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Research Article

The Case Number 112 of Congenital Chylous Ascites and Its Successful Treatment

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Abstract

Background: Congenital chylous ascites is a very rare condition that has been well recognized as early as the 1950s, and was mostly called "Congenital chyloperitoneum" during that time. Few cases have been reported during the 1960s, and in 1967, Craven et al attributed the development of congenital chylous ascites to obstruction of the cisterna chyli and chylous reflux into the peritoneal space and small intestine and they demonstrated lymphangiographically.

Patients and methods: The use of a unique dietary therapy for the treatment of congenital chylous ascites is briefly described and the relevant literature is described.

Results: An infant with congenital chylous ascites was seen at about the age of two and half months. He was initially treated with intravenous somatostatin infusion, and thereafter by repeated paracentesis, and continued to receive breast and bottle milk feedings. However, when he was first seen, ultrasound was still showing moderate ascites. The boy was successfully treated a combination of a unique form of very low-fat diet and octreotide acetate subcutaneous injections. Treatment was not associated with a negative effect on growth or development, and no evidence of nutritional deficiency was observed.

Conclusion: The case number 112 of congenital chylous ascites is reported and its unique therapeutic approach is described.

Keywords: congenital chylous ascites; Iraq

Introduction

Congenital chylous ascites is a very rare condition that has been well recognized as early as the 1950s, and was mostly called "Congenital chyloperitoneum" during that time [1, 2, 3]. Few cases have been reported during the 1960s [4, 5, 6], and in 1967, Craven et al attributed the development of congenital chylous ascites to obstruction of the cisterna chyli and chylous reflux into the peritoneal space and small intestine and they demonstrated lymphangiographically [6].

In 1975, Weber et al attributed the development of congenital chylous ascites to abdominal lymphdysplasia, and reported the 52nd case of this rare disorder [7].

In 1979, Flores and colleagues reported the occurrence of an autosomal recessive cases congenital chylous ascites in a brother and sister. The parents were first cousins [8].

About seventy-six cases of congenital chylous ascites were reported during half century from the fifties to the end of the 1990s [1-24].

Patients and methods

The use of a unique dietary therapy for the treatment of congenital chylous ascites is briefly described and the relevant literature is described.

Results

An infant with congenital chylous ascites was seen at about the age of two and half months. He was initially treated with intravenous somatostatin infusion, and thereafter by repeated paracentesis, and continued to receive

breast and bottle milk feedings. However, when he was first seen, ultrasound was still showing moderate ascites. The boy was treated a combination of very low-fat and octreotide acetate subcutaneous injections.

Low-fat infant formula was not easily or rapidly available, and the dietary treatment included replacing milk with rice milk with early gradual introduction of juices including orange, grape, and melon juices. Nutritional supplements included amino acids and royal jell capsule.

Octreotide acetate 50 microgram was given by subcutaneous injections weekly for one month, and thereafter, every 10 days for month.

Ascites was completely disappeared after one month of treatment, and didn't recur when the child was seen at about the age of nine months.

Treatment was not associated with a negative effect on growth or development, and no evidence of nutritional deficiency was observed.

Discussion

Congenital chylous ascites is a very rare disorder with about seventy-six cases reported in the medical literature during the 1950s, 1960s, 1970s, 1980s and 1990s, including the case reported by Matsuzaki et al in 1987 which was associated with infantile sialidosis [7-24].

In 1975, Weber et al reported a girl with neonatal chylous ascites that had been treated with repeated paracentesis until about the age of three years when treatment with a low fat diet was started, and after 8 weeks, ascites was no longer recurring [7].

In 1978, Portier et al reported the treatment of a difficult case of congenital chylous ascites by peritoneo-venous shunt [9]. Guttman and colleagues (1982) also reported the treatment of two patients with severe chylous ascites associated with generalized lymphatic malformations, and causing difficulty with breathing with peritoneo-venous shunts [13].

Alliët and colleagues (1992) reported the occurrence of congenital chylous ascites in girl with Down syndrome who experienced respiratory distress secondary at birth because of severe abdominal distension. The patient was treated with total parenteral nutrition and for10 weeks [18].

In 1996, Caty et al reported a successful treatment of congenital chylous ascites with a somatostatine analog [20].

By the end of the year 2010, there were about 89 cases of congenital chylous ascites reported in the medical literature [25-33].

Machmouchi et al (2000) reported four patients (3 males and one female) with congenital chylous ascites, three of them responded to low-fat diet and medium-chain triglycerides and multiple paracentesis. The fourth patient was a male who had a macro-digit affecting the right index and medius. He continued to have ascites after 10 weeks, but the ascites completely resolved during the third week of complete parenteral alimentation, and no recurrence occurred during one-year of follow-up [25].

Mitsunaga et al (2001) treated two patients with congenital chylous ascites initially conservatively with medium-chain triglyceride milk and total parenteral nutrition, but more than one month treatment was not effective. Thereafter, they successfully treated the patients by lymphatic duct ligation after the identification of the area of chylous leakage using a laparoscopic lymphoid dye test [26].

Lee (2002) reported a patient with congenital chylous ascites who was fully recovered after 26 days of total parenteral nutrition [27].

te Pas et al (2004) reported a boy with congenital chylous ascites who was initially treated with total parental feeding and later by diet with medium-chain triglycerides, and octeotride, a somatostatin analogue. The patient didn't respond to theses therapies and required surgical ligation of the leaking lymphatic duct [28].

