□ Case report □

A case of post-operative chylous ascites after a splenorenal shunt operation in a child with congenital hepatic fibrosis

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Chylous ascites is a rare condition caused by various diseases and conditions that interfere with the abdominal or retroperitoneal lymphatics, and uncommonly it can manifest as a post-operative complication after abdominal, retroperitoneal or mediastinal surgery. Chylous ascites can be diagnosed by a high triglyceride content in ascites. The authors experienced a 5-year-old girl with congenital hepatic fibrosis who presented with chylous ascites after a splenorenal shunt operation, who was successfully managed by fasting and total parenteral nutrition, followed by a lipid-free diet with medium chain triglyceride supplementation. Here, the authors report this case of post-operative chylous ascites after a splenorenal shunt (Warren shunt) operation with a review of the pertinent literature. **(Korean J Pediatr 2006;49:1106-1110)**

Key Words : Chylous ascites, Post-operative, Splenorenal shunt

Introduction

Chylous ascites involves the accumulation of chyle in the peritoneal cavity. It is a rare condition mostly caused by diseases or conditions that interfere with the abdominal or retroperitoneal lymphatic glands[1, 2]. Many factors are known as cause of chylous ascites, such as abdominal surgery, malignancy, blunt abdominal trauma, spontaneous bacterial peritonitis, cirrhosis, pelvic irradiation, peritoneal dialysis, abdominal tuberculosis, and congenital defects in lacrimal malformation[3].

Post-operative chylous ascites is a rare complication of retroperitoneal and mediastinal surgery that is caused by the unrecognized interruption of the major retroperitoneal lymphatic channels and lymphoperitoneal fistula formation[4]. Postoperative chylous ascites always represents a difficult patient treatment problem. Moreover, due to the rarity of this condition, its diagnosis and treatment are not well established[4]. And this rarity is more significant in pediatric patients[5].

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The common characteristics of chylous ascites are as follows; its appearance is usually described as milky or chylous; it is odorless; it has a high triglyceride content that is 2 to 8 fold that of plasma (range : 400-4,000 mg/dL); its specific gravity is greater than that of serum and its protein content greater than 3,000 mg/dL[3, 4, 6]. Its electrolyte level is usually similar to that of serum. A microscopic examination of fluid stained with Sudan Ⅲ shows fat globules and leukocytes with a lymphocytic predominance[3, 4, 6, 7]. Microbiologic studies are usually negative[3].

It has been reported that chylous ascites in children develops in many kinds of pathological conditions, such as congenital lymphatic disorders, malignancy, mesenteric lymphadenitis, liver cirrhosis, abdominal tuberculosis, and certain parasitic infections, and after abdominal surgery, trauma, radiation therapy in the pelvic space ,and peritoneal dialysis[1, 8]. However, most of cases chylous ascites in infants or children are still idiopathic[8].

Many cases of post-operative chylous ascites have been reported in adults, but few reports are available of post-operative chylous ascites in children, including cases of post-operative chylous ascites after an urologic operation[4, 9]. Moreover only one pediatric case report of postoperative chylous ascites after abdominal or retroperitoneal operation has been issued in Korea[10], despite many reports of pos-

toperative chylous ascites in adults.

We report a case of post-operative chylous ascites after with congenital hepatic fibrosis a splenorenal shunt (Warren shunt) operation in a child. This is the first pediatric case of postoperative chylous ascites after a Warren shunt operation reported in Korea.

Case Report

A 5-year-old girl was transferred from the department of pediatric surgery to the department of pediatrics because of poorly controlled post-operative ascites.

