International Journal of Medical and Pharmaceutical Case Reports
5(2): 1-5, 2015; Article no.IJMPCR.19624
ISSN: 2394-109X

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Congenital Unilateral Chylothorax in a Non-immune Hydropic Newborn with Down Syndrome

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Authors' contributions

This work was carried out in collaboration between all authors. Authors BSK and KK wrote the paper. Authors BS and BB collected references from literature and revised the article. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/IJMPCR/2015/19624
Editor(s):
(1) Erich Cosmi, Director of Maternal and Fetal Medicine Unit, Department of Woman and Child Health, University of Padua School of Medicine, Padua, Italy.
Reviewers:
(1) Jaspinder Kaur, Department of Obstetrics & Gynaecology, Punjab Institute of Medical Sciences, India.
(2) Somchai Amornyotin, Department of Anesthesiology, Mahidol University, Thailand.
Complete Peer review History: http://sciencedomain.org/review-history/10217

Case Study

Received 20th June 2015
Accepted 2nd July 2015
Published 16th July 2015

ABSTRACT

Chylothorax is defined as accumulation of lymph within the pleural space. It is the most common cause of pleural effusion causing respiratory distress in the first few days of life. Congenital chylothorax and Down syndrome are independently have been found in association with non-immune hydrops fetalis. On the other hand, these entities can seen together. The treatment of congenital chylothorax generally based on cessation of enteral feeding, initiation of total parenteral nutrition and introduction of medium chain triglycerides. Surgical intervention should be encountered when medical therapy fails. We present a hydropic newborn with Down syndrome and chylothorax who was successfully treated by chest drainage and conservative treatment.

Keywords: Congenital chylothorax; down syndrome; hydrops fetalis; neonate.

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1. INTRODUCTION

Chylothorax is defined as accumulation of lymph within the pleural space and is a potentially life-threatening condition in neonates [1] Generally, the causes of chylothorax in childhood can be classified as following; a) Congenital; as an isolated finding or associated with chromosomal anomalies (e.g. trisomy 21, Turner's syndrome), congenital malformations and disorders of the lymphatic system (e.g. Pulmonary lymphangiomatosis), b) Traumatic; post-surgery or non-iatrogenic, c) High central venous pressure; e.g. thrombosis, d) Malignancies; Lymphoma etc., e) Miscellaneous; e.g. tuberculosis and transdiaphragmatic movement of chylous ascites and idiopathic cases [1,2]. The prognosis is variable and treatment generally based on conservative methods [1,3].

Hydrops fetalis is fluid accumulation within the fetal tissue compartments and body cavities and is has been reported as an unusual manifestation of Down's syndrome [4]. Congenital chylothorax (CC) and Down syndrome are independently have been found in association with non-immune hydrops fetalis (NIHF) [4]. And also these entities can seen together [4]. We present a hydropic newborn with Down syndrome and chylothorax who was successfully treated by chest drainage and conservative treatment.

2. CASE REPORT

A fifteen-day old male newborn was referred to Private Istanbul Hospital for respiratuary distress. He had been born at term to a 29-year-old multiparous mother. The antenatal period was uneventful. He was the third child of a non-consanguineous marriage, born by normal vaginal delivery. He was intubated and accepted in another neonatal intensive care unit (NICU) due to severe respiratory distress after delivery. Initial chest graphy was showed diffuse reticulogranuler pattern. He was received surfactant and antibiotics. Despite of treatment his clinical status has been deteriorated so he was referred to our NICU. On admission, the neonate was intubated. Skin was edematous. There was also typical stigmata of Down syndrome.

Hemoglobin was 10.7 g/dl, leukocyte count was 10,770/mm^3 and thrombocyte count was 127,000/mm^3. Biochemical analysis showed plasma creatinine 0,7 mg/dl, glucose 77 mg/dl, triglyceride 58 mg/dl, albumin 2.2 g/dl, C-reactive protein 31.7 mg/l (N:0-5), TSH 48.7 mIU/ml (N:0.3-2.83 mIU/ml) and FT4:1.51 ng/dl (N:2.0-4.9 ng/dl). TORCH serology was negative. Chromosome analysis revealed the karyotype as 47,XY,+21. Echocardiography showed no structural anomaly.

