International Journal of Contemporary Pediatrics
Sreedevi R et al. Int J Contemp Pediatr. 2019 Sep;6(5):2203-2205
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20193699

Case Report

Congenital chylous ascites: a case report

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Received: 09 August 2019
Revised: 17 August 2019
Accepted: 20 August 2019

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ABSTRACT

Due to the rarity of congenital chylous ascites and the lack of standards in diagnosis and therapy, this disease constitutes a medical challenge and individual therapy seems to be extremely important. A late preterm newborn with antenatally diagnosed ascites was born and chylous ascites was diagnosed after feeds were started. The baby was managed initially with nil per oral, parenteral nutrition and octreotide, followed by adding MCT formula feeds. Considering the rarity of neonatal chylous ascites and the non-uniformity in management plans and follow up, more case reports need to be published. Also, MCT formula, the main stay of management has to be discontinued as soon as possible with gradual introduction into breast feeds or normal newborn formula milk as long chain fatty acids are essential for optimal brain growth in newborns.

Keywords: Chylous ascites, Long chain fatty acids, Medium chain triglycerides, Newborn, Octreotide, Parenteral nutrition

INTRODUCTION

Congenital chylous ascites (CCA) is a rare condition which results from abnormal development of the intra-abdominal lymphatic system. It is defined as the collection of milky fluid rich in triglyceride content inside the peritoneal cavity in children younger than three month of age.[1,2] CCA can be primary or secondary. While primary is due to mal-development of intra-abdominal lymphatic system, secondary is due to neoplastic or inflammatory processes.[3] Bits of Congenital chylous ascites (CCA) constitutes a medical challenge and individual therapy seems to be extremely important due to the lack of standardized management guidelines.[4] This article presents a case report of a newborn with primary congenital chylous ascites and a brief account of the current knowledge on the disease.

CASE REPORT

Preterm, female baby born at 36 weeks of gestation to a 28 yrs old primigravida mother (parents non consanguineous Indian origin) by cesarean section with birth weight 2.38 kg. Maternal history of Gestational diabetes on insulin and antenatally detected fetal ascites. No history of oligohydramnios. Baby had mild abdominal distension. Ultrasound scan (USG) of abdomen done soon after birth showed mild ascites with low level internal echoes. Both kidneys and pelvicalyceal system were normal. Under ultrasonic guidance about 8ml of ascitic fluid was tapped from the right lumbar region. Aspirate was straw colored and cloudy. Ascitic fluid was analyzed for counts, triglycerides, protein, amylase and LDH which were within normal limits. Ascitic fluid and blood cultures was sterile. Feeds were started on day 2 and gradually increased. Abdominal distension increased

from day 4 onwards, and USG abdomen showed moderate ascites. On day 5, Ultrasound guided tapping of 150 ml thick yellow fluid was done. Analysis of this fluid showed triglycerides 3304mg/dl, which was diagnostic of chylous ascites. Following that the plan of treatment was to keep the baby nil by mouth, start parenteral nutrition and trial of medium chain triglycerides (MCT) formula feeds once it is available (unavailable locally and had to be imported). Octreotide infusion was started on day 8 and gradually increased from 1mcg /kg to 10mcg/kg over the next few days. By day 12 because of the increasing abdominal distension, MRI abdomen was done which showed moderate ascites but no major abnormality in the lymphatic system. About 100 ml was drained following that. MCT feeds were started on day 16 and gradually increased to full feeds. USG abdomen had shown decreased quantity of fluid and hence octreotide was also tapered and stopped by day 36. Repeat USG abdomen showed minimal fluid and baby was discharged on day 40 of life.

At 2 months of age, baby was readmitted and shifted over from MCT formula to breast milk and supplementary term formula. No sonological evidence of reappearance of ascites was noticed. Baby is followed up regularly at the out-patient clinic with normal growth and development.

DISCUSSION

Chylous ascites in newborn, usually due to developmental abnormalities of the lymphatic system, presents with vomiting, scrotal or vaginal oedema from the presence of the fluid in the peritoneal cavity, increased pressure in the abdominal cavity and respiratory failure along with malabsorption of nutrients due to abnormal function of the intestinal tract.[4]

A key stage in the diagnostics of abdominal chylous ascites is ultrasound guided paracentesis. On aspirating, lymph is a cloudy milky or straw-colored fluid (the colour depends on the diet). A typical feature of the lymph is a high concentration of triglycerides [over 1000 mg/dl], significantly exceeding the levels found in the serum. The presence of chylomicrons is considered to be pathognomonic.[5]

Lymphangiography and lymphoscintigraphy are methods to show the site of lymph leakage. Invasive techniques – laparoscopy and laparotomy, remain a diagnostic and therapeutic tool in cases resistant to conservative treatment.[6]

Treatment

Treatment of abdominal chylous ascites is a multidirectional process. Diet based on specialist formulas rich in MCTs is a popular conservative method of treatment, considered as a first-line intervention. Its disadvantages include: a long time of use, i.e. from 4 to 6 months, necessary for the therapeutic effect, as well as estimated success rate of about 30%.[7] Next stage is the withdrawal of enteral nutrition, with introduction of parenteral nutrition. Octreotide is used in the treatment of diseases accompanied by lymph accumulation in the pleural cavities and peritoneal cavity. Case reports show reduction and regression of chylous ascites, observed as early as after a few days following octreotide infusion.[8] A probable mechanism of action includes a decrease in lymph production and its flow rate. Possible adverse effects of the therapy include disorders of carbohydrate metabolism, nausea, diarrhea, abdominal distension, and abnormal liver function and a potential relationship between octreotide and necrotising enterocolitis, as well as pulmonary hypertension.[9] Surgery is advised in cases resistant to conservative treatment, which is usually undertaken for 4-8 weeks. The use of fibrin glue, especially in the situation of multiple lymph leakage sites, seems to be a successful method supporting the classic surgical techniques.[10]

CONCLUSION

In the index case, baby responded well to conservative management. The source of the ascites could be a congenital defect in the lymphatic system which underwent spontaneous repair. Rather than going into detailed and invasive processes to diagnose the exact etiology of the origin of ascites, our plan was to try conservative treatment and go for further diagnostic procedures only if the conservative line fails. Especially in resource poor countries, this could be an easier and better alternative.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not Required

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Cite this article as: Sreedevi R, Paul J, Jose G, Majeed A. Congenital chylous ascites: a case report. Int J Contemp Pediatr 2019;6:2203-5.