

Congenital Chylous Ascites: A Rare Cause of Infantile Ascites Treated With MCT-Based Diet and Octreotide

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Abstract: Congenital chylous ascites (CCAs) are a rare disease that results from the accumulation of chylomicron-rich lymphatic fluid within the peritoneal cavity due to maldevelopment of the intra-abdominal lymphatic system. Medium-chain triglyceride (MCT)-based diet, total parenteral nutrition (TPN), and repeated paracentesis are considered supportive management for CCA. Cases unresponsive to conservative treatment usually require surgical intervention. We report a case of CCA in a premature neonate treated successfully with intravenous infusion of octreotide (synthetic somatostatin analog), after failing to respond to supportive therapies. Due to the lack of standards in diagnosis and treatment, this disease constitutes a medical challenge, and individual therapy seems to be noteworthy.

Key Words: Congenital chylous ascites, MCT-based diet, octreotide

INTRODUCTION

Congenital chylous ascites (CCAs) in infants are rare entities, conditioned by numerous factors and the changing dynamics of the disease. It results from the maldevelopment of the intra-abdominal lymphatic system, and causes accumulation of triglyceride-rich milky fluid (chyle) into the peritoneal cavity.¹ Because of the lack of diagnostic and therapeutic standards, this disease constitutes a medical challenge, and individual therapy seems essential.

CASE PRESENTATION

A 37-day-old-male infant born at 34 weeks to parents of consanguineous marriage was referred with the complaints of gradual abdominal distention along with scrotal swelling since the age of 20 days. The mother also complained of repeated vomiting for the same period. His weight (3.6 kg) and length (52 cm) were, respectively, at the third and tenth centile of the WHO growth standard. Mother was 26 years old primi, normotensive with no other medical complications (diabetes mellitus and hypothyroidism). However, she had a history of oligohydramnios based on prenatal ultrasonography (USG). On physical examination, the baby was vitally stable, had ascites and scrotal swelling but characteristically had no edema. The patient had no evidence of pleural effusion and cardiac anomalies. His other systemic examination revealed normal findings. USG of

the abdomen revealed moderate ascites and fluid collection in the bilateral scrotal sac. By doing abdominal paracentesis, about 150 ml of thick, milky white ascitic fluid was removed. Ascitic fluid analysis showed total count of WBC 180/mm³, Lymphocytes 77%, total protein 4.7 g/dl, triglyceride 5685 mg/dl, and lactate dehydrogenase 435 IU/L. The culture of ascitic fluid was normal. Infection screening, serum electrolytes, liver function test, and renal function test were normal.

The patient was diagnosed as a case of congenital chylous ascites; breastfeeding was stopped, and total parenteral nutrition was given for 48 hours. Then, the patient's nutrition was maintained by breast milk mixed with MCT oil from the third day of hospital admission. MCT-based formula milk, for example, Pregestimil, is not produced locally and is always imported at an exceptionally higher price in our country. Furthermore, during the COVID-19 pandemic, all exports were hampered. Hence, we used MCT oil mixed with breast milk as a diet for the patient. Our target was to achieve about 300–360 kcal/day (ie, 80 to 100 kcal/kg/day), containing approximately 23 g fat from a mixture of breast milk, MCT oil, and soybean oil.

All the oil components were mixed with the expressed breast milk and then fed to the baby. We also started fat-soluble vitamin supplementation (vitamin A, D, E, and K). However, the patient's clinical condition did not improve significantly. The baby was then started with continuous intravenous infusion of octreotide, on the fifth day of treatment, initially at a rate of 1 µg/kg/hour and gradually increased to 2 µg/kg/hour over 48 hours. His weight and abdominal girth were measured daily. The abdominal girth reduced from 43 to 35 cm over 5 days. Octreotide was continued in tapering dosage from day 7 and stopped on day 10. During the entire treatment course, no side effects of octreotide, such as diarrhea, hypoglycemia, or hypotension, were observed. Enteral feeding was well tolerated, and the baby was discharged home in good health condition.

On the last follow-up at the age of 4 months, the baby was well thriving on breastfeeding along with MCT oil and showed no recurrence of ascites. We adjusted the amount of feed according to patient's current body weight during follow-up and instructed the parents to continue this diet plan up to 6 months of age.