When she was 11 months old, hepatosplenomegaly, ascites, and pancytopenia were detected at a local hospital. After transfer to our hospital, she was diagnosed as having congenital hepatic fibrosis accompanied by autosomal recessive polycystic kidney disease by percutaneous liver biopsy and abdominal ultrasonography. Two years and eight months prior to the admission, esophagogastroscopy revealed esophageal varix grade Ⅲ. Two months later, bleeding from esophageal varices developed and the first endoscopic esophageal varix ligation (EVL) was performed. She underwent EVL four times again thereafter. When she was admitted for the last EVL, severe post-EVL variceal bleeding developed, which was treated for 6 days with intravenous somatostasin. At that time, it was also noted that the spleen was palpable below the umbilicus and pancytopenia due to hypersplenism. After consultations with pediatric surgeons, it was decided that she should undergo a porto-systemic shunt operation, because of sustained moderate pancytopenia (especially thrombocytopenia). Five days prior to operation, her common blood cell (CBC) count were white blood cell (WBC) count 2,470/mm^3, hemoglobin 9.9 g/dL, hematocrit 30.6%, platelets 51,000/mm^3. Her platelet count had fallen to 37,000/mm^3.

Sixteen days prior to transfer, she was admitted to department of pediatric surgery to undergo a Warren shunt operation, and 5 days after surgery a large amount of ascites developed. Despite the administration of diuretics, ascites was not controlled, and therefore, she was transferred to the department of pediatrics for the management of refractory ascites.

Medical history taking showed that she had been born as a term baby at gestation age 38 weeks by spontaneous vaginal delivery without any perinatal problems, and that she has been using a corticosteroid nebulizer regularly because of bronchial asthma, which was diagnosed 3 years before admission. There was no family history of hepatic or renal disease.

On the day of transfer (post-operative day 8), she looked mildly dyspneic. Her body weight was 19.9 kg (50-75 percentile), height 107.5 cm (10-25 percentile), and abdominal circumference 70.5 cm, and her blood pressure was 115/51 mmHg, pulse rate 112/min, respiration rate 32/min, and body temperature 37.5℃. Conjunctivae were not anemic and sclerae were not icteric. The oral cavity was clear and the tongue was not dehydrated. Pharyngeal injection and palatine tonsillar hypertrophy were not observed. The cervical lymph node was not palpable, and the chest wall expanded symmetrically without retraction, though and wheezing was heard in both lung fields. Her heartbeat was regular without murmur. Her abdomen was tense and distended. Bowel sounds were normoactive, and tenderness or rebound tenderness was absent. There was shifting dullness at her abdomen. Liver and spleen could not be palpated due to the ascites. Pitting edema, clubbing and cyanosis were not observed. Costovertebral angle tenderness was not observed. Her mental status was alert. No abnormal finding was apparent by neurological examination.

On the day of transfer, her laboratory findings were as follows; WBC count 11,100/mm^3, hemoglobin 12.2 g/dL, hematocrit 36.6%, platelets 156,000/mm^3, aspartate aminotransferase 28 IU/L, alanine aminotransferase 10 IU/L, total bilirubin 2.3 mg/dL, direct bilirubin 0.9 mg/dL, total protein 6.0 g/dL, albumin 3.2 g/dL, blood urea nitrogen 12 mg/dL, creatinine 0.4 mg/dL, glucose 63 mg/dL, cholesterol 80 mg/dL, and C-reactive protein 1.05 mg/dL. According to pre-operative coagulation function testing, her prothrombin time international normalized ratio (PT INR) was 1.7, and activated partial thromboplastin time (aPTT) was 48.7 sec. Routine urinalysis results were normal.

Seven days after transfer (post-operative day 15), ascites tapping was done. The ascites was grossly turbid and whitish-pink (Fig. 1). Red blood cell and WBC counts of ascites were 14,800/mm^3 and 117/mm^3, respectively. An analysis of the ascites revealed 1,283 mg/dL of protein, 123 mg/dL of glucose, 71 IU/L of lactate dehydrogenase, 11 IU/L of amylase, and 646 mg/dL of triglyceride. On the day of ascites tapping, serum triglyceride level was 51 mg/dL.

Abdomen Doppler sonography performed on post-operative day 5 revealed massive ascites and hepatomegaly.

Computed tomography (CT) angiography performed on post-operative day 15 showed a patent splenorenal shunt and massive ascites (Fig. 2A).

