Case Report

Management of Chylous Ascites with Surgery and Frusemide in a New Born

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Abstract

Chylous ascites is the accumulation of milky fat-rich chyle in the peritoneal cavity. It is a rare condition in children. Congenital malformations of the lymphatic vascular channel are the predominant cause in children. Diagnosis is often confirmed by paracentesis of the chylous ascitic fluid. Treatment is generally conservative except in rare refractory cases that surgery is required for closure of the leak site or excision of the lymphatic cyst. Here is a report on the management of chylous ascites in a 6-h-old girl presenting in acute respiratory distress with acute abdominal distention due rapid increase in chylous ascitic fluid volume that was managed with surgery and frusemide.

Keywords: Acute abdominal distention, chylous ascites, frusemide, surgery

Introduction

Chylous ascites is a rare clinical entity in children.[1] Incidence of about 1 in 20,000–1 in 187,000 admissions has been reported in a large tertiary hospital.[2,3] Most reports showed a slight male preponderance, ratio of 1.8:1 M>F.[4] The first report of chylous ascites was in a 2-year-old boy with tuberculosis by Morton in 1683.[5] Many aetiological factors have been described in adults, but the predominant cause in children is mainly due to congenital developmental disorders of the lymphatic channels, which is usually a hyperplasia or hypoplasia of the lymphatic channels.[6] Of which, hyperplasia of the primary lymphatic channels is responsible for majority of the congenital malformations. Rarely, chylous ascites may follow birth trauma, malignancy, surgery and cardiovascular diseases in children. And sometimes, it can also follow certain genetic syndromes like Turner syndrome, yellow nail syndrome and Klippel–Trenaunay–Weber syndrome.

In general, chylous ascites is asymptomatic except in rare cases of rapid increase of the lymph volume or superimposed bacterial infection that will present with obvious clinical symptoms of respiratory difficulty or peritonitis.[7]

This is a report on the management of chylous ascites in a 6-h-old girl presenting in respiratory distress with acute abdominal distention due to rapid increase of chylous peritoneal fluid that was managed with surgery and frusemide.

Case Report

A 6-hr old girl that was delivered after 39 weeks of pregnancy at an internally displaced persons camp by a 25-year-old primipara was brought into the special baby care unit of the University of Maiduguri Teaching Hospital in respiratory distress with massive abdominal distention. She refused breast milk and had about three episodes of nonbilious vomiting before presentation. The delivery was however spontaneous, and no history suggests it was prolonged. The pregnancy was uneventful, booked at 25 weeks' gestation at the camp clinic. She passed meconium within 4 h of her presentation through a normal anal opening. On examination, she was otherwise healthy, weighing 3.2 kg, with a heart rate of 150 beats per mins, respiration 68 cycles/min with flaring of the alae nasi, axillary temperature of 36.4°C of mercury. Her abdomen was grossly distended. There was no visible peristalsis. The

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abdominal girth was about 55 cm measured two-finger breadth above the umbilicus, Figure 1. No mass was palpable per abdomen. Ascites was demonstrated by fluid thrill. The bowel sound and the rectal examination were unremarkable. She had normal female external genitalia.

The packed cell volume was 58%, white blood count was 7.07 × 10 cells/mm^3, neutrophil 60%, lymphocyte 14%, eosinophil 3% and basophil 1%. Her liver function test and retroviral screen in the mother was normal. Abdominal ultrasound scan showed massive turbid free intraperitoneal fluid collection with floating bowel loops. The plain abdominal radiograph revealed free intraperitoneal fluid without evidence of intestinal obstruction, Figure 2. She had paracentesis which showed milky peritoneal aspirate.

Biochemical analysis of the ascitic fluid showed triglyceride level of 200 mg/dl, glucose 8.2 mmol/l and and protein 60 g/l. Microbiology of the ascitic fluid yielded no bacterial growth.

The patient was electively placed on intranasal oxygen support, prophylactic antibiotics (ceftriaxone and metronidazole) and 10% dextrose water.

She had exploratory laparotomy on the 5th day of admission. At laparotomy, the entire peritoneum was filled with milky peritoneal fluid, about 530 mls was drained, Figure 3a. The surfaces of small bowel are lined with dilated lymphatic channels, and there were multiple enlarged whitish lymph nodes on the mesentery, Figure 3b. The liver, spleen, pancreas and the kidneys were otherwise normal. Exploration of the retroperitoneum revealed no obvious point of leakage or opening.

