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Case report

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Chylous ascites in a patient with down syndrome: a case report

Abstract

Chylous ascites is the accumulation of chyle in the peritoneal cavity, it is a rare entity in the neonatal age. The causes are related to a delay in the maturation of the lymphatic capillaries at the level of the intestinal microvilli or their poor development, destruction of the lymphatic capillaries secondary to neoplasms, infections or inflammation. We present the case of a patient with a prenatal diagnosis of non-immune fetal hydrops with 46XY + 21 karyotype, diagnosed with chylous ascites on day 40 of life. The diagnostic approach, treatment and evolution in this rare entity are presented.

Keywords: chylous ascites, down syndrome, non-immune fetal hydrops, neonatal ascites

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Jorge Chang Edgar Alberto,[1] Villanueva Acosta Cecilia,[1] Morales Barquet Deneb Algedi,[2] Machuca Vaca Alfredo José,[3] Guerrero Kanan Ricardo Guillermo,[2] Torres García Laura Isela[1]

[1]Neonatologist resident, National Institute of Perinatology, Mexico
[2]Neonatologist, National Institute of Perinatology, Mexico
[3]Neonatal surgeon, National Institute of Perinatology, Mexico

Correspondence: Edgar Jorge Chang, Pediatric neonatologist, assigned to the toco-surgery unit, National Institute of Perinatology "Isidro Espinoza de los Reyes", Mexico City, Mexico, Tel +525528648509

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Introduction

Chylous ascites is the accumulation of chyle in the peritoneal cavity, characterized by a milky, odorless liquid with elevated levels of triglycerides. It is a rare condition in the neonatal age, with 112 cases reported worldwide as of March 2022.[1] Different theories regarding its origin have been proposed. In 1967, Craven and colleagues associated it with the obstruction of the chylous cistern (Pequet's cistern), which receives chyle rich in fats from the intestine and causes reflux into the peritoneal space. In 1975, Weber and collaborators attributed it to abdominal lymphatic dysplasia.[1]

The causes are related to a delay in the maturation of the lymphatic capillaries at the level of the intestinal microvilli or poor development of these capillaries, as well as destruction secondary to neoplasia, infections, or inflammation. Alterations in lymphatic capillary development may be associated with genetic syndromes, such as Turner Syndrome, Down syndrome, and Klippel-Trenaunay-Weber Syndrome.[2]

Background

Patients typically present with abdominal distension from birth or this condition can be detected prenatally by ultrasound. Chylous ascites can result in serious mechanical, nutritional, and immunological complications due to the loss of proteins, immunoglobulins, and lymphocytes. Therefore, it is crucial to monitor respiratory mechanics when managing the condition conservatively, and maintaining an adequate nutritional balance can help decrease lymph production and flow. Treatment includes paracentesis, total parenteral nutrition, enteral feeding based on medium-chain triglycerides, somatostatin analogues, and in refractory cases, surgical treatment.[1,2]

Clinical case

We present the case of a patient diagnosed with non-immune fetal hydrops (polyhydramnios, ascites, bilateral hydrocele) at week 27.3. A hydrops approach was conducted, ruling out infectious causes using a TORCH profile of amniotic fluid; Trisomy 21 was diagnosed due to Robertsonian translocation (karyotype 46,XY, DER(13;21)(Q10;Q10),+21). A cardiological assessment revealed a structurally healthy heart with preserved biventricular function.

The baby was born at 35.5 weeks of gestation due to preterm labor, with APGAR scores of 5 and 9 at the first and fifth minute, respectively. Advanced airway management was required due to irregular respiratory effort attributed to abdominal distension. Paracentesis was performed, draining 30 ml of cloudy fluid (turbid cytochemical/ xanthochromic, glucose 52, protein 28, leukocytes 1552, mononuclear 7, polymorphonuclear 93, DHL 146). (Figure 1).

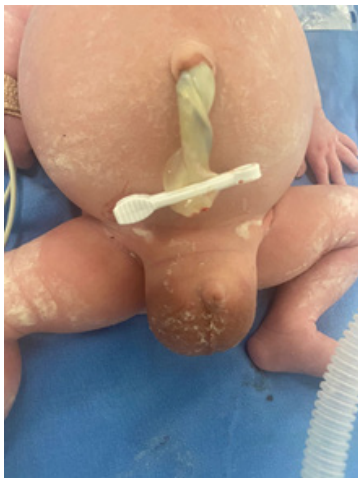

Figure 1 Abdominal distension and bilateral inguinal hernia.

Extubation occurred 24 hours after birth, and enteral feeding commenced at 48 hours of life with expressed human milk, later transitioning to mixed feeding due to signs of abdominal distension. On day 40 of extrauterine life, abdominal circumference increased by 4 cm, leading to a further assessment with abdominal x-ray and ultrasound, reporting significant ascites. A subsequent paracentesis

yielded 20 ml of milky fluid (Figure 2) (cloudy/milky cytochemical, glucose 102, proteins > 3000, leukocytes 264, mononuclear 98, polymorphonuclear 2, DHL 131, triglycerides > 525), confirming the diagnosis of chylous ascites. Exclusive feeding with a hydrolyzed breast milk substitute was initiated (TCM 55%).

