Surgical management of congenital chylous ascites

Ayman Albaghdady, Khaled M. El-Asmar, Mohamed Moussa and Sameh Abdelhay

Objective **The objective of this study was to present our experience regarding the diagnosis and therapeutic approach in cases of congenital chylous ascites that were surgically managed in our department in a 10-year period.**

Background **Congenital chylous ascites is a rare condition that constitutes a challenge for physicians. This condition is often refractory to therapy, and it is responsible for serious malnutrition and immunological deficiency because of losses of proteins and lymphocytes.**

Patients and methods **From 2007 to 2017, four cases of congenital chylous ascites were managed at our center. All cases were treated surgically after failure of medical treatment.**

Results **The first patient had no definite lymphatic leaks rather than presence of a moderate amount of milky ascitic fluid with dilated lymphatics at the root of the mesentery and at the intestinal wall and enlarged regional lymph nodes. The second one had leaking chylolymphatic cyst in the greater omentum that was excised. The third patient had no definite lymphatic leaks rather than thickened greater omentum, and thus, subtotal omentectomy was done. The fourth baby had no definite lymphatic leaks and developed reaccumulation of the ascitic fluid 1 month after surgery; insertion of a peritoneovenous shunt was indicated that succeeded to resolve the ascites. All children are growing up normally with no recurrence of the ascites on long-term follow-up.** ***Ann Pediatr Surg*** **14:56–59 © 2018 Annals of Pediatric Surgery.**

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Department of Pediatric Surgery, Ain Shams University, Cairo, Egypt

Correspondence to Khaled M. El-Asmar, MD, MRCS, FEBPS, 14 Mostafa Sadek El-Rafeay, Heliopolis, Cairo 11361, Egypt
Tel: + 20 100 122 2624; fax: + 20 483 0833;
e-mail: khaled.elasmar@med.asu.edu.eg

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Introduction

Congenital chylous ascites is an uncommon disease usually caused by obstruction or rupture of the peritoneal or retroperitoneal lymphatic glands. It is defined as the accumulation of chyle in the peritoneal cavity in infants younger than 3 months [1]. It is a serious disorder because of its remarkable mechanical, nutritional, and immunological consequences that resulted from the continuous losses of proteins and lymphocytes [2]. Many pathological conditions can result in this disease, including congenital defects of the lymphatic system; nonspecific bacterial, parasitic and tuberculous peritoneal infection; liver cirrhosis; malignant neoplasm; blunt abdominal trauma; and surgical injury [1]. The reported incidence of congenital chylous ascites is reported to be 1 : 20 000–1 : 187 000 live births [3]. Though the incidence of chylous ascites has increased in recent years, the treatment remains unsatisfactory in some cases because of prolonged duration of disease. Conservative treatment of chylous ascites, recommended in most patients, involves paracentesis, a medium chain triglyceride (MCT)-based diet, total parenteral nutrition (TPN), and recently used somatostatin therapy. Surgery is only recommended when conservative treatment fails [2]. Usually, the MCT-based diet is the first choice. TPN is recommended after dietary manipulation has failed, and somatostatin therapy is attempted only if chylous ascites has been refractory to all conservative measures. It usually takes several weeks to 2 months to close the lymphatic fistula adequately with routine conservative regimens [4].

Patients and methods

In this work, we retrospectively reviewed our department experience in managing patients presented with congenital chylous ascites during the past 10-year period. Three cases were referred to us for surgical intervention after failure of medical treatment, and one case was explored for excision of huge intra-abdominal cyst. Pediatric surgery department, Ain Shams University ethical committee approved to retrospectively analyze chylous ascites patients in the last 10 years.

Results

During the period between 2007 and 2017, four cases of congenital chylous ascites were surgically managed in the Pediatric Surgery Department, Ain Shams University Hospital.

The first case of a male infant weighing 4.3 kg presented at the age of 8 weeks with rapidly increasing abdominal distention from the first week of birth. Abdomen was grossly distended and measured 47 cm in girth with moderate bilateral hydrocele. He was dyspneic, and examination result of the cardiovascular and respiratory system was normal. Serum electrolytes, urea, and creatinine levels were normal. Hemoglobin and total blood count levels were normal. Total protein was 61 g/l, albumin was 37 g/l, globulin was 24 g/l, and total bilirubin was 10 mmol/l. Urine examination showed 0.1 g albumin/l. Chest radiograph result was normal. Ultrasound (US) of the abdomen showed large amount of fluid in the peritoneal cavity, inferior vena cava and hepatic veins were normal, and there was no organomegaly. Computed tomography-abdomen showed no abnormality rather than huge amount of intraperitoneal fluid (Fig. 1). Ascitic fluid analysis revealed total protein 22 g/l, glucose 5 mmol/l, triglycerides more than 110 mmol/l, cholesterol 19 mmol/l, and lactic

