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A rare case of congenital chylothorax in a Palestinian neonate

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ABSTRACT

Congenital chylothorax (CCT) is a rare condition which is characterized by an accumulation of lymphatic fluid in the pleural space and exposes the newborn to respiratory distress and losing of proteins, coagulation factors and immunoglobulins. These cases are liable to have sepsis and high mortality rate. We report a case of a female fetus in Gaza delivered at 36 weeks gestational age diagnosed with CCT. The antenatal ultrasonography showed right sided significant pleural effusion and at birth; she had severe respiratory distress. The baby was intubated at birth and right-sided chest tube drain inserted to drain the pleural fluid which was chylus. The case responded partially to intravenous (IV) octreotide and sildenafil. Chylus stopped completely after 2 days of treatment with octreotide and medium chains triglyceride (MCT) oil-based formula feeding. **Conclusion**: A female newborn diagnosed with right-sided pleural effusion by antenatal scan was confirmed to have congenital chylothorax postnataly. The infant responded to IV octreotide and MCT based formula feeding.

1. Introduction

Congenital chylothorax (CCT) is a rare condition and its incidence is 1:8600 to 1:10000 live birth [1]. The condition is characterized by the accumulation of lymph in the pleural space which could cause pressure on the intrathoracic organs. The drained chyle contains lymphocytes, proteins, coagulation factors, and fluid. These babies are liable to have difficulty in breathing, sepsis, bleeding, and dehydration. The mortality rate of CCT is 20–60%. CCT could be associated with a different syndromes like trisomy, monosomy and X-linked myotubular myopathy, missense mutation; in integrin α9β1,11,12 and Gorham–Stout [2,3].

1.1. Background

Congenital chylothorax is rare and if not diagnosed and treated in time appropriately, it has a high fatal outcomes and serious complications [4,5]. We report a case diagnosed antenatally at 36 weeks gestation with pleural effusion and after birth congenital chylothorax was confirmed after insertion of a chest drain. The case responded completely and resolved after IV octreotide and MCT formula milk feeding but partially responded to IV octreotide and oral sildenafil.

2. Case report

A female fetus was found at 36 weeks of gestational age to have a significant right-sided pleural effusion; leading to mediastinum shift to the left side (picture 1–2). A multidisciplinary team meeting including the obstetricians, neonatologists, and pediatric surgeons suspected congenital right sided chylothorax and recommended delivery by cesarean section at 37 weeks gestation to prepare for resuscitation and appropriate investigations includes the genetic studies. A female baby was delivered at the 37 weeks gestational age by cesarean section. Apgar score was 4 and 6 at 1 and 5 minutes respectively. On physical examination, there was no dysmorphic features and external genitalia was normal. Birth weight was 2700 g and head circumference was 34 cm. At birth, the baby had severe respiratory distress and hemodynamically was unstable. The baby was intubated immediately and right-sided chest tube drain was inserted. After chest drain insertion, the baby started to stabilize on mechanical ventilation and oxygen saturation improved to 98%. Chest X-ray showed chest drain in place (picture 3) and during the first 24 hours 250ml yellowish fluid drained from the right-sided of the chest. The baby was extubated after 24 hours and placed on nasal cannula with a low flow of 2l/min and oxygen 25% and there was no clinical evidence of increased of breathing. Since then, the baby neurologically

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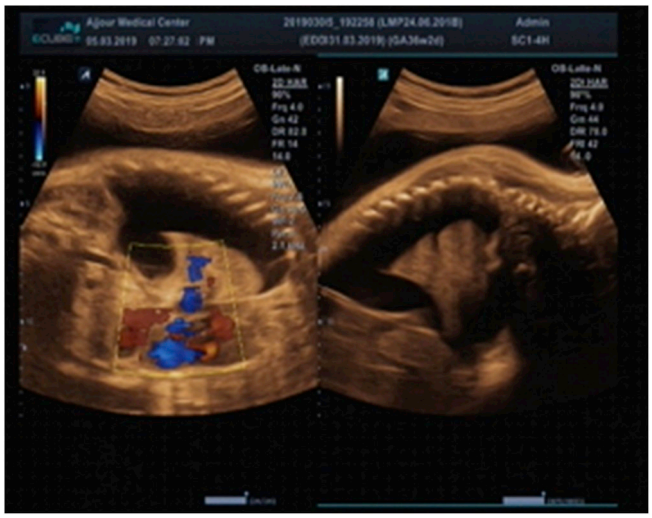
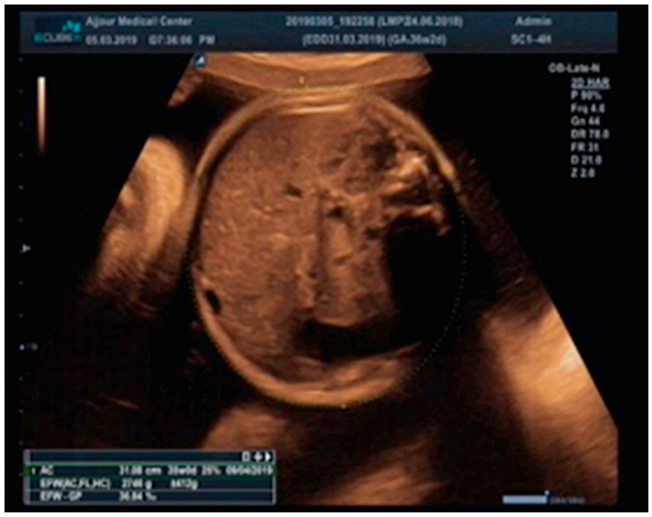
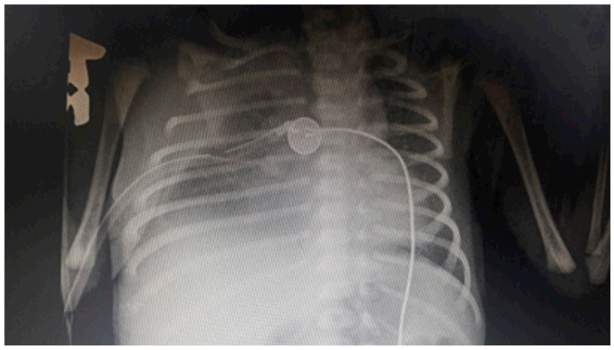