Figure 2 Milky fluid of ascites.

Due to persistent ascites, octreotide was started as an infusion of 1 mcg/kg/hour on day 55 of extrauterine life, gradually increasing to a maximum dose of 10 mcg/kg/hour. An intestinal and colon transit study was performed by enema to rule out intestinal malrotation.

At 59 days of extrauterine life, a special formula was changed (TCM 90%) but poorly tolerated due to frequent diarrheal stools, prompting a switch to an elemental diet with a concentration of 15% until achieving tolerance of 20%, with gradual increases in medium-chain triglycerides.

We performed an immunoglobulin determination, which reported IgG 348 mg/dL (610 - 1500) and IgM 28 mg/dL (6 - 30), which confirms the decrease in serum IgG associated with lymphatic leakage.

Fluid was obtained (cytochemical analysis: cloudy/milky, glucose 99, protein 4770, leukocytes 4080, mononuclear 95%, polymorphonuclear 5%, triglycerides 1049). We monitored triglyceride levels in the ascites fluid after starting elemental feeding, observing a significant decrease, with a report of 28 mg/dL after 4 weeks of treatment, in addition to an 8 cm reduction in abdominal perimeter.

Lymphoscintigraphy with Tc-99m rhenium Nano colloid showed areas of abnormal radiotracer concentration in the lymphatic channels of the external iliac chains, compatible with lymphatic ectasia. These findings do not rule out a coexisting leak at this level.

The patient was discharged at 157 days of life with adequate tolerance to the enteral route through special elemental feeding.

We report a complication of healthcare-associated sepsis, with a positive culture for *Klebsiella pneumoniae*, requiring antibiotic treatment with vancomycin and amikacin.

Discussion

Congenital chylous ascites is primarily related to inadequate drainage due to malformation in the lymphatic pathway in 45-60% of cases. However, up to 50% of newborns present no identifiable cause of chylous ascites, with lymphatic leakage due to lack of maturation being suspected.

The diagnosis should be suspected from the first ascitic fluid obtained, which may be collected prenatally or postnatally. Key cytochemical data include increased cellularity > 1000 cells/mL with lymphocyte predominance, protein levels > 2.5 g/dL, lactate dehydrogenase > 110 IU/L, and triglycerides > 200 mg/mL. In our patient's case, there were suggestive data from the paracentesis performed at birth, but we confirmed the diagnosis 55 days later when we obtained distinctly milky fluid.[3–6]

There is no established treatment for congenital chylous effusion, therefore, Tamaoka et al, recommend a combination of strategies (respiratory care, nutritional management, pharmacological interevention),[7] also the drainage is required to improve the respiratory work in case that the ventilation is severely affected, the same interventions that we realized with our patient.

In 20-25% of cases, external compression causing lymphatic obstruction has been observed, for example in cases of malrotation, incarcerated hernia, or malignancy. For this reason, despite conservative treatment and persistence of ascites, we decided to perform an intestinal transit study and colon enema to rule out anatomical causes. Long et al.,[5] reported a series of 10 chylous ascites cases, where they diagnosed intestinal malrotation solely by barium colonic drainage, identifying the ileo-colic junction in the upper right quadrant in 7 out of 10 patients. Nine out of 10 patients required the Ladd procedure.

As described by Lopez,[3] our patient was initially treated conservatively with a hydrolyzed diet with a predominance of medium-chain triglycerides (55% MCT). However, due to poor progress, we escalated treatment to the pharmacological method, with a maximum dose of octreotide at 10 mcg/kg/hour. Due to inadequate response, we changed to an elemental diet with added MCT, which resulted in clinical improvement.

Lymphangiography in neonates is a useful tool for visualizing normal and abnormal lymphatic anatomy, specially for the planning of interventional radiology or surgical treatments.[8] In our case, lymphangiography was performed to determine the etiology and persistence of the condition, and the conclusive report indicated lymphatic stasis at the iliac level. We decided to continue with conservative treatment, as the literature suggests that surgical treatment is not indicated in the absence of severe complications.

We also report infectious complications associated with healthcare, similar to those described by Sooklin.[2] This is linked to a decrease in immunoglobulins, which is attributed to the loss of proteins via chylomicrons

There are new pharmacological therapies to treat chylous effusion in adults, such as midodrine, it may be useful in congenital chylous in neonates, but further studies are needed, although Tamaoka reported its use in treating a patient.[7]

Conclusions

Since congenital chylous ascites is a rare condition in the neonatal stage and can be confused with other causes of ascites, it is important to consider the chemical data suggestive of this entity, even in the absence of the classic milky appearance or elevated triglyceride levels in the ascitic fluid. Early treatment with diets rich in medium-chain triglycerides is a key factor for the patient's prognosis, as it reduces the formation of chylomicrons.

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Conflicts of interest

The authors declare no conflicts of interest.

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