Chylous ascites in an infant, a case report and literature review

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ABSTRACT

Background: Chylous ascites (CA) is very rare in the pediatric population. It is defined as whitish ascitic fluid with lymphocyte predominance. Congenital CA is probably the most common cause of CA in children. We report a case of an infant with CA; the cause of which remains unidentified, with full recovery. We have included a relevant literature review as well.

Case Presentation: A previously well 4-month-old baby girl was referred for worsening ascites. Despite extensive workup, no cause was found. Treatment options including diuretics, medium chain triglyceride formula (MCT) feeds, abdominal paracentesis, and total parenteral nutrition (TPN) were attempted. She recovered fully with no residual morbidity.

Conclusion: CA very rare in the pediatric population. Extensive workup excluded known causes and the yield was negative. Treatment is mainly supportive. MCT formula, fasting, and TPN were reported to have variable effects. Surgery could required in limited cases.

Keywords: Congenital ascites, chylous, TPN, abdominal distension, case report.

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Introduction

Chylous ascites (CA) is a rare condition with unclear etiology in most cases [1,2]. It is defined by the presence of whitish ascetic fluid rich in chylomicron and predominant lymphocyte count. It is seen mainly following damage to the lymphatic vessels or the thoracic duct following cardiothoracic surgery in addition to other rare causes (Table 1). Congenital chylous ascites (CCA) is probably the most common cause of CA in children.

Case Report

A 4-month-old female infant, previously well, was referred from the local hospital for worsening ascites. She was born by normal delivery. Antenatal scans showed Oligohydramnios, pericardial effusion, ascites, and hyper-echogenic bowel. Abdominal ultrasound and echocardiography post-delivery were normal. She was breast fed with bottle feeds top-ups. She was thriving and not on any medications. Immunizations were up to date and there was no family history of note. She remained well till 14 weeks of age when she presented to the local hospital with abdominal distension. Initial blood tests including full blood count, renal and liver profile, and electrolytes were normal, except for the C-reactive protein which was 51 mg/dL. An ultrasound scan (US) of the abdomen followed by abdominal MRI showed massive ascites and irregular liver margin with no abdominal or pelvic mass. She had a normal Barium study. Spironolactone was then started in addition to medium chain triglycerides (MCT) formula feeds which were not tolerated. On arrival at our hospital, she was not dysmorphic, non-icteric with a distended, soft abdomen, and 3 cm palpable liver, and huge ascites. Cardio-respiratory exam was normal. There was no peripheral oedema. MCT feeds were changed from bolus to continuous with fluid restriction. Full work up of ascites was performed (Table 1). The aim was to out rule common causes. These include metabolic causes, galactosemia, Alpha one antitrypsin deficiency, pancreatitis, congenital and acquired infections, and tumors. The abdominal tap showed chylous fluid with massive leukocytosis; 98% was lymphocytic. No organism was seen. Fluid Albumin was 29 mg/dL, Total Protein 41 mg/dL, Triglycerides 18.4 mg/dL (High), Fluid electrolytes, Lactate dehydrogenase, Amylase and cholesterol were all normal. She developed bronchiolitis required oxygen and was found to be Adenovirus positive on nasopharyngeal aspirate. Chest X-ray showed left-sided pleural effusion. Echocardiography showed trivial pericardial effusion. She underwent abdominal laparoscopy, which was converted to laparotomy to allow better visualization, and 1,200 ml

Table 1. *List of investigations and results.*

TEST	RESULT
FBC	Normal
LFT & coagulation screen	Normal
Renal function	Normal
AFP	18
Galactosemia screen	Negative
VLCFA	Normal
Transferrin Isoforms	Normal
Hepatitis screen	Negative
Karyotype	46XX
Faecal calprotectin	<50
Faecal elastase	Normal
Alpha-1 Antitrypsin phenotype	Pi M
Metabolic screen	Normal

of fluid was drained. No cause of ascites was identified, and two drains were inserted. Abdominal lymph node biopsies showed reactive changes. Losses dried up after a few days. She was kept fasting and started total parenteral nutrition (TPN). Two weeks later oral MCT formula was introduced slowly and was well tolerated. She was discharged home on MCT formula and fat-free solids. The abdominal US before discharge showed no re-accumulation of ascites. She was readmitted for upper and lower GI endoscopy after a dietary fat challenge. These were normal with no evidence of lymphangectasia. She remained well since with no recurrence.

