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Case Report

Congenital Chylothorax as a Cause of Non Immune Hydrops: A Case Report

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Abstract

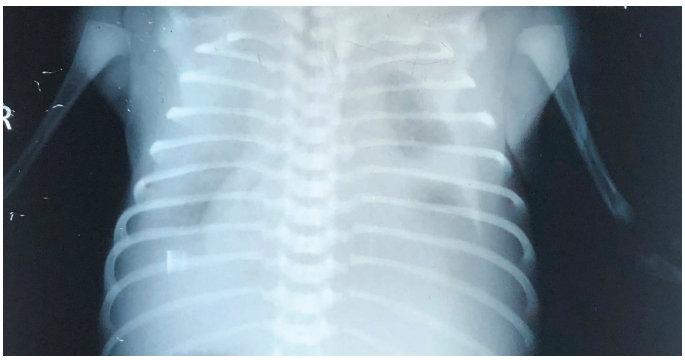
Chylothorax is a rare cause of non immune hydrops fetalis & presents with respiratory distress at birth. We present a late preterm diagnosed antenatally as hydrops with chylothorax with bilateral pleural effusion and respiratory distress at birth requiring mechanical ventilation. Baby was managed successfully with intercostal drainage, octreotide infusion & MCT milk formula.

Introduction

Chylothorax is accumulation of lymph / chyle in the pleural space. Congenital chylothorax, although uncommon, is the most common cause of pleural effusion in the neonatal period. It is classically a disorder of infants at or near term.[1] Most cases are idiopathic. Males are affected twice as frequently as females.[1] It occurs spontaneously due to lymphatic malformation or is associated with birth trauma to thoracic duct. It produces respiratory compromise, nutritional failure and immunological depletion contributing to sepsis.[2] The diagnosis of chylothorax is classically established by pleural fluid analysis and entails a triglyceride level > 110 mg / dl, pleural fluid: serum cholesterol ratio < 1.0, pleural fluid: serum triglyceride ratio > 1.0, > 1000 cells / mm^3 with marked lymphocyte predominance.[3] Congenital chylothorax if not diagnosed and treated on time, has poor prognosis. We report a case diagnosed antenatally at 36 weeks gestation with pleural effusion, pericardial effusion and ascites. Baby responded and chylothorax resolved after chest drainage, I/V octreotide and MCT formula milk.

Case Report

A 3.2 kg male newborn baby with a gestational age of 36 weeks was born to LSCS to a 30 year old second gravida mother from non consanguineous marriage. Antenatal scan done at 34 weeks was suggestive of hydrops fetalis (Fetal ascites, bilatera pleural and pericardial effusion) and mild polyhydramnios. The child did not cry at birth. Baby required resuscitation at birth. Apgar score at 1 & at 5 minutes was 3 & 7 respectively. Baby was kept on mechanical ventilator subsequently. On examination there was no facial dysmorphism or evidence of cardiac murmur. Bilateral pleural effusion was detected on chest X ray after delivery which was confirmed on chest ultrasound (Fig 1).



Bilateral chest tubes were inserted and 150 ml fluid was drained. The total protein content in the chylous fluid was 14.3 g / L. Pleural fluid triglycerides (TG) and cholesterol were 391.4 mg / dl and 19.4 mg / dl respectively, serum TG and cholesterol were 124.9 mg / dl and 108 mg / dl respectively. Thus, pleural fluid : serum cholesterol ratio was < 1.0 and that of pleural fluid : serum TG ratio > 1.0. It had a total cell count of 15000 / mm^3 with lymphocyte predominance. CBC, PBF was not suggestive of haemolysis. CRP, reticulocyte count and thyroid profile were normal. Serology was negative for TORCH infections. Cranial ultrasound was normal and abdominal USG revealed ascites. ECHO was suggestive of trivial tricuspid regurgitation and minimal pericardial effusion. Injection octreotide was started @ 1 microgram / kg / hour at fifth day of life and gradually increased to 10 ug / kg / hour. Along with this, feeding with very low fat formula milk mixed with MCT powder was started. 80 - 100 ml / kg / day of chylous fluid was drained daily which was replaced with equal amount of NS. Pigtail insertion was done on the right side after confirming a moderate amount of pleural fluid and 120 ml chylous fluid was drained. The daily chest tube drain output reduced gradually and stopped by 30 days. Antibiotic therapy was tailored according to blood culture reports and culture of ET secretions. Baby was weaned off mechanical ventilation after 30 days. Chest tubes were removed after 35 days. Octreotide was tapered and stopped by day 38. Repeat ultrasound chest and abdomen were suggestive of no residual fluid. Child was discharged at day of life 48 with MCT formula along with breast milk and is doing well on follow up.