The mesenteric lymph node was biopsied, and the peritoneum was copiously lavaged with normal saline and closed. The immediate postoperative abdominal girth was 34 cm. The immediate postoperative period was uneventful. However, abdomen gradually started distending after breastfeeding commencing breast milk, and by the 10th day on breast milk the abdominal girth was 38 cm, Figure 4a. She was allowed breast feeding to continue but commence on low-dose frusemide 0.5 mg/kg. She responded very well to the frusemide as evident by the gradual decrease in the abdominal girth which was 36.2 cm by the 2nd week of its commencement before she was discharged.

Her follow-up serum albumin and protein after 2 weeks were 34 g/L and 60 g/L, respectively. Moreover, the ultrasound scan

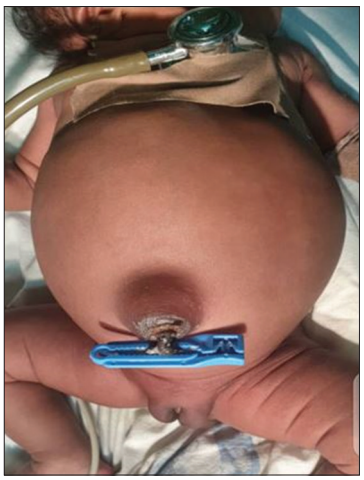

Figure 1: Abdominal distention from massive accumulation of peritoneal fluid

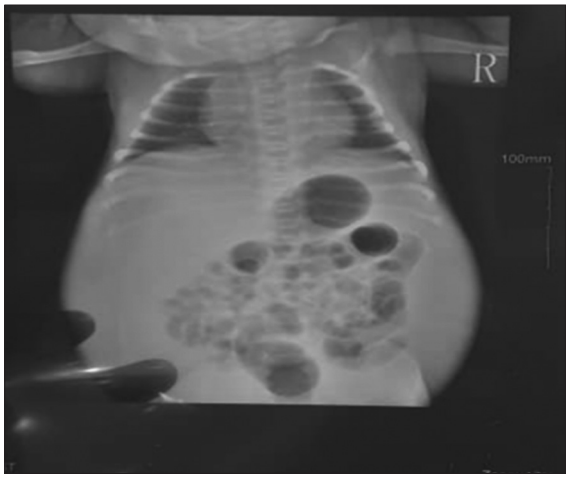

Figure 2: Plain abdominal radiograph showing massive intraperitoneal fluid without evidence of bowel obstruction

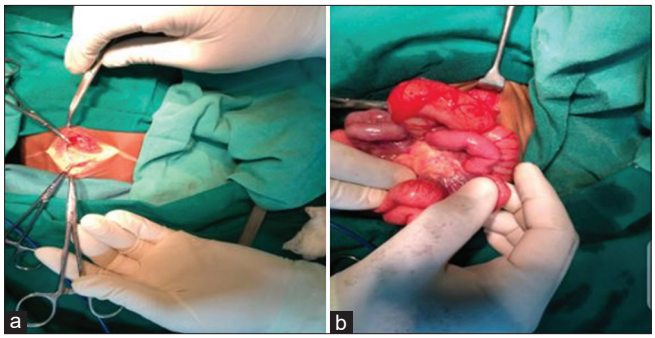

Figure 3: Intraoperative findings, (a) showing accumulation of milky peritoneal fluid, (b) dilated mesenteric lymph nodes

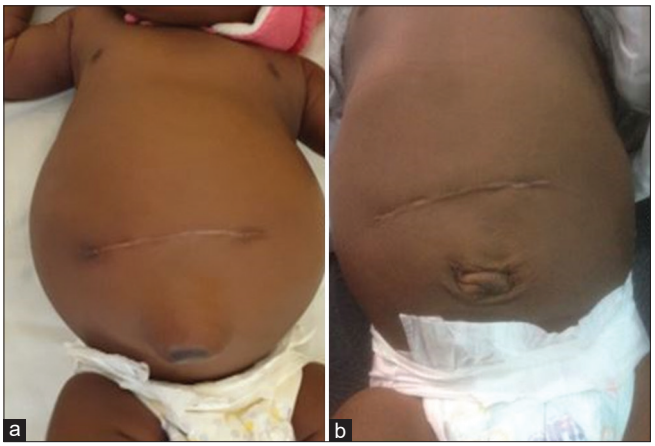

Figure 4: Postoperative abdominal distension (a) before commencement of frusemide (b) at first clinic visit

showed normal bowel loops with clear peritoneal fluid. The abdominal girth at then was 34.3 cm, Figure 4b. Pathological examination of the mesenteric lymph node predominantly showed lymphocytes. The frusemide was stopped at her second clinic visit. She afterward had uneventful followed up monthly for about 6 months before defaulting.

