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Successful treatment for intractable chylous ascites in a child using a peritoneovenous shunt

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Abstract Intractable post-operative chylous ascites had been managed successfully using a peritoneovenous shunt (PVshunt). A 4-year-old girl with neuroblastoma originated from the right adrenal gland was admitted to our hospital. Following the preoperative chemotherapy, tumor resection, and lymph node dissection of the abdominal paraaortic region were carried out. Post-operative radiation therapy 9.6 gray to the tumor bed and to the paraaortic region and a high dose chemotherapy supported by auto bone marrow transplantation were completed. Three months later some enlarged lymph nodes along the duodeno-hepatic ligament were detected and these had gradually increased in size. Lymph node dissection along the hepatic artery and the abdominal aorta was carried out. Pathological examination of the specimen showed reactive lymph node swelling. Chylous ascites developed several days after surgery. Despite the medium-chain triglycerides meal or total parental nutrition, the ascites persisted for more than 80 days. Multiple paracenteses were mandatory. A PV shunt was implanted and the ascites was resolved by the fourth post-operative day. Thirty months later, the vascular end tube of the shunt was ligated. As ascites had not accumulated for 2 weeks, the PV shunt was removed. The patient has been doing well without recurrence of ascites or neuroblastoma for 12 years. As PV shunts were mostly used for long lasting disease, it has not been referred as to how to know when the shunt

should be removed. If the shunt is inserted for transient management of ascites, less invasive methods of investigation to know when to remove the shunt need to be developed.

Keywords Chylous · Ascites · Peritoneovenous shunt · Child

Introduction

It has been reported that chylothorax or chyloperitoneum in children developed in many pathological conditions, such as congenital lymphatic disorders, thoracocardiac operation, abdominal surgery, malignancy, trauma, liver cirrhosis, radiation therapy in the pelvic space, peritoneal dialysis, and abdominal tuberculosis [1]. While spontaneous recovery has occurred in most post-operative chylous effusions, repeated paracenteses may cause considerable loss of calories, protein, and lymphocytes. A case with intractable post-operative chylous ascites successfully managed using a peritoneovenous shunt (PV shunt) is reported.

Case report

A 4-year-old girl with neuroblastoma was admitted to our hospital. The radiological evaluation showed a huge calcificated right adrenal tumor invading the liver and paraaortic lymph node swelling. Further evaluations did not show any distant metastasis including bone, bone marrow, lung, or brain. Following the preoperative chemotherapy with cyclophosphamide (CPA), vincristine (VCR), adriamycin (ADR) and *cis*-platinum (CDDP), tumor resection, and lymph node dissection of the abdominal paraaortic region were carried out. Post-operative chemo-radiotherapy was performed. Radiation (9.6 gray) to the tumor bed, paraaortic area and high-dose chemotherapy of CDDP, VP-16, and melphalan supported by auto bone marrow transplanta-

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tion were completed. The patient demonstrated good tolerance, recovered from the bone marrow suppression, and was discharged from the hospital. Total doses of the agents administered were as follows: VCR 3.0 mg/m², CPA 2,500 mg/m², CDDP 460 mg/m², ADR 245 mg/m², VP-16 200 mg/m², melphalan 180 mg/m². Three months later, some enlarged lymph nodes along the duodeno-hepatic ligament were detected by abdominal ultrasound echogram (ECHO) and these gradually increased in size. Lymph node dissection along the hepatic artery and abdominal aorta was carried out again. Pathological examination of the resected specimen showed reactive lymph node swelling without evidence of malignancy. The patient complained abdominal fullness, pain, and nausea several days after operation. Physical examination showed a notable abdominal distension and fluctuation. Abdominal ECHO detected a considerable amount of ascites and abdominal paracentesis was performed. The analysis of evacuated ascites showed the following: specific gravity 1.034, chylomicrone (+), cell count 6,000/mm³, lymphocyte 85%, and monocyte 11%. While the medium-chain triglycerides (MCT) meal or total parental nutrition (TPN) was administered, the ascites persisted for 80 days. Repeated paracenteses were mandatory and a considerable amount of fresh frozen plasma needed to be supplied. As both the cytological examination for malignant cells and the bacterial examination were negative, the evacuated ascites was re-infused through the central venous access. Neither cardiac congestion nor disseminated intravascular coagulation (DIC) nor sepsis developed. A PVshunt (Denver Biomaterials Inc, Evergreen, CO) was implanted. The vascular end tube was placed in the superior vena cava through the right internal jugular vein. The ascites was resolved by the fourth post-operative day. The patient was subsequently discharged on the seventh day after shunt implantation. As ascites had not developed for 20 months after the implantation, radiological studies were performed to evaluate if the shunt was patent and functioning. While the abdominal ECHO did not show accumulation of ascites, the radiopaque material, which was injected into the shunt tube using a fine needle, moved to the central vein in a few minutes. Ten months later, the vascular end tube of the shunt was ligated. As ascites had not accumulated over 2 weeks, the shunt was removed. Neither ascites nor recurrence of neuroblastoma has been observed for 12 years. And the patient has been doing well.