Discussion

Neonatal chylothorax is an uncommon condition associated with high health care utilisation. Its incidence ranges from 1 : 8600 to 1 : 10,000 live births with a high mortality of 20 - 60%.[4] It can be congenital or acquired, the latter most commonly occurs after damage to thoracic duct during surgery. Most cases of congenital chylothorax occur without a clear etiology, termed as "idiopathic congenital chylothorax." It may be associated with lymphangiomatosis, lymphangiectasia, CHD, chromosomal abnormalities (Turner, Noonan and Down syndrome), mediastinal malignancies or H- type TEF.[5] Rupture of the thoracic duct due to hyperextension of the spinal column or secondary to increased systemic venous pressure during birth may also be the underlying cause. Right sided chylothorax is more frequently observed. CECT chest may help in finding the anatomical cause of chylothorax which may detect lymphatic malformation. Lymphangiography and lymphoscintigraphy both require administration of contrast medium and are not easily available.[6]

Diagnosis of chylothorax is made by WBC`s of > 1000 / ul with lymphocyte predominance (> 70%) and a TG level > 110 mg / dl^3. Pleural fluid in our case had TG levels of 391.4 mg / dl and TLC count of 15000 / $mm.^3$

Polyhydramnios in chylothorax, as also seen in our case, is probably due to mediastinal and oesophageal compression with obstruction of physiological foetal swallowing. Polyhydramnios with increase in intra amniotic pressure may reduce drainage of pleural effusion and may lead to preterm labour.[7]

Routine management of chylothorax involves treatment of the cause, drainage of fluid by chest tube insertion and rarely surgery. Octreotide is a somatostatin analogue that causes mild vasoconstriction of splanchnic vessels, including those in venous hepatic flow. This results in decreased gastric, pancreatic, intestinal secretions as well as intestinal absorption.[4] However, side effects of octreotide have been reported in up to 14% neonates in the form of hyperglycaemia, NEC, transient cholestasis, transient hypothyroidism, injection site pain, nausea, vomiting, diarrhoea, constipation, pulmonary hypertension and systemic hypotension.[5,6] Our neonate had conjugated hyperbilirubinemia towards the latter part of the hospital stay which could be due to octreotide, sepsis and partial parenteral nutrition.

Review of literature shows mixed results after octreotide with resolution in some and failure in few of cases. Timing of initiation, dosage, duration and frequency varied markedly with longest duration being reported by Coulter et al at 151 days.[8] It has been used at a dose of 0.3 ug / kg / hour for upto 10 days[8]. Coulter et al reported use of IV octreotide at 6 ug / kg / hour for 16 days followed by 0.05 ug / kg / dose thrice daily subcutaneously for 151 days.[8] Unresponsive cases are treated by surgery after 10 days. In our case we gave inj octreotide for 33 days with maximum dose of 10 ug / kg / hour which was gradually tapered. Antenatal management of chylothorax comprises pleuroperitoneal or pleuroamniotic shunt to prevent pulmonary hypoplasia. Optimal antenatal management and timing are still controversial. Surgical approach includes thoracoscopic pleurodesis, pleuroperitoneal shunt, ligation of thoracic duct and creation of thoracic duct to azygous vein anastomosis.[5] Only a minority of patients require surgical intervention and there are no clear evidence-based guidelines as to when such interventions should be used.[9]

MCT is easily absorbed across intestinal mucosa and delivered to portal vein without going through intestinal lymph vessels and thoracic duct. Reduction of long chain fat decreases lymphatic pressure, thus helping in decrement of chylothorax.[10] Poor neonatal outcome is associated with hydrops, bilateral effusion, pulmonary hypoplasia, preterm labour and absence of prenatal treatment.

Conclusions

Neonatal chylothorax as a cause of nonimmune hydrops is uncommon. Treatment is quite cumbersome, prolonged and includes stabilisation with mechanical ventilation, pleural drainage, fluid, electrolyte replacement, MCT feeds, administration of octreotide, anticipation and aggressive management of infections, prevention of nutrient deficiencies and prolonged hospital stay. Prospective registry of chylothorax patients and a multicentre randomised controlled trial to assess timing, duration,

dose, safety and efficacy of octreotide is the way forward.

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