On admission chest X-ray graphy showed no effusion but after two days a radiopaque image in the left hemithorax has seen (Fig. 1). Thorax ultrasound confirmed the pleural effusion, so a diagnostic thoracentesis performed. The biochemical analysis of the dense, yellow fluid revealed the following: protein 3.59 g/dl, albumin 1.93 g/dl, LDH: 329 IU/L, triglyceride 719 mg/dl, leukocytes 2,390/mm^3; on microscopy there was 30% granulocytes and 70% lymphocytes. The fluid was considered as chylous effusion. A chest tube inserted then about 200 ml yellow fluid was drained in first day. Culture was negative. The infant was put on ventilator on SIMV mode and antibiotics were started. Hypothyroidism was diagnosed and L-levothyroxine was started. Enteral feeding was stopped and total parenteral nutrition (TPN) was started. Respiratory distress remained and infant could not extubated during first 10 days. Chest tube was removed after 5 days when there was no drainage for 48 hours and effusion has been recovered (Fig. 2). By day 15 of hospitalization, respiratory distress resolved and infant extubated however he has to account for continuous positive airway pressure (CPAP) for 7 days. Medium-chain fatty acids (MCT) was started (GOLDEN GOAT 1-manifactured by DGC, New Zelland) and increased to 120-130 ml/kg/day gradually. The patient was discharged with an MCT-rich formula diet on 55th day. He was followed for 6 months and was in good condition.

3. DISCUSSİON

The word ‘chyle’ means a milky coloured fluid composed of chylomicrons, immune cells and proteins [1,5]. Firstly, chyle is clear and yellow, becoming milky after beginning of enteral feding [6]. The diagnosis of chylothorax depands on definition the chyle in plevral effusion by a triglyceride level >1.1 mmol/L (110 mg/dl) and a cell count >1,000 cells/mm^3 with a predominance of lymphocytes (>%80) as described by Staats et al. [5]. It has been reported that the range of age of occurrence was 5-46 days for nonacquired chylothorax and was 2-44 days for postoperative chylothorax [7]. This finding is consistent to our case that chylothorax has been determined by 17th day of life.