From the day of transfer, the dose of diuretics was increased to the maximum dosage to reduce the ascites, but this resulted in no improvement. Under a diagnosis of chylous ascites based on tapping results, a lipid-free diet and medium-chained triglyceride (MCT) supplementation was started on post-operative day 16. However, she failed to respond to diet therapy, and her abdominal circumference increased despite a fat-free diet. At post-operative day 22, we decided to stop the fat-free diet with MCT supplementation, and to start nill per os (NPO) with total parenteral nutrition (TPN). Three days after the initiation of TPN, abdominal circumference began to decrease. Ten days later after the initiation of TPN, the administration of oral diuretics was stopped, and 18 days after the initiation of TPN, a semi-blended fat-free diet was applied with reduced TPN, which resulted in no further abdominal circumference gain. One month later, her abdominal circumference had reduced to 57.5 cm (Fig. 2B), and normal fat-free diet with MCT oil supplementation was applied with TPN discontinuance. Moreover, because her abdominal circumference did not increase again, she was discharged from hospital at post-operative day 58. She continued fat-free diet with MCT supplementation after discharge, and stopped at 1 year after operation due to no more progression of chylous ascites. Two years postoperatively, she is being followed in the outpatient department, and she remains well without chylous ascites.

Fig. 1. Gross finding of ascites with whitish-pink color, suggesting chylous ascites.

Discussion

Chylous ascites is an uncommon condition that is usually caused by diseases that interfere with the abdominal or retroperitoneal lymphatic glands[1, 2]. Postoperative chylous ascites is a rare complication of retroperitoneal and mediastinal surgery that is caused by the unrecognized interruption of the major retroperitoneal lymphatic channels and lymphoperitoneal fistula formation[4]. Postoperative chylous ascites usually develops as a result of operative trauma to the thoracic duct, cisterna chyli or their major tributes in combination with increased chyle production and obstruction of lymphatic drainage from the abdomen[3, 4, 6].

Although various vascular surgical procedures, including aortofemoral bypass, resection and replacement of the inferior vena cava as part of surgery to remove malignant

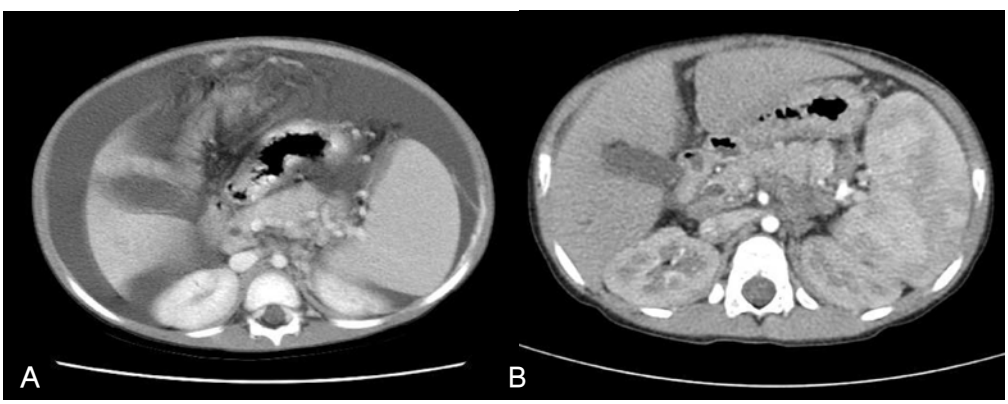

Fig. 2. Abdominal CT findings before (A) and after treatment (B). Fig 2A shows massive ascites without other intra-abdominal abnormality. (B) shows a reduced amount of ascites and a patent splenorenal shunt.

tumors, a Warren shunt and mesocaval shunt procedures may cause chylous complications[3, 4, 6], abdominal aortic surgery is the most common cause of postoperative chylous ascites, causing 81% of all chylous complications due to surgical injury to the retroperitoneal lymphatic glands or cisterna chyli[4].

Paracentesis and fluid examination are essential for the diagnosis of chylous ascites[3, 4]. The common characteristic and diagnostic criterion of chylous ascites is a milky ascitic fluid with a high triglyceride content of usually more than 200 mg/dL or 2 to 8 fold higher than that of plasma[4]. Moreover it has not been reported that the criteria of chylous ascites in children differs from that in adults. In our case, serum triglyceride level was 51 mg/dL and the triglyceride level in ascites was 646 mg/dL (more than 10 folds higher), which is consistent with the diagnostic criteria for chylous ascites.