Discussion

Ascites is defined as the pathological accumulation of fluid within the peritoneal cavity. It develops as a response to various pathological processes. While most adult cases are mainly secondary to malignancy, cirrhosis, or tuberculosis infection; the cause in most pediatric cases is rarely identified [1]. Press et al. [3] reported four cases of CA in children. Three were secondary to congenital lymphatic anomalies. An abnormal antenatal US is sometimes the only abnormality identified [3], as in our case. CCA was also reported in association with gut mal-rotation [4]. CA can also occur post liver transplant [5], following Sirolimus immunosuppression, post kidney transplantation [6], and with acute idiopathic pancreatitis [7]. Infections, such as tuberculosis, mycobacterium avium, and *Escherichia coli*, in addition to congenital infections such as parvovirus, CMV, and syphilis could rarely cause CA [8]. The diagnosis depends on the findings of paracentesis. Investigations yield could be minimal as described in our case. Lymphatic imaging, although used in adults is rarely indicated in children due to low diagnostic yield. Treatment is mainly supportive and is based on fasting and TPN for variable durations, diuretics, and MCT feeds. Very few patients require surgery. Some authors suggested that extensive investigations are not indicated in cases of CCA [3]. Surgery depends on the underlying diagnosis and must be selective. It should consider the severity of the condition. Surgical options include resection of localized lymphangiectasia, suturing of lymphatic fistula and even shunting.

Table 2. *Causes of Chylous ascites.*

- Cirrhosis - Up to 0.5% of patients with ascites from cirrhosis may have chylous ascites.
- Congenital defects of lacteal formation
- Chromosomal; e.g Turner syndrome
- Abdominal surgery or abdominal trauma
- Tumour related e.g. Lymphoma, carcinoid
- Infections e.g Spontaneous bacterial peritonitis or Abdominal tuberculosis
- Pelvic irradiation
- Peritoneal dialysis

Conclusion

CA is a very rare condition in the pediatric population. Diagnosis relies on the presence of confirmation of chylous ascitic fluid on abdominal paracentesis. Investigations should aim to exclude known causes including Turner syndrome, lymphangiectasia, lymphatic obstruction, mal-rotation, and infections. Imaging options include barium meal and follow-through, abdominal CT, and lymphatic scintigraphy, which is rarely required due to rarity in the pediatric population. Treatment is mainly supportive and is based on fasting, TPN diuretics, and MCT feeds. Our case represents one of very few cases in the literature of CA presented in infancy and although may be congenital we could not identify the underlying cause.

What is new

Chylous ascites is rare in infants. Extensive workup is required although it may not lead to a diagnosis. Treatment options are drainage, diuretics, MCT formula, TPN, and rarely surgery.

List of Abbreviations

CA	Chylous ascites
CCA	Congenital chylous ascites
MCT	Medium chain triglyceride formula
TPN	Total parenteral nutrition
US	Ultrasound scan

Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

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Consent for publication

Written informed consent was obtained from the patient/from the parents/from the next kin of the patient (if the patient is deceased or unable to provide consent for publication).

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	4-month-old female
2	**Final diagnosis**	Chylous ascites
3	**Symptoms**	Abdominal distension
4	**Medications**	Symptomatic treatment given
5	**Clinical procedure**	Abdominal paracentesis, TPN, Laparoscopy and Laparotomy
6	**Specialty**	Pediatric gastroenterology and neonatology