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CASE REPORT

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Congenital chylous as cites in a 3-month-old infant in Zaria: a case report

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Abstract: Congenital chylous ascites (CCA) is a rare disease that results from the mal-development of the intra-abdominal lymphatic system. 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 3-month-old girl diagnosed with congenital chylous ascites. She was managed initially with nil per oral, parenteral nutrition, medium chain triglyceride (MCT) containing oil and abdominal paracentesis, followed by octreotide. Medium chain triglyceride formula, the main stay of management was discontinued with gradual reintroduction of breast feeds. This case was selected due to the rarity of CCA and the lack of standards in the diagnosis and therapy.

Keywords: Chylous ascites, infant, medium chain trigycerides, octreotide, parenteral nutrition

Introduction

Congenital chylous ascites (CCA) 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]It can be primary or secondary.[3,4] Primary CCA, is due to mal-development of intra-abdominal lymphatic system, while the, secondary is due to neoplastic or inflammatory processes. Congenital chylous ascites constitutes a medical challenge and individualized therapy seems to be extremely important due to the lack of standardized management guidelines.[5] We present a case report of a 3-month-old girl with primary congenital chylous ascites and a brief account of the current knowledge on the disease.

Case Summary: B R

A 6-months-old girl who was first admitted at three months of age with a history of weight loss and body swelling of two months duration, difficulty breathing and fever of six weeks duration.

She was noticed to be losing weight progressively despite a good appetite. She was given bottled water in addition to breastfeeding on demand. Each breastfeeding session lasted 5 to 10 minutes. A breast milk substitute (correctly constituted) was introduced at 6 weeks of age due to weight loss and this was given thrice daily, about 30mls per feed. She had no history of cough or undue sweating. There was no known contact with individuals with chronic cough. She received BCG vaccine on the 7thday of life.

Body swelling was noticed at about, the same time. It began from the abdomen and slowly progressed over a two-week-period to involve the lower limbs. Limb swelling was painless and slowly progressive till presentation.

She had occasional bouts of non- projectile, non-bilious and non-bloody postprandial vomiting which persisted till presentation. No jaundice, diarrhoea or constipation was noticed. Stools looked normal and there was no bleeding from body orifices, history suggestive of skin itching or eruptions. Urine also appeared normal in colour and volume.

This was her first experience of body swelling. There was no antecedent history of trauma to the abdomen or abdominal surgery. Family history did not reveal similar illness, renal disease, visual or hearing impairment.

Difficulty in breathing is not associated with bluish discolouration of the lips.

Fever started two weeks prior to presentation described as low grade and continuous and persisted till presentation.

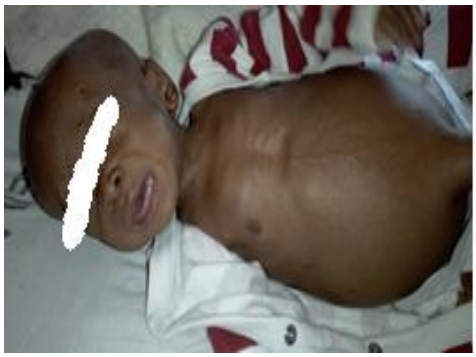
The baby was given over the counter medications (paracetamol and other syrups) and occasionally herbal concoction (made from soaked back of trees and roots). She was brought to our facility on worsening of symp-

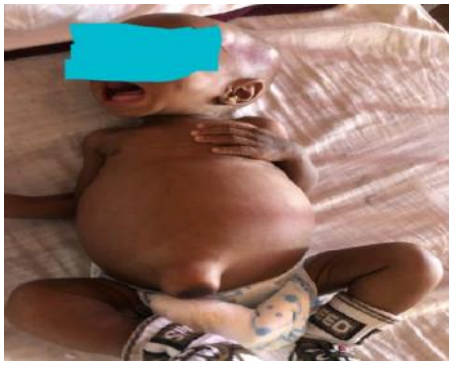
toms.
She is a product of term gestation, supervised at a private health facility. Pregnancy was not adversely eventful and obstetric scan done at booking showed only normal findings. Delivery was normal and the baby.
Cried immediately after birth. Her birth weight was 2.5kg. She passed meconium and urine on the first day of life and has been passing stool and urine normally since then.

At three-months, she was acutely ill, wasted, moderately pale with obvious abdominal distension, bilateral pitting lower limb oedema and sacral oedema. She was anicteric, afebrile(35.8^{oc}), well hydrated and had no dysmorphic facies.
Weight(4.2kg) and length (55cm) both below fifth percentile.
Abdomen was uniformly distended with a girth of 44cm, had visible distended anterior wall veins, an umblical hernia with a 3cm ring defect and a right inguinal hernia of about 3 x 3cm dimension. Ascites was demonstrable by fluid thrill and bowel sounds were normoactive. She had normal female external genitalia. Digital rectal examination showed normal sphincteric tone with well-formed stool. Urine was amber coloured, urinalysis showed normal findings.
Respiratory rate was 62cpm (tachypnoeic) and she was dyspnoeic,but breath sounds were normal. Cardiovascular findings were normal.

