

TREATMENT OF CONGENITAL CHYLOUS ASCITES WITH SOMATOSTATIN IN A PREMATURE NEWBORN WITH DOWN SYNDROME

Case Report

DOWN SENDROMLU BİR PREMATÜRE YENİDOĞANDA KONJENİTAL ŞİLÖZ ASSİTİN SOMATOSTATİNLE TEDAVİSİ

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ABSTRACT

Chylous ascites commonly occurs secondary to surgical trauma, and primary congenital chylous ascites is very rare. Conventional therapy consists of cessation of oral intake, total parenteral nutrition, medium chain triglyceride containing diet and paracentesis. Here we describe a premature newborn with Down syndrome with primary chylous ascites. The baby was finally treated with somatostatin.

Key words: *Congenital chylous ascites; Down syndrome; somatostatin; prematurity.*

ÖZET

Konjenital şilöz assit çok nadir görülen bir durumdur. Genellikle cerrahiye ikincil olarak görülmekle birlikte primer vakalar çok daha nadir ve sporadiktir. Geleneksel tedavide parasentez, oral alımın kesilip total parenteral beslenme şeklinde beslenme ve MCT içeren diyet yer alır. Burada prematüre olarak doğan ve Down sendromu bulguları olan bebekte daha sonra spontan olarak gelişen ve somatostatin ile tedavi edilen şilöz assit olgusu sunulacaktır.

Anahtar sözcükler: *Konjenital şilöz assit; Down sendromu; somatostatin; premature*

INTRODUCTION

Chylous ascites (CA) is a rare form of ascites formation caused by the accumulation of chyle with a triglyceride content >200 mg/dl in the abdominal cavity (1). It may be due to the obstruction of the lymph flow, exudation of chyle or traumatic thoracic duct obstruction causing direct leakage (2). Trauma is the most common cause in children, mainly due to peritoneal or intraabdominal surgery. Therapeutic strategies consist of cessation of oral intake, administering total parenteral nutrition (TPN), median chain triglyceride (MCT) rich diet or TPN and drainage of the

chylous material. In case of treatment failure, somatostatin and its analogue octreotide may be administered to treat chylothorax and chylous ascites in both adults and children (3,4).

Here we report a premature newborn with Down syndrome with a prenatal diagnosis of ascites, which was revealed to be CA.

CASE REPORT

A 2.5 kg female infant was delivered by cesarean section at 34 weeks of gestation because of suspected hydrops fetalis. Prenatal ultrasonography revealed congenital ascites. The Apgar scores were 6, 7, and 8 at 1, 5 and 10 minutes, respectively. Dysmorphic features compatible with Down syndrome were present on physical examination. The abdomen was massively distended and dull to percussion (**Figure 1**).



Vital functions were stable except for tachypnea. Complete blood count revealed normal leucocyte, hemoglobin and platelet levels. Serum electrolytes, liver and renal function tests and blood coagulation tests were within normal ranges. Ultrasonography of the abdomen showed massive ascites with normal liver and spleen. No cardiac abnormalities were revealed by echocardiography.

The diagnosis of hydrops fetalis was eliminated by these tests. Percutaneous abdominal paracentesis performed at

15th hours after delivery revealed non-purulent, clear-colored fluid, resembled to transudatic ascites. Chromosome analysis revealed the karyotype as 47,XX,+21. The patient was exclusively fed with human milk and formula. As the abdominal girth consistently increased, a repeat paracentesis was performed on 5th day. The fluid was milky this time, and laboratory analysis revealed the contents of triglyceride as 1162mg/dl and white blood cells (WBC) as 13480/mm³ with 60% lymphocytes. The first paracentesis revealed transuda and after the baby was feeding enterally, the ascitic fluid turned to chylous material.. So the enteral feeding was stopped and TPN was administered. Since abdominal girth did not resolve at 7th day, somatostatin (SS) infusion was initiated at a dose of 3 mcg/kg/h, and was gradually increased to 9 mcg/kg/h, The newborn was monitored for blood glucose and no serious side effect was seen. The abdominal girth regressed on the second day of somatostatin treatment. Somatostatin therapy was stopped gradually 10 days after administration. Unfortunately, the baby died due to lower tract respiratory infection on the 46th day of hospitalization.

DISCUSSION

Ascites in newborn is usually related to other diseases, and congenital or primary chylous ascites is a very rare entity (5). The pathogenesis of primary CA is poorly understood and genetic factors are thought to be involved (6). Chylous ascites frequently presents as progressive and painless abdominal distention. Most often, the diagnosis of chylous ascites is not considered before performing a paracentesis. The triglyceride levels in ascitic fluid are very important in defining chylous ascites. Triglyceride values are typically above 200 mg/dL. If possible, lymphangiography and lymphoscintigraphy, former being the gold standard, can reveal the cause of lymphatic obstruction (7). However none

of those tests could be done for our patient due to technical problems.

There is not yet an established treatment protocol for primary CA. Treatment of congenital CA is primarily conservative, regarding reduced intestinal fat absorption and chylous flow. Medium-chain triglycerides are given because they are transported as free fatty acids and glycerol directly to the liver via the portal vein. The use of a low-fat diet with medium-chain triglyceride supplementation therefore reduces the production and flow of chyle (6).

Somatostatin has been widely used for chylothorax both in pediatric and adult patients. There are few reports on the beneficial effect of Octreotide in primary CA of premature newborns. (7,8). It has been speculated that somatostatin improves chylous ascites by inhibition of lymph fluid excretion through specific receptors found in the lymphatic vessels (6). There is yet no consensus about the dosing, mode of administration and treatment duration with somatostatin. In the presented case we use intravenous route, started with a dose of 3 mcg/kg/h and increased to 9 mcg/kg/h for 10 days adjusted to the abdominal girth. There was no complication such as hypoglycemia, vomiting diarrhea or necrotising enterocolitis due to SS treatment.

The coexistence of congenital chylous ascites and Down syndrome is reported in a female term infant from USA (9). The patient was treated with TPN and paracentesis and fully recovered after 10 weeks of treatment. A thorough research in English literature revealed that this is the first premature case with Down syndrome and primary CA.

In conclusion, congenital chylous ascites is a very rare condition which may be associated with congenital or genetic abnormalities such as Down syndrome. The management consists of cessation of oral intake, paracentesis, TPN, MCT administration, initiation of or somatostatin. We successfully treated a

premature newborn with Down syndrome and primary CA with TPN and somatostatin, although the patient died later due to unrelated causes.

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