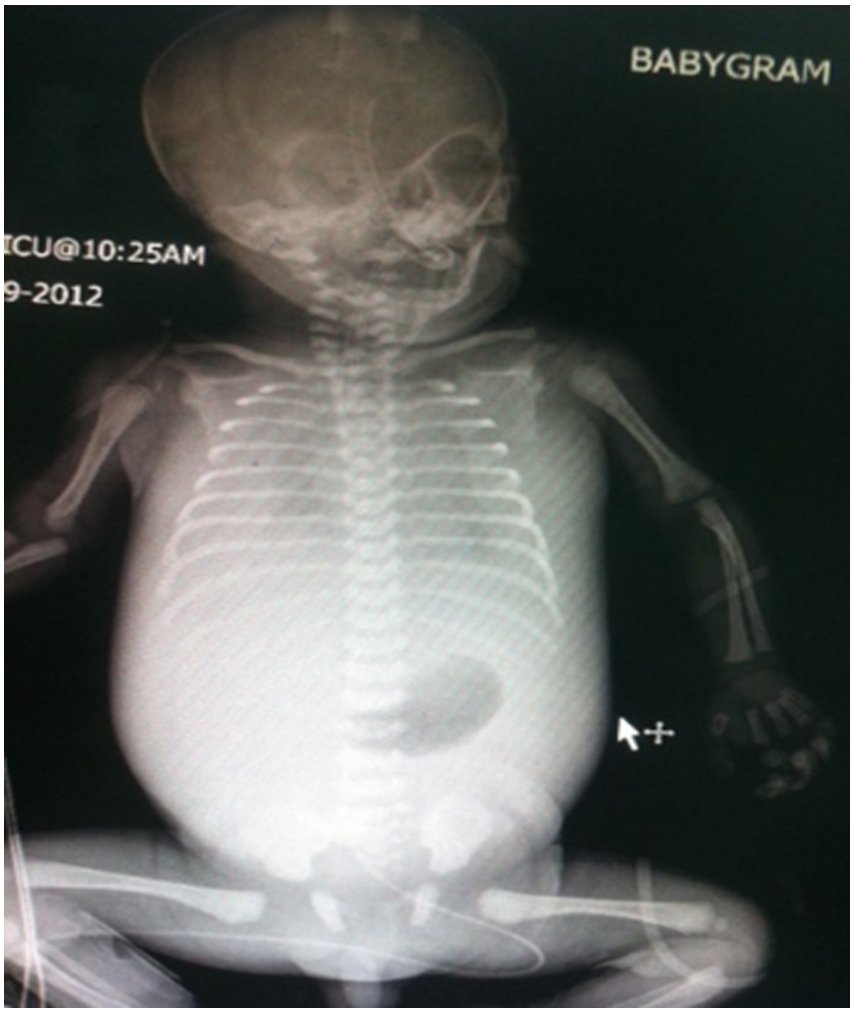

Figure 1 - Babygraph of the patient showing generalized edema, ascites and massive bilateral pleural effusion.

At the age of 5 days, milk feeding was started through orogastric tube and increased gradually reaching full feeding within one week. At that moment, thoracostomy tube drainage had changed to a milky fluid with a lymphocytic pleocytosis and elevated triglycerides, its triglyceride content was 1.51mmol/L, cholesterol content was 0.86 mmol/L and the ratio of pleural fluid cholesterol to triglyceride was, therefore, less than 1 which is diagnostic of chylothorax of chylothorax (Figure 2).