Abdominal ultrasound scan showed massive ascites with normal liver, spleen, kidneys and gall-bladder. Abdominal paracentesis yielded milky ascitic fluid with biochemistry as follows: total protein=35g/l, albumin=12g/l, cholesterol of 4.1mmol/l and triglycerides of 6.1mmol/l (540mg/dl) confirming chylous ascites. Abdominal CT scan confirmed normal abdominal organs with massive ascites. She had a haematocrit of 28% and total leucocyte count of 23.9x10^{9}/l. Blood biochemistry was essentially normal; except for hypoalbuminaemia (18g/l). Sepsis screen and tuberculosis screen were negative. Lymphoscintigraphy was not done due to its unavailability.
She was admitted, had blood transfusion and was placed on bowel rest with parenteral nutrition, prophylactic antibiotics and medium chain triglyceride oil and micronutrient supplementation. Abdominal girth decreased from 44cm to 40cm within one week. Enteral feeds were successfully recommenced without increase in abdominal girth. Lower limbs oedema completely resolved and the child was discharged home after three weeks of admission.

She was however re-admitted two weeks later due to recurrence of abdominal distension and respiratory distress. Enteral feeds were again withdrawn, and parenteral nutrition was reinstituted, She was also managed with intermittent abdominal paracentesis and prophylactic antibiotics. Subcutaneous octreotide 40µg 8hourly (at 1µg/kg/hour) was commenced on the 14th day of admission.
Abdominal girth gradually reduced to 38cm and remained normal after re-commencement of enteral feeding (breast milk) three weeks into admission. MCT based formula was not available hence child was introduced to regular formula at six months of age. She achieved neck control at four months and was able to sit with support at six months.
She was discharged on subcutaneous octreotide after seven weeks of admission and is still being followed up at the Paediatric Gastroenterology Clinic.





Discussion

Congenital chylous ascites is rare. It results from leakage of lipid rich lymph into the peritoneal cavity. This usually occurs due to trauma and rupture of the lymphatics or increased peritoneal lymphatic pressure secondary to obstruction. The most common (45 to 60%) cause is malformation of the lymphatic vessels which can be either atresia or stenosis of the major lacteals.[1,6]
Abdominal paracentesis is the most useful diagnostic step[4,5] The findings in our patient were consistent with the diagnosis of chylous ascites. Our patient was diagnosed at the age of three months. Huang et al diagnosed chylous ascites in a 50-day-old male baby[2] while, Mahmoud et al diagnosed three neonates antenatally at 34, 35 and 37-weeke gestation resp0ectively.[1]
The earlier the diagnosis, the better the prognosis.[1]
Previous investigators suggested that CCA should be treated conservatively at first. [7] In a series of 103 Japanese patients 63.5% were cured by conservative treatment. The average treatment was 63.5-days.[8] The goal of conservative treatment is to provide "gut rest" and decrease the intestinal secretions, conservative manage-

ments includes enteral feeding with a formular high in medium chain triglyceride (MCT), low in long chain triglyceride and enhanced in protein contents, and repeated abdominal paracentesis,even just clear water have been shown to increase the lymphatic duct lymph flow.[8] Liao et al[9] reported one case of neonatal chylous ascites that was put on strict MCT formular. The abdominal girth continued to increase and warranted readmission at 8-month of age. Theoretically TPN is superior to enteral feedings because the bowel is bypassed. Abdominal paracentesis can rapidly relieve respiratory insufficiency and abdominal discomfort. However, it may induce critical losses of fluid, lymphocytes, proteins, coagulation factors and antibodies, hence increases chances of infections, reduces immune function and may results in more ascites. It did happen in our patient. Some studies suggested prolonged non operative management avoiding paracentesis if possible. Surgical intervention was recommended if 1 to 2-months of conservative approach failed.

The success of the operation depended on identifying the site of leakage of the lymph duct.[1,5,8] In our patient, total parenteral nutrition combined with intravenous octreotide provided satisfactory relief. In this case, a marked reduction of chylous ascites was observed after octreotide infusion and abdominal girth returned to normal within 14- days. These results suggested that TPN along with octreotide can stop lymphatic leakage and clinical symptoms improved rapidly and effectively. 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, diarrhoea, abdominal distension and abnormal liver function.

Caty et al have reported a successful treatment of congenital chylous ascites by a somatostatin analogue.[6]

After discharge from the hospital, patient is supposed to be followed up in the hospital clinic to observe for increasing abdominal distension and growth monitoring which was done in our patient.

Conclusion

Congenital chylous ascites is a complex condition which can be managed as conservative or surgical. Our patient responded well to conservative management only. Our plan was to try conservative treatment and go for further diagnostic procedure only if the conservative line fails.

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