Picture 1. Female fetus was diagnosed at 36 weeks gestational age that had significant pleural effusion in the right side which pushing the mediastinum to the left side (picture 1–2).



Picture 2. Pleural effusion in the right side pushed the mediastinum to the left side.



Picture 3. Chest X-ray showed that pleural fluid collection and chest drain in place.

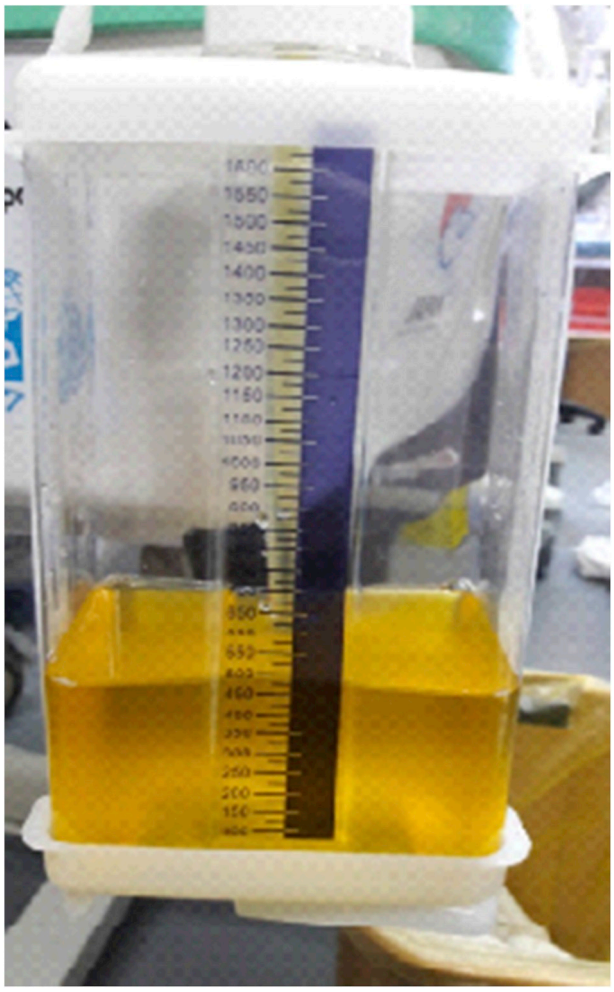
and hemodynamically was stable. She was fed expressed breast milk by orogastric feeding tube. Brain ultrasonography, abdominal ultrasonography, and echocardiography were normal. On the second day of age, 50–100ml of fluid drained and octreotide IV infusion at a rate of 1 μg/kg/hour was started with slow daily increment. On day 7 of age, The octreotide infusion rate was 3 μg/kg/hour. Oral sildenafil 2mg/kg every 6 h was initiated since the second day of age till the day 13 of age. When the baby was fully fed via orogastric tube using expressed breast milk, the chest drainage started to become milky in color (picture 4–5). At age of 8 days, the baby orogastric feeding stopped and total parenteral nutrition started whilst maintaining IV octreotide infusion at a rate of 3 μg/kg/hour. At this stage, the chest drain amount was less (about 30–40 ml/daily) but not stopped completely. At the age of 14 days, pregestimil milk formula started (MCT based formula) and IV octreotide infusion at a rate of 3 μg/kg/hour continued. After 48 hours of this regimen, chyle drainage stopped completely and the IV octreotide was gradually decreased over 48 hours before discontinued completely. The baby was discharged home in a good condition at the age of 20 days. At the age of 40 days, the pregestimil formula was discontinued and the baby received regular formula milk without the relapse of chylothorax.

*Investigations*Results of blood, pleural fluid and other investigations.