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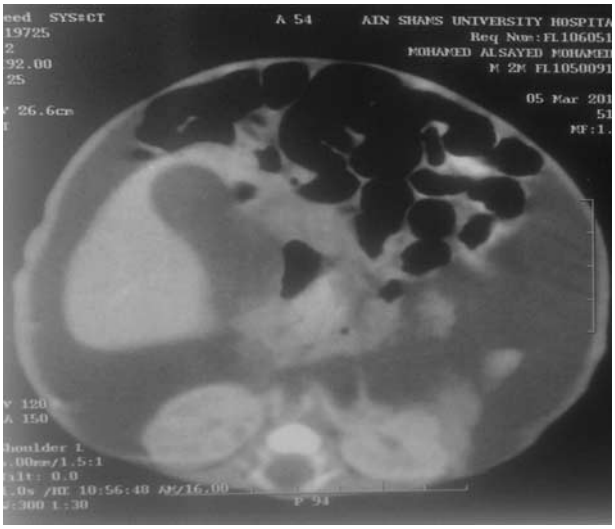
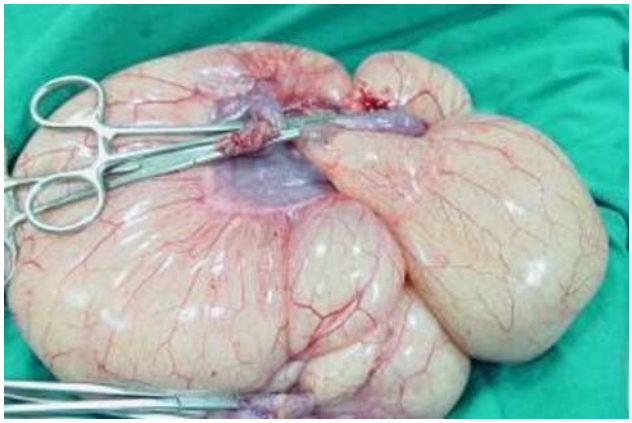
Fig. 1



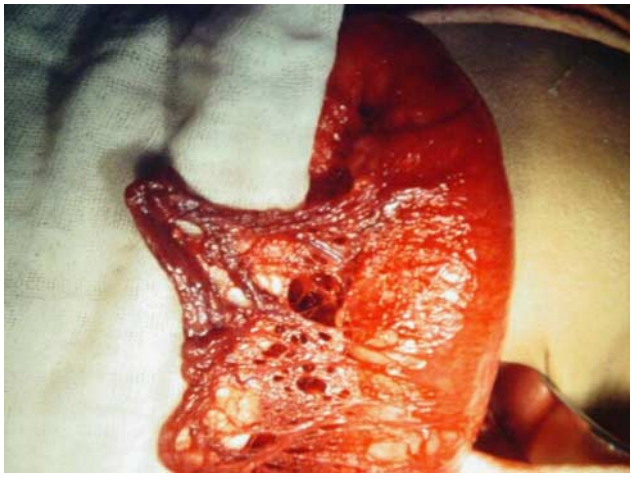
Abdominal computed tomographic findings show massive ascites without other abdominal abnormalities.

Fig. 2



Intraoperative findings of excised greater omental cyst.

Fig. 3



Intraoperative findings of thickened greater omentum.

dehydrogenase165 IU/l. Cytology of the ascitic fluid showed large number of lymphocytes. The child was given TPN for 10 weeks. During the initial period of treatment, paracentesis was performed to relieve dyspnea. Introduction of high MCT milk formula was attempted; however, the ascites did not resolve. Laparotomy revealed presence of a moderate amount of milky ascitic fluid and dilated lymphatics at the root of the mesentery and at the intestinal wall with enlarged regional lymph nodes. Post-operatively, he was given MCT formula exclusively for 4 months. As the ascites did not recur, normal diet was gradually introduced. At 2 years of age, he was well, with no recurrence of the ascites.