Chylous ascites in pediatric patients is a extremely rare condition[5], and a few cases of chylous ascites caused as a primary or a congenital diseases in children have been reported in Korea[8, 11-14], but there was only one case report of post-operative chylous ascites after Kasai operation in children[10], although a number of reports have been issued on post-operative chylous ascites in adults[15]. Therefore, this is the second case report of post-operative chylous ascites in Korean children.

The mainstay of nonoperative treatment for nonmalignant chylous ascites in various etiologies is a low fat diet with MCT supplementation, but if it is no use, total parenteral nutrition can be used[4]. Surgical treatment, including direct repair of leaking lymphatic glands and peritovenous shunt placement, should be reserved for patients that fail to respond to conservative management with total parenteral nutrition[16]. The management of post-operative chylous ascites is similar to that of chylous ascites with other causes. Because of diet control difficulties, the preferred nonoperative therapy for chylous ascites in pediatric patients is total parenteral nutrition[4]. Its feasibility for managing chylous ascites in infants and children is well established, despite non-availability of published experiences[5, 16, 17]. In the present case, we initially started a lipid-free diet with MCT supplementation, but abdominal circumference did not reduced on this diet therapy. Thus, a lipid-free diet was found to be ineffective at controlling post-operative chylous ascites. However, NPO supported by TPN was found to be highly effective at improving post-operative chylous ascites in the present case. Moreover, it appears that a lipid-free diet with MCT supplementation was effective at preventing the production of chylous ascites after TPN therapy. Recently, it has been reported that somatostatin (or octreotide) is effective at managing chylous ascites, but experience to date in pediatric patients is very limited[4, 10, 18, 19], and was used when conservative management failed. This therapy was not considered in this patient, because she was responsive to TPN and subsequent fat-free diet with MCT supplementation.

The general outcome of chylous ascites depends mainly on the nature of the underlying pathologic condition causing the lymphatic leakage[4]. Moreover, despite an increased risk of associated complications, post-operative chylous ascites carries a more favorable prognosis with significantly lower mortality rate[4].

Some case reports have described chylous ascites after a Warren shunt operation in adults[20-23]. Maywood et al.[23] suggested that post-operative chylous ascites subsequent to a Warren shunt procedure might not be a rare phenomenon, and Aalami et al.[24] reported that Warren shunts are a particularly common cause of postoperative chylous ascites, because of extensive dissection along the course of the intestinal lymphatics. The prognoses of these patients were varied and depended on their underlying diseases. However, there was no case report of chylous ascites after Warren shunt operation in pediatric patient, and no report has addressed the prognosis of chylous ascites after Warren shunt in pediatric patients.

The authors experienced a case of post-operative chylous ascites after Warren shunt operation, which improved after TPN and the installation of a subsequent lipid-free diet with MCT supplementation. Chylous ascites is a very uncommon condition in pediatric patients, especially after surgery. However, if a patient who has undergone abdominal or mediastinal surgery shows refractory ascites unresponsive to medical treatment, including diuretics, this rare complication should be considered.

한 글 요 약

선천성 간섬유화증에서 비-신장 문합수술 후에 발생한 소아의 유미성 복수증 1례

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유미성 복수는 복강이나 후복강의 림프순환계에 이상이 발생하는 질환이나 각종 원인에 의해서 발생하는 드문 질환으로서, 대개 혈장 내 triglyceride보다 2-8배 이상의 높은 복수 내 triglyceride의 함량을 보이는 경우 진단이 가능하다. 유미성 복수는 여러 가지 원인에 의해서 발생할 수 있지만, 수술 후 부작용으로서 복강이나 후복강 및 종격동의 수술 후 발생하는 경우가 드물게 있다. 저자들은 선천성 간섬유화증을 가진 소아 환자에서 비-신장 단락수술(Warren 단락술) 후에 발생하여 금식 및 총정맥영양에 이어 저지방식이를 시행한 후 호전된 유미성 복수증 1례를 경험하였기에 문헌고찰과 함께 이를 보고하는 바이다.

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