Discussion

The life-threatening clinical feature in chylous ascites is usually respiratory embarrassment caused by massive abdominal distention splinting the diaphragm as in index case. And sometimes, there would be associated vomiting, diarrhoea and loss of appetite. Vomiting and diarrhoea are usually the most frequent associated clinical manifestations seen in 28% and 17%, respectively.[8] Usually, this is due to increased susceptibility of the chylous fluid to infection. The principal pathogens are often *Escherichia coli, Pseudomonas aeruginosa, Staphylococcus epidermidis* and *Staphylococcus aureus*.[9] Rarely, patients may also have associated abdominal pain that usually follows tracking of the chylous fluid along the right paracolic gutter to accumulate in the right iliac fossa, therefore mimicking acute appendicitis. This is a very uncommon phenomenon in neonates.[10]

Neuroblastoma, Wilms' tumour, hydrometrocolpos, intestinal perforation, bladder perforation, congenital nephrosis or renal vein thrombosis were the differentials diagnosis we initially considered to be responsible for the massive abdominal distention in the index case. This was before ultrasound scan was able to narrow the diagnosis to chylous ascites. Afterwards, abdominal paracenteses were performed, which was able to confirm the diagnosis. Lymphangiography is usually the gold standard of diagnosis of chylous ascites, but it has been replaced with lymphoscintigraphy due to the attendant difficulty and risk associated with lymphangiography in neonates such as allergic reaction, fever, infection, inflammation of the lymphatic vessels. Lymphangiography and lymphoscintigraphy both visualise the leak site in chylous ascites. Magnetic resonance imaging and laparoscopy are other valuable tools in the diagnosis and of chylous ascites. Laparoscopy can be used for both diagnostic and therapeutic purposes.[11] A complete blood count and a retroviral screening is usually required in the clinical work of chylous ascites. This is to avoid missing the reported associations between lymphopenia, neutropenia, anaemia, human immune deficiency virus in about 0.5-1% of patients with chylous ascites.[12] The lymphopenia is probably due to sequestration of lymphocytes into the chyle, while neutropenia may be due to margination of the white blood cells rather than true neutropenia. Anaemia associated with chylous ascites is usually secondary to malabsorption of nutrients required for maintenance of red blood cell integrity, while HIV infection has been known to predispose infective chylous ascites in developing countries.[13]

The treatment of chylous ascites is generally conservative, usually for 4–8 weeks.[14] The treatment is aim at correcting the underlying cause of the chylous ascites, decrease the rate of chyle formation and provide low fat-high protein diet via parenteral route. The conservative measures are dietary restriction of low fat-high protein medium chain triglycerides in isolation or in combinations with somatostatin, octreotide or etilefrine agents that will usually decrease the rate of chyle fluid formation.[15] In the index case, breast milk was used in combination with frusemide, due to nonavailability of low-fat medium chain triglyceride in our setup.

Repeated paracentesis is not usually encouraged to decrease the rate of chyle formation due to peritonitis and hypoproteinaemia in chylous ascites. Surgery is usually employed if conservative measures fail. Surgery is employed to excise congenital mesenteric chylous cyst, close possible chylous openings or leak site. In the index case, no leak site or congenital chylous mesenteric cyst was found, as typical of majority of congenital chylous ascites in children.[16] However, we were able to copiously lavaged the peritoneum and biopsy mesenteric lymph node. The mesenteric lymph node biopsy will exclude infective or malignant aetiology of the chylous ascites. Furthermore, surgery is also expedient for insertion of peritoneovenous shunt, electrocauterisation of leaky lacteals and application of fibrin glue with absorbable mesh after dissection of the leaky zone for refractory chylous ascites.[17]

Conclusion

Nutritional requirement in chylous ascites can be in combination with a diuretic agent that will decrease the rate of chyle formation in absence of low fat-high protein diets. Furthermore, surgery may not always identify the cause of chylous ascites but may provide insight into the likely aetiology from the tissue biopsy. Repeated paracentesis should be discouraged due to risk of peritonitis and hypoproteinemia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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