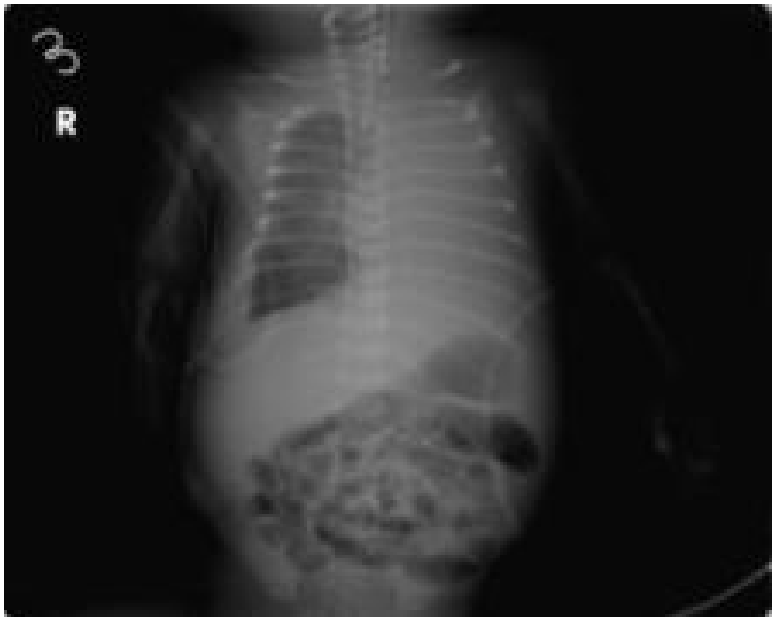

Fig. 1. Radiopaque image in the left hemithorax on day 2 of hospitalization

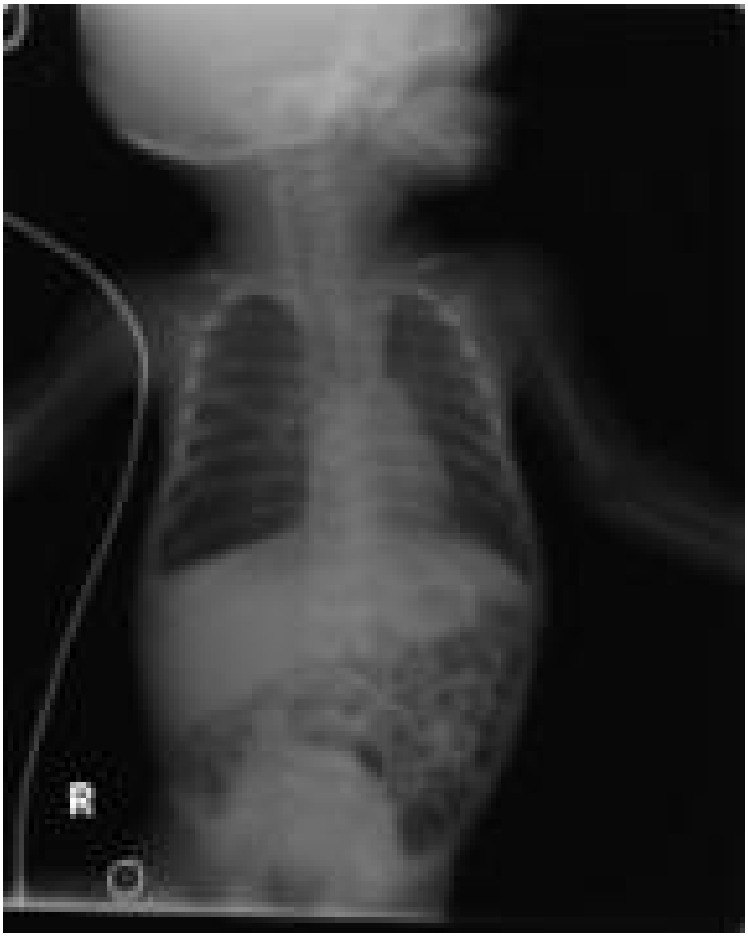

Fig. 2. Recovery of chylothorax after treatment by chest tube drainage

Down syndrome in association with non-immune hydrops fetalis and CC has been well described in English-language medical literature in a few reports but Down syndrome with absence of cardiac defects and other major surgical malformations in association with NIHF and CC is an unusual entity [3,4]. The pathogenesis of generalized edema in Down syndrome and CC is not well established but protein leakage into the pleural cavity and/or elevation of intrathoracic pressure with subsequent obstruction of venous return had been charged [3].

Treatment is based on minimizing the chyle flow in the thoracic duct to wait for spontaneous healing of the leakage site [8]. Conservative treatment modality consists of pleural drainage; decreasing lymphatic flow by interruption of enteral feeding, initiation of TPN and introduction of MCT and also assisted ventilation in cases of respiratory distress [8]. It has been suggested that a single thoracentesis might be adequate because it will lead to expansion of lungs and will tamponades the duct or lymphatic defect [6]. But mostly, continuous drainage with a chest tube is indicated, due to respiratory distress or the accumulation of effusion [8].

Recently, somatostatin and its analogue, octreotide, which inhibits lymph secretion, has been successfully used specially in cases who did not respond initial treatment [8]. Although conservative therapy was adequate in most patients, surgical intervention (e.g duct ligation) is recommended after a 5 to 7 days course of medical therapy has failed [5,6]. MCT can bypass the intestinal lymphatics and are absorbed directly into the portal vein system [9]. So MCT-rich diet will cause a marked reduction of volume and lipid concentrations in the pleural effusion as well as can provide adequate nutrition [9]. We used TPN for first 15 days of hospitalization with minimal enteral nutrition then maintained with oral MCT-enriched diet and consequently encountered no reoccurence of chyle accumulation. However, it has been recently suggested that it is not necessary to give MCT diet as the first line feeding formula and MCT or parenteral nutrition should be used only when the CC worsens after beginning enteral feding [9].

4. CONCLUSION

The survival rate of newborns with CC and NIHF is variable and is mostly dependent on underlying disorders. Here we report a newborn with a rare and complex clinical entity. We found an association between NIHF with unilateral chylothorax and Down syndrome. We want to emphasize that, chest tube drainage could be performed successfully in whom other therapy modalities failed to minimize liquid accumulation, specially in patients with marked respiratory distress.

CONSENT

It is not applicable.

ETHICAL APPROVAL

Ethical approval is not necessary for a case report in Private Istanbul Hospital, Van, Turkey.

COMPETING INTERESTS

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/10217