DISCUSSION

Congenital chylous ascites is a disease with a severe prognosis with etiology that varies according to age groups.¹ Chylous ascites in newborns are usually due to developmental abnormalities of the lymphatic system. It presents with vomiting, scrotal, or vaginal edema from the fluid in the peritoneal cavity.² Due to multifactorial etiopathogenesis, the diagnostic process should be individualized. A key stage in the diagnostics of abdominal chylous ascites is ultrasound-guided paracentesis. On aspiration, lymph is a cloudy milky or straw-colored fluid depending 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 in the aspirating fluid is pathognomonic. Diagnosis of malformation of the lymphatics is suspected when other etiological evaluations are negative. Lymphangiography and lymphoscintigraphy are methods to show the site of lymph leakage. However, only

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TABLE 1. Diet plan for the baby maintaining daily energy and fat requirements

Ingredient	Amount (daily requirement in ml)	Energy (approximately in kcal)	Proportion of Energy (% of daily requirement)	Fat content (approximately in g)	Proportion of Fat (% of daily requirement)
Breast milk	400–450	280–315	78%–88%	15–16	65%–70%
MCT oil (saturated fat)	5–7.5	38.3–57.5	11%–16%	4.7–7.0	20%–30%
Soybean oil (essential fatty acid)	2 (approximately)	7.2–10.8	2%–3%	1.0–1.1	4%–5%

**FIGURE 1.** A) Patient condition on admission. B) Ascitic fluid collected by paracentesis. C) Patient condition on the fifth day of treatment.**FIGURE 2.** Patient condition on follow-up at the age of 4 months.

one center in our country performs lymphangiography, which was not operating during the COVID-19 pandemic. Hence, we did not advise the patient for lymphangiography. Invasive techniques like laparoscopy and laparotomy remain a diagnostic and therapeutic tool in cases resistant to conservative treatment.³

Treatment of abdominal chylous ascites is a multidirectional process. MCT-based diet is a popular conservative treatment method, considered a first-line intervention. Its disadvantages include prolonged use (ie, from 4 to 6 months) necessary for the therapeutic effect and an estimated success rate of about 30%.⁴ The next stage is the withdrawal of enteral nutrition, with the introduction of parenteral nutrition. Octreotide is used to treat 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.⁵ A probable mechanism of action of octreotide includes a decrease in lymph production and its flow rate. It also decreases the absorption of fats and lowers the triglyceride concentration in thoracic duct.¹ Since its first successful use to treat the chylous effusion by Caty et al, marked variation was found regarding its dosing regimen, modes of drug administration, and duration of treatment in the existing literature.^{2,6–9}

In the reported case, the patient benefited from the administration of breast milk mixed with MCT oil and octreotide. Both Purkait et al¹ and Huang et al⁹ showed successful treatment with a similar regimen as ours. Purkait et al¹ started administering Octreotide at 1 $\mu\text{g}/\text{kg}/\text{hour}$ and gradually increased to 2 $\mu\text{g}/\text{kg}/\text{hour}$ over 48 hours on the seventh day with a significant reduction in the abdominal girth over three days. They continued Octreotide in tapering dosage from day 7 and stopped on day 10.¹ Huang et al⁹ started Octreotide at 0.5 $\mu\text{g}/\text{kg}/\text{hour}$ and gradually increased to 2 $\mu\text{g}/\text{kg}/\text{hour}$ on day 3 of this treatment, tapered on day 7, and discontinued on day 9. They found

that the abdominal girth of their patient was decreased after 24 hours of starting octreotide.⁹

On follow-up of our patient, his abdominal girth was within the normal limit, without any clinical decompensation. This therapy's possible adverse effects include abnormal carbohydrate metabolism, nausea, diarrhea, abdominal distension, abnormal liver function, necrotizing enterocolitis, and pulmonary hypertension.¹⁰ However, our patient did not develop any significant adverse effects. In cases resistant to conservative treatment, surgery is advised, which is usually undertaken for 4–8 weeks.¹⁰ Though, surgical correction of CCA requires expertise and is associated with increased mortality.

CONCLUSION

In this index case, with an early introduction of octreotide, the conservative treatment seems advantageous, as it shortens the time of treatment and hospitalization. Especially in resource poor countries, this could be a more accessible and better alternative.

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