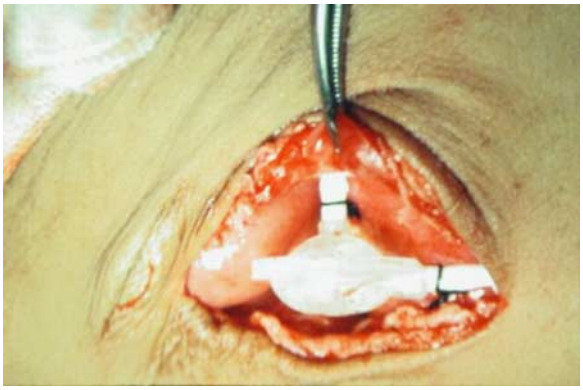
The second case was a 28-day-old full-term male baby who presented to the emergency unit because of abdominal colic and vomiting. Abdominal US revealed a cystic formation (45 × 32 mm) near the transverse colon, and MRI additionally showed edema of the intestinal wall. The ascitic fluid, obtained by paracentesis, revealed 75% lymphocytes and triglycerides of 4490 mg/dl. The patient had been subjected to exploratory laparotomy. The peritoneal cavity was filled with milky fluid. A leaking chylolymphatic cyst was found in the greater omentum (Fig. 2). The mesentery was thickened with dilated lymphatics. The cyst was completely excised with subtotal omentectomy. The pathologic examination showed thick, vascularized, fibrous connective tissue, consistent with chylolymphatic cyst wall. Postoperative period was uneventful. On the fourth postoperative day, the patient started MCT milk diet and discharged from the hospital 1 week later. Follow-up US (1st, 3rd, 6th, and 12th month postoperatively) showed no ascitic fluid. The baby shifted to breast milk fed from the seventh month of age, and he is growing up normally for 3 years of follow-up.

The third case was a girl born prematurely at 32 weeks of gestation by cesarean birth because of fetal ascites and hydrops. The ascitic fluid obtained by paracentesis was straw colored, with 8160 cells/l (85% lymphocytes). Diagnostic evaluation including pelviabdominal US and abdominal computed tomography showed a huge amount of ascitic fluid, and there were no suggestive findings of any other abnormality. Lymphoscintigraphy failed to reveal the site of leakage. Initial treatment was conservative, with TPN, octreotide, and multiple abdominal paracentesis. Finally, an abdominal drainage tube was inserted. The amount of ascetic fluid produced was 700 ml/24 h. Surgical intervention was decided at age of 50 days as there was no significant decrease of the daily chyle amount. During exploratory laparotomy, there was thickening of the greater omentum (Fig. 3) with no other abnormalities found intraperitoneal or retroperitoneal. Subtotal omentectomy was done, and a drainage tube was placed in the abdominal cavity. The amount of ascitic fluid dramatically decreased post-operatively. The child started MCT milk diet on the 10th day postoperatively and was discharged 3 weeks after surgery. Follow-up abdominal US at first, third, and sixth month after discharge revealed no reaccumulation

of ascitic fluid. The child has normal growth and has been fed normally since the eighth month of life with follow-up till 4 years of age.

Our fourth case was a 9-week-old male infant weighing 4.5 kg presented with rapidly increasing abdominal distention from the age of the 2 weeks. Abdomen was grossly distended, and the patient was dyspneic with normal examination of the cardiovascular and respiratory system. Abdominal US detected a considerable amount of ascites, and abdominal paracentesis was performed. The analysis of evacuated ascites showed the following: specific gravity 1.035, chylomicrons (+), cell count 6000/mm^3, mostly lymphocyte 82%, and monocyte 10%. Chylous ascites was diagnosed and TPN was administered, but the ascites persisted for 30 days. Repeated paracenteses were mandatory, and a considerable amount of fresh frozen plasma was given. Surgical intervention was done and revealed presence of a copious amount of milky fluid between the bowel loops, which was identified by the biochemical analysis as chyle. The intraperitoneal organs had no remarkable lesions, and the retroperitoneal space was explored by mobilizing the left and right colonic flexure. After doing this maneuver, a large amount of milky fluid was released from the retroperitoneal space; the exact source of which could not be identified. The operation was completed by a thorough lavage of the peritoneal cavity and the placement of drains (right paracolic gutter and pelvic space). After laparotomy, the patient received TPN and intravenous octreotide for 7 days. Chyle output from the drains was progressively diminished, and the patient was discharged from our department after 11 days with no postoperative complications. Patient presented after 1 month of discharge from the hospital with reaccumulation of the ascitic fluid again. Reexploration was done, and a peritoneovenous shunt (Denver Biomaterials Inc., Evergreen, Colorado, USA) was implanted (Fig. 4). The vascular end tube was placed in the superior vena cava through the right internal jugular vein. The ascites was resolved by the fourth postoperative day. The patient was subsequently discharged on the seventh day after shunt implantation. The patient developed chronic calcular cholecystitis 18 months later and was subjected to cholecystectomy. It was noted that the ascitic fluid had not developed for 18 months after the shunt implantation. The shunt was removed during the cholecystectomy surgery. There was no recurrence of ascites for 9 years of follow-up, and the patient has been doing well.