	Result	Normal range
CBC		
WBC	30000×10^9/L	5000-30000×10^9/L
Hemoglobin	16.5 g/dl	13–20 g/dl
Platelet	294×10^9/L	150-450×10^9/L
Blood chemistry		
Serum urea	10 mg/dl	15–50 mg/dl
Serum creatinine	0.3 mg/dl	130–150 mg/dl
Serum sodium	138 mmol/l	130–150 mmol/l
Serum potassium	5.1 mmol/l	3.5–6.0 mmol/l
Serum chloride	110 mmol/l	93–112 mmol/l
Serum calcium	11.17 mg/dl	8.4–10.6 mg/dl
Serum total protein	5.7 g/dl	5–8 g/dl
Serum albumin	3.2 g/dl	3.5–5.2 g/dl
Serum cholesterol	152 mg/dl	130–200 mg/dl
Serum triglyceride	70 mg/dl	40–200 mg/dl
AST	37 U/L	0–40 U/L
ALT	20 U/L	0–42 U/L
Serum alkaline phosphatase	453 U/L	48–406 U/L
Serum LDH	1020 U/L	0–480 U/L
Blood sugar	90 mg/dl	50–110 mg/dl
Pleural fluids investigations		
Pleural drainage cells	4700 cells (85% lymphocyte, 15 neutrophil)	
Pleural drainage glucose	60 mg/dl	
Pleural drainage cholesterol	80 mg/dl	
Pleural drainage triglyceride	230 mg/dl	
Pleural drainage total protein	4.5 g/dl	
Pleural drainage albumin	3 g/dl	
Pleural drainage LDH	1200 U/L	
Pleural drainage electrolytes		
- Sodium	- 136 mmol/l	
- Potassium	- 4.2 mmol/l	
- Chloride	- 112 mmol/l	
- Calcium	- 10.18 mg/dl	
Pleural drainage culture	no growth	
Other investigations		
Serum TORCH	Negative	
Blood culture	no growth	
Karyotyping	46, XX	

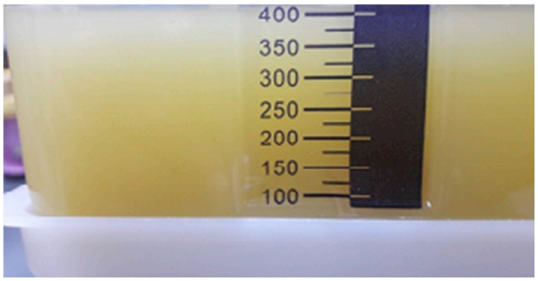
3. Discussion

Multidisciplinary approach before the delivery, the anticipation of diagnosis, planning of delivery by cesarean section and neonatologist



Picture 4. When the baby reached full feeding by expressed breast milk, chest drainage started to become milky in color (picture 4–5). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

attendance at delivery with the availability of suitable neonatal resuscitation equipment and trained staff are all important factors contributed to proper management, reduced morbidity and mortality [6,7]. Chest drain fluid was a characteristic of chylothorax. It was milky in appearance and the composition of electrolytes was the same as serum. The total protein content was 4.5g/L (the protein contains of chyle is usually >3g/L), and contains 4700 cells of which 85% lymphocyte and 15% neutrophils which is consistent with chyle. Chyle has absolute cell count >1000 cells/L and the lymphocyte count ranges from 400 to 6800/mm^3 [8]. Pleural drainage triglyceride was 230mg/L which is also diagnostic of chylous fluid which tends to be more than 110mg/dl [9]. Physical examination was normal, blood culture and pleural drainage culture did not show any growth. Brain ultrasound, echocardiography,



picture 5. Chest drainage turned into milky shape.

and abdominal ultrasound were normal. Karyotyping showed normal female, karyotype 46,XX. When the case was managed initially by IV octreotide [10,11] and oral sildenafil [12], the chylothorax drainage decreased but did not stop completely. Then the baby has kept nothing per mouth and TPN was added to regimen but the chylothorax drainage continued. The case was successfully treated by octreotide and MCT based formula milk (pregestimil). Adding of pregestimil to previous regimen enhanced recovery and improved baby's health resulting in successful treatment and stopping of the chylothorax [13]. Medium-chain triglyceride (MCT) is easily absorbed across the intestinal mucosa and delivered to the portal vein without going through the intestinal lymph vessels. The MCT based formula reduces accumulation of chyle in the pleural space without going through the intestinal lymph vessels and the thoracic duct. MCT oil-based diet decreases long-chain fat usage, which is absorbed and transmitted through lymphatic vessels. Reduction of long-chain fat usage leads to a decrease in lymphatic pressure and lymphatic flow, thus helps in a decrement of chylothorax [14].

4. Conclusion

A female full-term newborn was antenatally diagnosed with severe right-sided pleural effusion, was successfully managed by a combination of multidisciplinary team approach, preparation and appropriate resuscitation and management. Physical and biochemical characteristics of drainage were typical of chylothorax. Physical examination and karyotyping were normal. The case was a rare congenital idiopathic chylothorax failed to respond to TPN, sildenafil and octreotide infusion. However, responded and totally recovered to regimen of octreotide infusion with MCT formula feeding (pregestimil).

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2019.100937.

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