Journal of Pediatric Surgery Case Reports 31 (2018) 60–63

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

Successful use of doxycycline pleurodesis in non-immune hydrops fetalis secondary to congenital chylothorax

Vilmaris Quinones Cardona, MD[a], Michelle Clark, MD[a], Margaret Gilfillan, MD[a,*], Randy Stevens, MD[b]

[a] Department of Pediatrics, Drexel University College of Medicine[1], St. Christopher's Hospital for Children, Philadelphia, PA, USA
[b] Department of Surgery, Drexel University College of Medicine, St. Christopher's Hospital for Children, Philadelphia, PA, USA

ARTICLE INFO

Keywords:
Non-immune hydrops
Congenital chylothorax
Pleurodesis
Doxycycline

ABSTRACT

Congenital chylothorax is a rare presentation of nonimmune hydrops fetalis (NIHF). We report a case of congenital chylothorax presenting as NIHF managed successfully with chemical pleurodesis. A term male infant presented at birth with anasarca and respiratory failure secondary to bilateral pleural effusions. The infant was initially managed with ventilation and tube thoracocentesis however re-accumulation of pleural fluid occurred when feeds were started. Pleural fluid analysis was consistent with congenital chylothorax. Pleural fluid re-accumulated despite conservative management with tube thoracocentesis, nutritional interventions and octreotide therapy but responded to chemical pleurodesis with doxycycline. To our knowledge, this is the first case of congenital chylothorax treated successfully with chemical pleurodesis utilizing doxycycline as the sclerosing agent. This case adds to the literature by providing support for doxycycline as an alternative agent for chemical pleurodesis in the management of congenital chylothorax.

1. Introduction

Chylothorax is a rare condition in children, with an estimated prevalence between 1:8,600 and 1:15,000 births [1]. It is the most common form of pleural effusion in the neonate, and is defined by an accumulation of chyle within the pleural space [2]. Chylothorax can either be acquired or congenital, with congenital cases being rare and often idiopathic, occurring more commonly in males and more likely affecting the right side of the chest [2,3]. The incidence of congenital chylothorax is estimated to be 1:20,000 pregnancies [4]. Nonimmune hydrops fetalis (NIHF) secondary to congenital chylothorax is an extremely rare but potentially devastating condition with mortality rates as high as 98% [5]. Neonates with chylothorax typically present with respiratory failure and pleural effusion on chest radiograph. The diagnosis is confirmed by biochemical analysis of pleural fluid. Patients with persistent chylothorax can lose considerable amounts of fat, fat-soluble vitamins, proteins, electrolytes, immunoglobulins, and T-lymphocytes resulting in malnutrition, significant shifts in fluid and electrolyte balance, and an impaired immune system [4]. Although several different conservative and surgical treatment options exist, management of individual cases must be determined by the clinical situation and local availability of treatment options, as there is limited evidence to provide sufficient guidance [4]. This case highlights the successful use of chemical pleurodesis with doxycycline in a neonate with NIHF secondary to congenital chylothorax following the failure of conservative management with tube thoracostomy, medium-chain triglycerides (MCT), total parenteral nutrition (TPN), and octreotide.

2. Case report

A 4600 g male infant was born at a gestational age of 37 weeks and 2 days to a 28 year old G3P2002 mother with no significant medical history. He was delivered via C-section for failure to progress following an uncomplicated pregnancy and unexpectedly required intubation in the delivery room due to respiratory depression. Initial examination findings included marked diffuse subcutaneous edema and bilateral diminished breath sounds. Apgar scores were 1, 2 and 2 at 1, 5 and 10 min respectively.

On admission, the infant went into cardio-respiratory arrest and radiographs revealed bilateral pleural effusions and ascites (Fig. 1). Emergent bilateral thoracocentesis and paracentesis yielded a total of 136 ml of straw-colored pleural fluid and 26 ml of straw-colored peritoneal fluid with subsequent improvement in hemodynamics. Pleural fluid analysis revealed a lymphocytic predominance with low protein

* Corresponding author. St Christopher's Hospital for Children, 160 E.Erie Ave, Philadelphia, 19134, USA.
E-mail address: mag465@drexel.edu (M. Gilfillan).
[1] Drexel University IRB No: 1704005327.

https://doi.org/10.1016/j.epsc.2017.12.013
Received 9 October 2017; Received in revised form 11 December 2017; Accepted 16 December 2017
Available online 18 December 2017

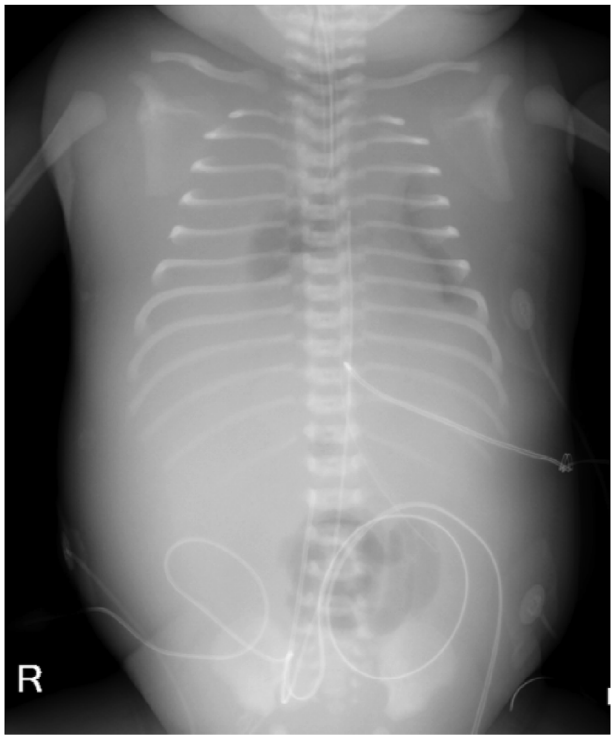

Fig. 1. Chest and abdominal radiograph demonstrating subcutaneous edema, bilateral effusions and centralized bowel loops suggestive of ascites.

