A rare case of congenital chylothorax in a Palestinian neonate

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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. Apper 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...
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 6h 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.

| CBC | Result | Normal range |
|-----|--------|--------------|
| WBC | 30000  | 5000–100000/L |
| Hemoglobin | 16.5 g/dl | 10–16 g/dl |
| Platelet | 294 10^9/L | 150–450 10^9/L |

**Blood chemistry**

| Parameter       | Result   | Normal range |
|-----------------|----------|--------------|
| Serum urea      | 10 mg/dl | 2–7 mg/dl    |
| Serum creatinine| 0.3 mg/dl| 0.5–1.0 mg/dl|
| Serum sodium    | 138 mmol/l| 135–145 mmol/l|
| Serum potassium | 5.1 mmol/l| 3.5–5.0 mmol/l|
| Serum chloride  | 110 mmol/l| 95–110 mmol/l|
| Serum calcium   | 11.7 mg/dl| 4.5–6.5 mg/dl|
| Serum total protein | 5.7 g/dl | 3.5–4.5 g/dl |
| Serum albumin   | 3.2 g/dl | 3.0–4.5 g/dl |
| Serum cholesterol| 152 mg/dl| 140–200 mg/dl|
| Serum triglyceride | 70 mg/dl | 40–150 mg/dl |
| AST             | 37 U/L   | 0–65 U/L     |
| ALT             | 20 U/L   | 0–60 U/L     |
| Serum alkaline phosphatase | 453 U/L | 30–400 U/L   |
| Serum LDH       | 1020 U/L | 0–500 U/L    |
| Blood sugar     | 90 mg/dl | 40–110 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**

| Test         | Result |
|--------------|--------|
| 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...
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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