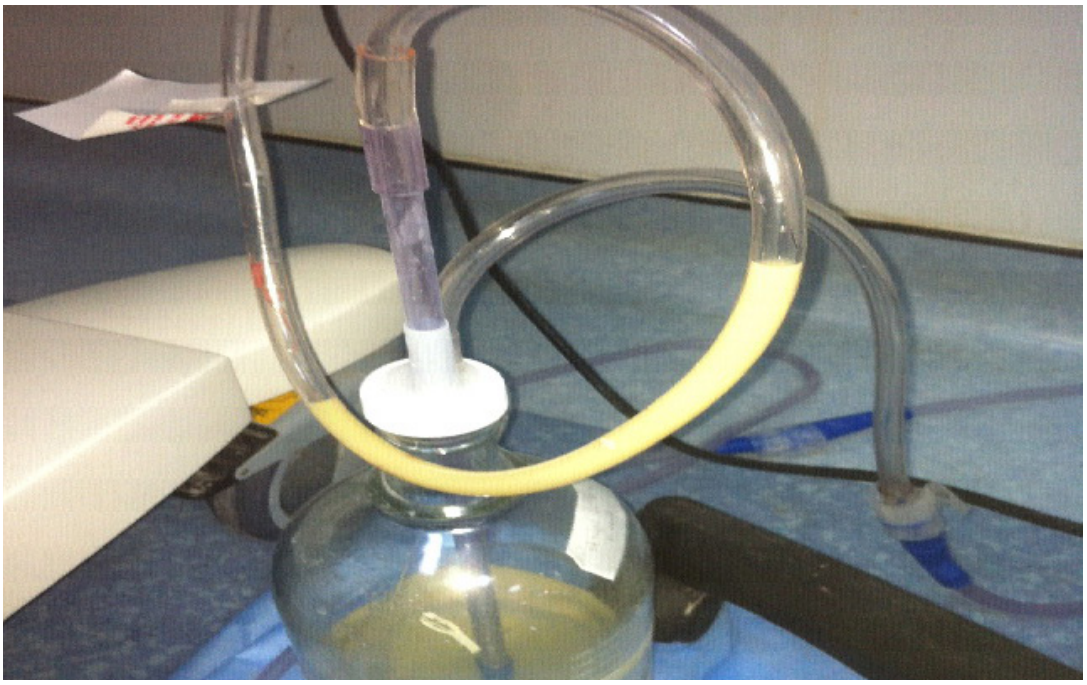

Figure 2 - Thoracostomy tube drainage showing milky fluid (chyle).

The oral feeding was replaced by TPN and intralipid. The patient had bilateral pneumothoraces drained with chest tubes (Figure 3). Then he was started on octreotide infusion 1µg/kg/hour and titrated up by daily increments till reaching 10µg/kg/hour by the 10th day , then and continued for three weeks. The thoracostomy drainage stopped by the third day of starting octreotide infusion and the tubes were removed. By the end of the third week a repeat chest radiograph demonstrated no pleural effusion in the bilateral thoracic cavities (Figure 4). Octreotide infusion was discontinued, oral feeding restarted until he was fully enterally fed without collection of chyle in the pleural space. He was discharged home after one week of full feeding. When followed up in the neonatal outpatient clinic, he was found doing well without recurrence of chylothorax.

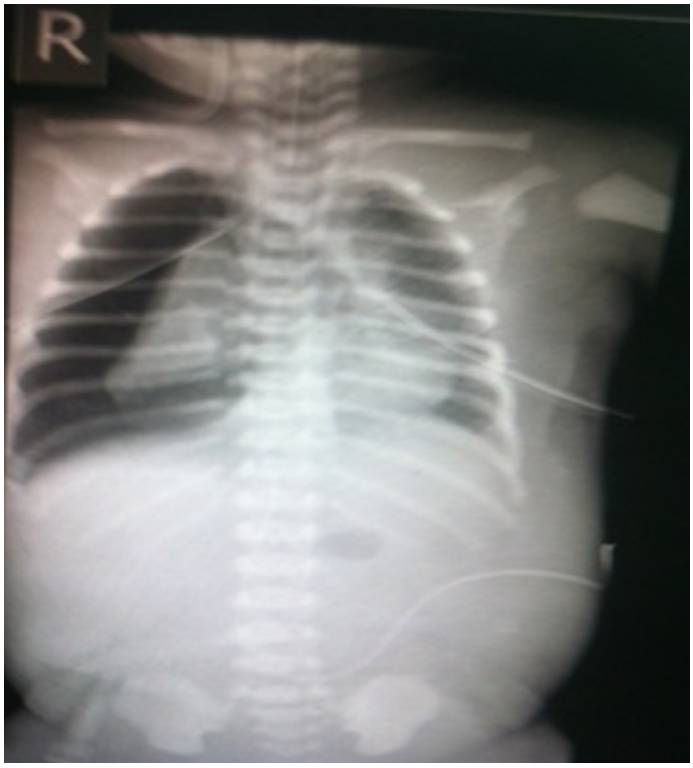

Figure 3 – Chest X-ray before treatment: Bilateral pneumothoraces.