Table 1
Pleural fluid analysis results.

Pleural Fluid Analysis	Birth	After starting feeds
Albumin	1.4 g/dL	1.3 g/dL
Glucose	93 mg/dL	88 mg/dL
LDH	114 units/L	142 units/L
Protein	1.8 g/dL	1.8 g/dL
Triglycerides	Not done	677 mg/dL
%Neutrophils	12	0
%Eosinophils	1	4
%Lymphocytes	84	92
%Macrophages	2	4
%Reactive lymphocyte	1	0

and LDH (Table 1). The patient was transferred to our institution for further management of hydrops fetalis and respiratory failure.

Upon arrival, bilateral tube thoracostomies were placed for continuing drainage of pleural fluid. Over the course of his first week of life, the pleural effusions gradually diminished in size and the infant was successfully extubated on day 7. The infant also tolerated introduction of enteral nutrition, however as he advanced towards full feeds, his tube thoracostomy output increased with a biochemical profile consistent with chylothorax (Table 1). Feedings were switched to MCT formula on day 12 in response to these results. Virology (CMV, EBV, HSV 1 and 2, rubella, Parvovirus B19, toxoplasma) and syphilis analyses were negative. Chromosomal microarray, ophthalmologic exam, echocardiography, head, abdominal and renal ultrasounds were normal.

Multiple tube thoracostomy replacements were required due to frequent occlusion and re-accumulation of pleural fluid. Octreotide infusion was started on day 22 at 1 mcg/kg/hr and slowly titrated to 3 mcg/kg/hr. No improvement was noted in chest tube output and octreotide infusion was stopped after 4 days due to symptoms concerning for necrotizing enterocolitis. Abdominal radiographs were within normal limits, however given the critical nature of his illness, enteral rest was continued. The infant's course was complicated with MSSA bacteremia and pleuritis leading to a decompensation in his respiratory status on day 24 requiring re-intubation. At this time, his IgG level was measured at < 200 mg/dL and a single dose of IVIG was administered. The infection was subsequently treated with a 10 day course of naficillin. In addition to IVIG, the patient also received frequent albumin replacements due to hypoalbuminemia and high tube thoracostomy output.

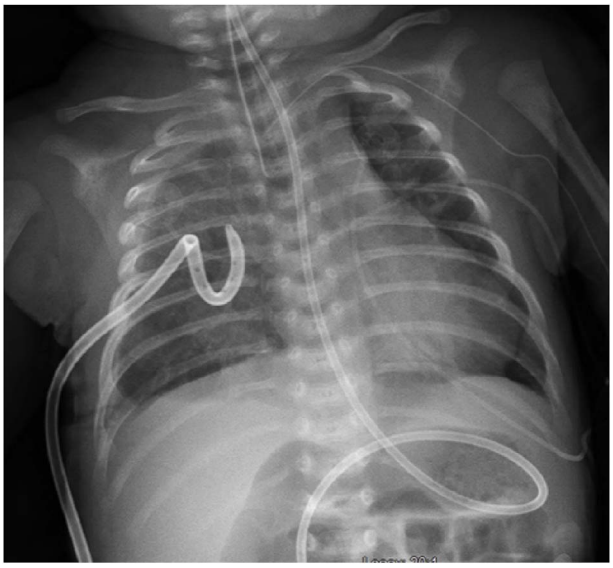

Fig. 2. Chest radiograph after chemical pleurodesis demonstrated a pigtail catheter in right hemithorax with a small residual apical pleural effusion.

Despite enteral rest, TPN, supplemental albumin and maintenance of thoracostomy tube patency and drainage, the infant continued to have over 30 ml/kg/day of chest tube output. Cardiothoracic surgery was consulted and recommended chemical pleurodesis with doxycycline which was initiated on day 32 (see pleurodesis technique). On day 2 and 3 of pleurodesis there was minimal thoracostomy output and chest radiograph showed a persistent apical pleural effusion. Due to concerns for possible thoracostomy tube obstruction, the pigtail catheter was replaced which did not yield pleural fluid and led to a small pneumothorax. Pleurodesis was stopped after 3 days given marked improvement of the right pleural effusion (Fig. 2). The thoracostomy output remained minimal while feeds were advanced. He was extubated on day 43, and the pigtail catheter was removed on day 44 without reaccumulation of pleural fluid. He was eventually discharged home in stable condition with the diagnosis of NIHF secondary to congenital chylothorax.

3. Pleurodesis technique

Chemical pleurodesis was performed with 1.5 mg/kg lidocaine and 4 mg/kg of doxycycline with the intention of a 5-day treatment. Chest x-ray was obtained prior to procedure to confirm correct placement of the pigtail catheter. Fentanyl was given for procedural analgesia. A 20 ml mixture of lidocaine, doxycycline and sterile water was instilled over 10 min in a sterile manner via an 8F pigtail catheter with close monitoring of hemodynamics and pain. The mixture was left to dwell with repositioning every 15 min for 2 h after which the pigtail was placed back to suction. Tube thoracostomy output was recorded and chest x-rays were repeated daily to monitor pleural effusion size. This was repeated daily for 3 consecutive days. Please refer to the procedural check list included in appendix A for further details.

4. Discussion

Management strategies of congenital chylothorax range from