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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; Treatment; Nutritiona

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 [1-3]. Few cases have been reported during the 1960s [4-6], 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].

Alliet 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 macrodigit 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 [9] and lymphatic duct ligation [11,28,32]. Successful treatment with rather lengthy period has also been reported total parenteral nutrition [20,27,29].

Successful treatment of congenital chylous ascites with a somatostatin analog has also been reported [22,42,50], but was not successful all the time [30,39,49].

Treatment with low-fat diet and medium-chain triglycerides may not always be successful [27,49,50].

Treatment with total parenteral nutrition with or without medium-chain triglyceride milk also may not always be successful [28,30,39,42,49].

Conclusion

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

Conflict of interest

None.

Bibliography

1. Hurez A and Leveque C. "Le chylopéritoine congénital [Congenital chyloperitoneum". *Gazette Médicale de France* 59.11 (1952): 669-672.

2. Janet H., *et al*. "Chyloperitoine congenital [Congenital chyloperitoneum]". *Nourrisson* 41.1 (1953): 1-9.

3. Niznikowska-Marks MJ. "Przypadek wrodzonego wylewu mleczu do otrzewnej [A case of congenital chyloperitoneum]". *Pediatria Polska* 29.11 (1954): 1117-1120.

4. Alvarado A. "Congenital chylous ascites". *Clinic Proceedings Children's Hospital of the District of Columbia* 18 (1962): 109-113.

5. Freundlich E., *et al*. "Congenital chylous ascites and chylothorax. Case report". *The Israel Medical Association Journal* 21 (1962): 171-7.

6. Craven CE., *et al*. "Congenital chylous ascites: lymphangiographic demonstration of obstruction of the cisterna chyli and chylous reflux into the peritoneal space and small intestine". *The Journal of Pediatrics* 70.3 (1967): 340-345.

7. Weber HP., *et al*. "Congenitaler chylöser Ascites. Eigene Beobachtung und Literaturübersicht über 51 Kinder [Congenital chylous ascites. Case reports and review of 51 recorded cases". *Klinische Pädiatrie* 187.4 (1975): 370-376.

8. Flores S., *et al*. "Chylous ascites in sibs from a consanguineous marriage". *American Journal of Medical Genetics* 3.2 (1979): 145-148.

9. Portier F., *et al*. "Ascite chyleuse congénitale irréductible. Traitement par dérivation péritonéo-veineuse [Irreducible congenital chylous ascites. Therapy by peritoneo-venous shunt]". *Nouvelle Presse Medicale* 7.44 (1978): 4048.

10. Evers KG and Huth F. "Congenital lymphatic vessel dysplasia with intestinal lymphangiectasis and chylous ascites in twins". *Monatsschr Kinderheilkd* 125 (1977): 526–529.

11. Pearl J., *et al*. "Chylous ascites: the first reported surgical cure by direct ligation". *Journal of Pediatric Surgery* 12 (1977): 687-691.

12. Volkman RW. "Chylous ascites: the first reported surgical cure by direct ligation". *Journal of Pediatric Surgery* 13 (1978): 351.

13. Guttman FM., *et al*. "Experience with peritoneo-venous shunting for congenital chylous ascites in infants and children". *Journal of Pediatric Surgery* 17.4 (1982): 368-372.

14. Hogeman PH., *et al*. "Chyloperitoneum in a newborn infant". *Tijdschr Kindergeneeskd* 51 (1983): 143-146.

15. Matsuzaki K., *et al*. "A case of infantile sialidosis associated with congenital chylous ascites". *No To Hattatsu* 19.3 (1987): 249-253.

16. Forg A., *et al*. "Medical treatment of chylous ascites: apropos of a case". *Pediatrics* 42 (1987): 253-254.

17. Asada M., *et al*. "A case of congenital chylous ascites]". *Nihon Sanka Fujinka Gakkai Zasshi* 41.2 (1989): 231-233.

18. Pallas Alonso CR., *et al*. "Ascitis quilosa congénita. A propósito de un caso [Congenital chylous ascites. Report of a case]". *Anales Españoles de Pediatría* 33.5 (1990): 487-488.

19. Liao HB., *et al*. "Neonatal chylous ascites: report of two case". *Acta Paediatrica Sinica* 31 (1990): 42-52.

20. Alliët P., *et al*. "Chylous ascites: total parenteral nutrition as primary therapeutic modality". *European Journal of Pediatrics* 151.3 (1992): 213-214.

21. Woods CG., *et al*. "Recessively determined chylous ascites: a case report and possible mouse model". *Clinical Dysmorphology* 1 (1992): 179-181.

22. Caty MG., *et al*. "Successful treatment of congenital chylous ascites with a somatostatine analog". *Pediatric Surgery International* 11 (1996): 396-397.

23. Chye JK., *et al*. "Neonatal chylous ascites: report of three cases and review of the literature". *Pediatric Surgery International* 12 (1997): 296-298.

24. D'Agastino S., *et al*. "Neonatal chylous ascites: a case report". *La Pediatria Medica e Chirurgica* 20 (1998): 285-286.