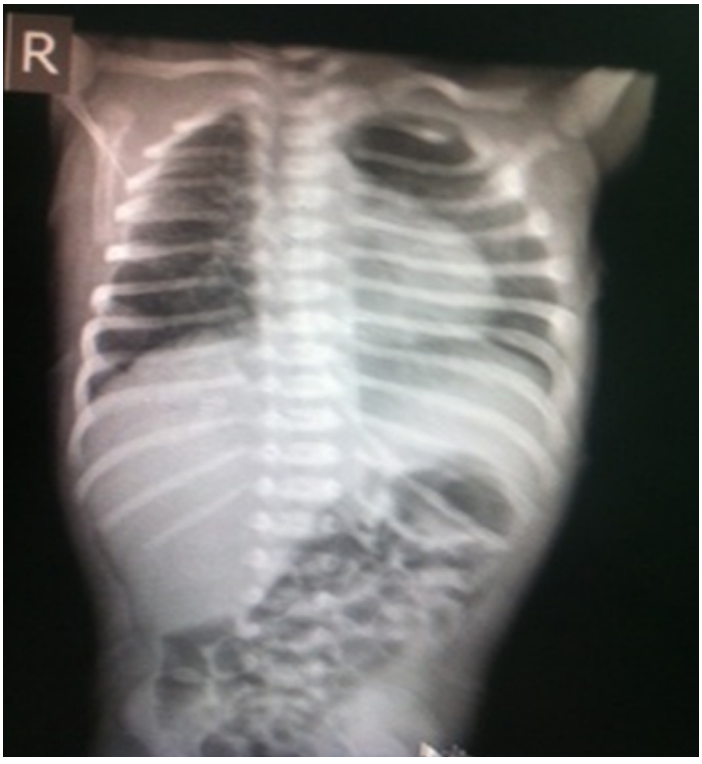

Figure 4 - Chest X-ray after treatment: No fluid or air leak.

DISCUSSION

The initial treatment of chylothorax is medical but in the case of continuation of drainage despite 2 to 5 weeks of TPN, alternative options are advocated and should be attempted like surgical ligation of thoracic duct, pleuroperitoneal shunt, pleurectomy, or chemical pleurosis [3-5]. The chylothorax in our first case did not respond to octreotide infusion therapy and thoracostomy drainage continued after four weeks.

Neonatal chylothorax is an uncommon, serious clinical problem usually complicated by hydrops fetalis in utero, and has substantial morbidity and occasional mortality. In our patients, both had severe hydrops, complicated in the second by birth asphyxia. There were no known predisposing risk factors that could lead to this congenital chylothorax in our patients. We consider it as idiopathic congenital chylothorax because the etiology is unknown. Neonatal chylothorax is usually a transient condition and resolves spontaneously with the cessation of lymph flow in the lymphatic duct for some time. This is achieved by starting TPN, withholding oral feeds and starting octreotide infusion. Octreotide is a synthetic somatostatin analogue that acts on somatostatin receptors in the splanchnic circulation, and decreases lymph fluid production through a multiple known inhibitory effects on the gastrointestinal (GI) tract [6]. These include inhibition of pancreatic secretion, decreased gallbladder contractility, slowing of intestinal transit time, inhibition of absorption of glucose and amino acids, and inhibition of GI peptide hormone secretion [7,8]. In turn, thoracostomy can lead to loss of lymphocytes, proteins, coagulation factors, and antibodies as well as lymphatic fluid, resulting in increased risk of occurrence of complications like hypoproteinemia, coagulopathy, lymphopenia, hypogammaglobulinemia, sepsis, and ventilator-related pulmonary injury [3-5]. If thoracostomy drainage persisted for more than five weeks, as in Case 1, alternative treatment lines of octreotide therapy should be considered and the patient should be referred for surgery. Chylothorax in the second case resolved completely with octreotide infusion therapy. Rapid resolution of pleural drainage was achieved with administration of octreotide in both cases without side effects, such as hypotension or hyperglycemia, throughout the whole treatment period. There was no recurrence in the second case who was discharged home earlier than the first one who required surgery.

Some babies with persistent chylothoraces (15%) eventually die, usually from complications of therapy. Standard management of neonatal chylothorax usually entails prolonged hospitalization and frequently requires surgical intervention. Administration of octreotide in our second case led to a more rapid resolution of pleural drainage, no recurrence, and early hospital discharge. Octreotide immediately diminished chyle production, without observed adverse effects but still its use as a therapy in neonatal chylothorax merits further investigation.

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