Fig. 4



Intraoperative findings of peritoneovenous shunt.

Discussion

Chylous ascites is uncommon in children. It is, however, relatively more common in infancy [5]. Congenital malformations of lymphatic system like congenital lymphangiomatosis, congenital chylous cysts, and atresia of the lymphatic ducts are responsible for 39% of cases [6,7]. Idiopathic condition accounts for 30% of cases, and this condition is said to have leaky lymphatics [8]. 'Leaky lymphatics' appears to be caused by delayed maturation or hypoplasia of lacteals which allows chyle to leak into the peritoneal cavity [8,9]. Conservative and symptomatic measures like TPN, high MCT diet, and repeated paracentesis form the initial management. In refractory cases, the administration of somatostatin or its analog octreotide is needed [10–15]. It is not completely understood how these drugs work in these cases. It is speculated that they reduce the absorption of fatty acids from the intestine and decrease the gastric, pancreatic and enteral secretions, the intestinal motility, and the visceral blood flow and so they decrease the production of chyle. It is also believed that these drugs inhibit specific receptors found in the normal lymphatic vessels of the intestinal wall, and they prevent the excretion of lymph [11,12,15]. The aim of conservative management is to provide time for the leaky lymphatics to get obliterated and for new lymphovenous channels to get established [9]. The time required for the conservative management to be effective in congenital chylous ascites is highly variable and may require 10 weeks or more [16]. If the conservative treatment is not sufficient to resolve the ascites and/or fails to provide sustained relief after the introduction of normal diet, surgery is the appropriate choice. It was found that 58% of children with intractable chylous ascites have a lesion amenable to surgical intervention [17]. In our series, there was no definite cause in three cases rather than thickened greater omentum in one case and greater omental leaking chylolymphatic cyst in another case. Important for the success of the operation is to recognize the leakage site [17]. Localization of the chyle leaking point intraoperatively is often difficult. Preoperative lymphangiography, lymphoid scintigraphy, or more simply and safer the oral administration of a lipophilic dye, – for example, Sudan Black 6 h before surgery may facilitate the identification of the responsible site of chylous leakage [17,18]. With the advance of imaging modalities, a noninvasive MRI lymphangiography that allows precise imaging of thoracic lymphatic vessels without contrast-enhancing agents is available now in some centers [19]. Recently, localization of leaking point and lymphatic duct ligation could be performed under laparoscopic guidance with minimal invasiveness. Laparoscopy may have the advantages of exploring the peritoneal cavity in its natural state, in addition to have a magnified image of the abdominal cavity [17,20].

When nonoperative and operative therapy fails, peritoneovenous shunts placement is indicated with some

success [21]. High levels of cell counts and protein contents in the chylous ascites increase the risk of shunt obstruction. Denver shunt has a valve mechanism, which prevents backflow of blood, and the valve chamber lies in the subcutaneous tissue can be compressed to promote flow and to relieve blockage. Other complications reported were cardiac disseminated intravascular coagulation, and perforation of the coronary sinus [22]. Sooriakumaran *et al.* [23] reviewed 11 children who underwent shunting over a 17-year period. In all of the children, ascites resolved except for a child of 4 years old with lymphohistiocytosis. They recommended elective removal of the shunt after 1 year as five shunts were removed 1–3 years after insertion without recurrence of ascites. In the presented case, ascites had not accumulated for 18 months, and the shunt was still patent and functioning. Another modality for management of refractory cases is the fibrin glue, which has been used recently to seal the area of lymph exudation in the abdominal cavity especially when the exact leaking point that is amenable to suture could not be identified [15,18,24,25].

Conclusion

Congenital chylous ascites is an uncommon pathology. It can be suspected early with antenatal US, and the diagnosis is confirmed postnatally by analysis of the ascitic fluid. It is desirable to give an adequate trial of conservative treatment specially if there is good response as laparotomy may not always reveal a correctable surgical abnormality. Surgery should be reserved for those who have an identifiable surgically correctable lesion or when conservative approach fails to provide sustained relief after the introduction of normal diet. When nonoperative and operative therapy fails, peritoneovenous shunts should be considered.

Conflicts of interest

There are no conflicts of interest.

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