Discussion

Post-operative chylous effusion results from injury of the thoracic duct, cisterna chyli, or its intestinal tributaries. While chylous ascites may develop after operation for lymphangioma, abdominal aortic surgery, or lymph node dissection in the retroperitoneal space, it is likely to subside spontaneously. Blalock [2] reported that for the lymphatic leakage to persist, not only injury of the

thoracic duct but also impairment of the collateral pathway formation must be present. Two cases with chylous ascites after radiation therapy or chemotherapy were reported. In one case, the chylous ascites persisted after the retroperitoneal lymph node dissection and two courses of post-operative dactinomycin therapy [3]. Another case had a combined treatment of radiation and chemotherapy [4]. In the presented case, the radiation and the multi-agent chemotherapy before the second lymph node dissection might impair healing of the injured lymphatics and formation of a collateral circuit.

Although most of the patients with post-operative chylous effusion were managed by paracentesis, MCT milk, low fatty acid meal or TPN successfully, surgical treatment such as ligation of the injured lymphatics or a PV shunt was indicated on some occasions. While many cases of successful ligation have been reported, usually it is difficult to locate the leaking lymphatics during the operation. The literature review of case reports revealed 17 cases of children under 12 years who underwent implantation of a PV shunt (Table 1) [5–17]. Seven shunts in four patients were obstructed before ascites was resolved. All of these children had congenital lymphatic disorders and chylous ascites. High levels of cell counts and protein contents in the chylous ascites increase the risk of shunt obstruction. Denver shunt has a valve mechanism, which prevents back flow of blood and the valve chamber lies in the subcutaneous tissue can be compressed to promote flow and to relieve blockage. Other complications reported were cardiac congestion, DIC, sepsis, nephritis, dissemination of malignant cells, and perforation of the coronary sinus. Shunting for malignant ascites has a higher incidence of DIC and malignant cells dissemination. In the reported case, both the pathological examination of the dissected lymph nodes and analysis of the ascites were negative for malignant cells or bacteria. The evacuated ascites had been infused to the central vein before the shunting operation. As any serious complication such as sepsis, DIC or pulmonary congestion had not developed, the shunt was inserted. Ascites had been controlled well without complications. Sooriakumaran et al. [18] reviewed 11 children who underwent shunting over a 17-year period. In all of the children ascites resolved except for a child of 4 years old with lymphohistiocytosis. Since PV shunts had been used mostly in adults with chronic disease, it has not been referred how to know when the shunt should be removed [19, 20]. Sooriakumaran et al. [18] recommended elective removal of the shunt after 1 year as five shunts were removed 1–3 years after insertion without recurrence of ascites. In the presented case, ascites had not accumulated for 20 months and the shunt was still patent and functioning [20, 21]. We ligated the infusion catheter and observed the patient for 2 weeks to confirm whether ascites should be managed without a shunt. A PV shunt is recommended in the management of ascites unresponsive to medical treatment. If the shunt is inserted for transient management of the ascites, less invasive

Table 1 Case reports of a peritoneovenous shunt in children

Case	References	Age	Sex	Etiology of ascites	Character of ascites	Duration of shunting	Complication
1	Klein [5]	1 Y	F	Budd-Chiari synd.	Serous	15 months	–
2	Chang [6]	12 Y	M	Lymphangiomas	Chylous	6 months	Obstruction
3	Altman [7]	10 M	M	Biliary atresia	Serous	?	?
4	Ryan [8]	10 Y	F	Multiple hygromas	Chylous	24 months	Obstruction
5	Guttman [9]	7 W	M	Generalized lymphatic malformation	Chylous	> 16 months	Obstruction
6	Guttman	6 Y	M	Generalized lymphatic malformation	Chylous	10 days	Obstruction
7	Press [10]	2 Y	F	Congenital lymphangiectasia	Chylous	1 month	Obstruction
8	Shah [11]	2 Y	M	Liver cirrhosis	Chylous	5 month	Obstruction
9	Man [12]	7 M	M	Idiopathic	?	?	?
10	Man	5 M	F	Hepatitis liver cirrhosis	Serous	1 day	Coronary sinus perforation
11	Man	4 Y	F	Peritonitis carcinomatosa	Chylous	> 4 years	–
12	Man	9 Y	F	Wilms' tumor post-op	Chylous	> 6 months	–
13	Kerr [13]	7 Y	M	Leukemia	Chylous	2.5 months	–
14	Salcedo [14]	9 Y	F	Hepatitis	Chylous	?	–
15	Gracia [15]	Neonate	F	Budd-Chiari synd.	Chylous	> 9 months	–
16	Gillan [16]	6 W	M	Hepatitis	Serous	8 weeks	DIC, congestion, nephritis, sepsis
17	Pettitt [17]	Neonate	M	Lymphatic malformation	Serous	2 months	Congestion
18	Presented case	5 Y	F	Neuroblastoma post-op	Chylous	> 6 months	–
						30 months	–

Y years old, M months old, W weeks old, F female, M male

methods of investigation to know when to remove the shunt need to be developed.

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