Huang and Xu (2005) the treatment of congenital chylous ascites with total parenteral nutrition and somatostatin [29].

Siahanidou et al (2009) reported the occurrence of gall stones in a patient with congenital chylous ascites.

Karagol et al (2010) emphasized the rarity of congenital chylous ascites and reported a patient neonate.

During March, 2022, there were about 111 cases of congenital chylous ascites reported in the medical literature [34-49].

In 2011, Huang et al reported a premature neonate with congenital chylous ascites who was treated successfully with intravenous infusion of octreotide, a synthetic somatostatin analogue after failure to response to traditional supportive therapies [34].

Spagnol et al (2011) reported two patients with refractory congenital chylous ascites treated surgically successfully by cauterization, and fibrin glue application [35].

Mouravas et al (2012) reported the successful treatment of 4 patients having congenital chylous ascites including one patient treated conservatively and three required surgery. Two patients were treated with excision of intra-abdominal cysts and one patient was treated with ligation of the left lumbar lymphatic trunk and cisterna chyli and the use of fibrin glue [36].

Olivieri et al (2012) reported the treatment of a patient with congenital chylous ascites with a failed conservative management, but thereafter was treated successfully with fasting, total parenteral nutrition and octreotide for a period of 15 days followed by medium chain triglyceride feeds. On day 25, breast feeding started, and was discharged on day 33 [37].

Bhattacharya and colleagues (2012) reported the second case of congenital chylous ascites occurring in association with Down's syndrome, and was also associated with lymphedema [38].

Moreira Dde et al (2013) reported a patient with recurrent chylous ascites who didn't respond to medical treatment. Exploratory laparotomy couldn't identify lymph leakage site that could be ligated. The patient was treated successfully by the placement of a hemostatic oxidized cellulose mesh on lymph exudation areas, with a thin layer of fibrinogen/thrombin glue over the mesh [39].

Purkait et al (2014**)** reported a 2 and half month infant with congenital chylous ascites who didn't respond to treatment with medium-chain triglycerides formula and paracentesis, but was successfully treated with octreotide intravenous infusion [40].

Sadovnikova and colleagues (2015) emphasized the variable pathogenesis of congenital chylous ascites and reported a patient with congenital chylous ascites caused by peritoneal cavity lymphangioma [41].

Cao et al (2016) reported two patients congenital chylous ascites treated successfully with rice soup and parenteral nutrition [42].

Ermarth (2016) reported the association of congenital chylous ascites caused by primary lymphatic defect with additional vascular anomaly, with Type VI Ehlers-Danlos Syndrome. The patient was treated with octreotide and total parenteral nutritional [43].

Long et al (2018) reported a patient with congenital chylous ascites caused mesenteric lymphatic obstruction associated with intestinal malrotation [44].

Xu et al (2018) reported the association of congenital chylous ascites phacomatosis pigmentovascularis which is associated with pigmentary nevus and capillary malformation in a five-month old [45].

Carr et al (2019) reported the successful treatment of four patients who had refractory chylous ascites with complete retroperitoneal exposure with application of fibrin glue and Vicryl mesh [46].

Saxena et al (2020) a patient with congenital chylous ascites refractory to initial treatment with medium-chain triglyceride based diet followed treatment with total parenteral nutrition plus octreotide. Thereafter, the patient was treated successfully surgically using fibrin glue the area of mesenteric lymphatic leakage [47].

Sooklin et al (2020) a patient with severe congenital chylous ascites in a preterm neonate who experienced severe respiratory distress at birth and required immediate paracentesis. Treatment with medium-chain triglyceride was not satisfactory, but octreotide treatment was successful [48].

Tamaoka et al (2021) reported the third case of congenital chylous ascites occurring in association with Down syndrome. The case was refractory to traditional therapies, but was successfully treated with midodrine which is an alpha-1 adrenoreceptor agonist [49].

Successful dietary treatment of congenital chylous ascites with a low fat diet was probably first reported in 1975 by Weber et al. A variety of surgical treatments have been reported including peritoneo-venous shunts (Portier et al, 1978) and lymphatic duct ligation (Pearl and colleague, 1977; Mitsunaga et al, 2001; Densupsoontorn et al, 2009). Successful treatment with rather lengthy period has also been reported total parenteral nutrition (Alliët and colleagues, 1992; Machmouchi et al, 2000; Lee, 2002).

Successful treatment of congenital chylous ascites with a somatostatin analog has also been reported (Caty et al, 1996; Purkait et al, 2014; Sooklin et al, 2020), but was not successful all the time (te Pas et al, 2004; Olivieri, et al, 2012; Saxena et al. 2020).

Treatment with low-fat diet and medium-chain triglycerides may not always be successful (Machmouchi et al, 2000; Saxena et al, 2020; Sooklin et al, 2020).

Treatment with total parenteral nutrition with or without medium-chain triglyceride milk also may not always be successful (Mitsunaga et al, 2001; te Pas et al, 2004; Olivieri, et al, 2012; Purkait et al ,2014; Saxena et al, 2020).

Conclusion

The case number 112 of congenital chylous ascites is reported and its unique therapeutic approach is described.

Conflict of interest: None.

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