25. Bappal B., *et al*. "Successful surgical treatment of congenital chylous ascites". *Indian Pediatrics* 35.3 (1998): 276-277.

26. Bagolan P., *et al*. "Fetal tachycardia and chylous ascites". *British Journal of Obstetrics and Gynaecology* 106 (1999): 376-378.

27. Machmouchi M., *et al*. "Congenital chylous ascites: report of four cases and review of the literature". *Annals of Saudi Medicine* 20.5-6 (2000): 436-439.

28. Mitsunaga T., *et al*. "Successful surgical treatment of two cases of congenital chylous ascites". *Journal of Pediatric Surgery* 36.11 (2001): 1717-1719.

29. Lee YY., *et al*. "Total parenteral nutrition as a primary therapeutic modality for congenital chylous ascites: report of one case". *Acta Paediatr Taiwan* 43.4 (2002): 214-216.

30. Te Pas AB., *et al*. "Intractable congenital chylous ascites". *Acta Paediatrica* 93.10 (2004): 1403-1405.

31. Huang Y and Xu H. "[Treatment of congenital chylous ascites using total parenteral nutrition and somatostatin: a case report]". *Zhonghua Er Ke Za Zhi* 43.2 (2005): 152-153.

32. Densupsoontorn N., *et al*. "Congenital chylous ascites: the roles of fibrin glue and CD31". *Acta Paediatrica* 98.11 (2009): 1847-1849.

33. Siahanidou T., *et al*. "Cholelithiasis in a neonate with chylous ascites". *Journal of Perinatology* 30.3 (2010): 227-230.

34. Karagol BS., *et al*. "Therapeutic management of neonatal chylous ascites: report of a case and review of the literature". *Acta Paediatrica* 99.9 (2010): 1307-1310.

35. Melo-Filho AA., *et al*. "Refractory congenital chylous ascites". *Indian Journal of Pediatrics* 77.11 (2010): 1335-1337.

36. Huang Y., *et al*. "Successful management of congenital chylous ascites in a premature infant using somatostatin analogue". *Indian Journal of Pediatrics* 78.3 (2011): 345-347.

37. Spagnol L., *et al*. "Preoperative administration of Sudan III and successful treatment of persistent chylous ascites in a neonate". *Journal of Pediatric Surgery* 46.5 (2011): 994-997.

38. Mouravas V., *et al*. "Diagnosis and management of congenital neonatal chylous ascites". *Hippokratia* 16.2 (2012): 175-180.

39. Olivieri C., *et al*. "Successful management of congenital chylous ascites with early octreotide and total parenteral nutrition in a newborn". *BMJ Case Report* 2012 (2021): bcr2012006196.

40. Bhattacharya S., *et al*. "Congenital chylous ascites and lymphedema in Down's syndrome". *Indian Journal of Pediatrics* 79.11 (2012): 1532.

41. Moreira Dde A., *et al*. "Congenital chylous ascites: a report of a case treated with hemostatic cellulose and fibrin glue". *Journal of Pediatric Surgery* 48.2 (2013): e17-19.

42. Purkait R., *et al*. "Congenital chylous ascites treated successfully with MCT-Based formula and octreotide". *Journal of Indian Association of Pediatric Surgeons* 19.3 (2014): 175-177.

43. Sadovnikova IV., *et al*. "Congenital chylous ascites: A case report]". *Eksp Klin Gastroenterol* 1 (2015): 84-87.

44. Cao Y., *et al*. "Parenteral nutrition combined with rice soup can be a safe and effective intervention for congenital chylous ascites". *Asia Pacific Journal of Clinical Nutrition* 25.3 (2016): 631-635.

45. Ermarth AK., *et al*. "Congenital Chylous Ascites and Ehlers-Danlos Syndrome Type VI". *ACG Case Reports Journal* 3.4 (2016): e186.

46. Long L., *et al*. "Congenital chylous ascites in infants: another presentation of intestinal malrotation". *Journal of Pediatric Surgery* 53.3 (2018): 537-539.

47. Xu S., *et al*. "A female infant with phacomatosis pigmentovascularis and congenital chylous ascites: A case report". *Medicine (Baltimore)* 97.34 (2018): e12012.

48. Carr BD., *et al*. "Retroperitoneal exploration with Vicryl mesh and fibrin tissue sealant for refractory chylous ascites". *Journal of Pediatric Surgery* 54.3 (2019): 604-607.

49. Saxena R., *et al*. "Refractory congenital chylous ascites: role of fibrin glue in its management". *Journal of Indian Association of Pediatric Surgeons* 25.4 (2020): 245-247.

50. Sooklin L., *et al*. "Management of large congenital chylous ascites in a preterm infant: fetal and neonatal interventions". *BMJ Case Reports* 13.9 (2020): e235849.

51. Tamaoka S., *et al*. "Midodrine, an oral alpha-1 adrenoreceptor agonist, successfully treated refractory congenital chylous pleural effusion and ascites in a neonate". *Chest* 159.4 (2021): e189-e191.