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ORIGINAL ARTICLE

The surgical experience for retroperitoneal, mesenteric and omental cyst in children

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Purpose: Intra-abdominal cystic masses originating from the retroperitoneum, mesentery or omentum are very rare and mostly benign tumors, but sometimes present as a complicated cyst encasing the major organs. **Methods:** We analyzed the clinical findings, histologic diagnosis, and surgical outcomes in children who underwent operation for retroperitoneal, omental, and mesenteric cyst from 1998 to 2010, retrospectively. **Results:** Twenty-three patients (male, 12; female, 11) underwent the operation at a median age of 46 months (range, 9 days to 16 years). Among them, 17 cysts presented one or two symptoms such as abdominal mass, abdominal pain or abdominal distension. The median duration of symptoms was 7 days (range, 1 day to 365 days). Five were detected prenatally. Ten cysts were found in retroperitoneum, 8 in the omentum and 5 in the mesentery. The median diameter was 13 cm (range, 3 to 30 cm). Twenty cysts were completely removed. Five mesenteric cysts required bowel resection and anastomosis. Three of retroperitoneal cysts were impossible to complete excise because of location and extensiveness. Pathologically, 20 cysts were lymphangioma and 3 were pseudocyst. The morbidity was one of adhesive ileus and the mortality was one who had extensive retroperitoneal cyst with mesenteric cyst. He died from sepsis. During follow-up period, there was no recurrence. **Conclusion:** Preoperative diagnosis and localization for these cysts are very difficult. Complete excision was possible in almost all cases despite the size, bringing a favorable outcome. The possibility of this disease entity should be considered as the cause of acute abdomen.

Key Words: Retroperitoneal cyst, Mesenteric cyst, Omental cyst, Lymphangioma, Pseudocyst

INTRODUCTION

An intra-abdominal cystic mass can originate not only from solid organs such as the liver, spleen, and pancreas but also from retroperitoneum, omentum, and mesentery. Retroperitoneal, omental, and mesenteric cysts are very rare intra-abdominal tumors, occurring in one of 105,000 hospitalized patients [1]. Many authors consider these cysts are derived from the same embryological structures, and with similar pathogenesis, as ectopic lymphatic tissue [1-3]. These cystic tumors are mostly benign lesions, but can present as a complicated large cyst encasing other organs. We reviewed the cases of retroperitoneal, omental, and mesenteric cysts in children. From this study, we ana-

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lyzed the clinical findings, histological diagnosis, and surgical outcome.

METHODS

We had a retrospective study of children operated upon for retroperitoneal, omental, and mesenteric cysts from January 1998 to December 2010. Intra-abdominal cystic mass from pancreas, liver, spleen, ovary, and gastrointestinal tract was excluded. The medical records were reviewed to determine the age, symptoms, preoperative diagnosis, operative findings, and histologic findings. We noted location, size and character of the contents of mass from operative records. The location of cyst was confirmed during surgery. The median follow-up period was 52 months (range, 8 to 183 months).

RESULTS

Among 23 patients, there were 11 females and 12 males. The median age at the time of operation was 46 months, ranging from 9 days to 16 years. Surgery was performed during the neonatal period in 3 patients and on four infants. Five patients were detected and followed from the prenatal period, 4 of whom were diagnosed as intra-abdominal cystic mass and one as fetal hydrops. Seventeen

Table 1. The clinical findings of retroperitoneal, omental, and mesenteric cysts

Variable	Retroperitoneum (n = 10)	Omentum (n = 8)	Mesentery (n = 5)
Operation age			
Neonates	1	0	2
Infants	3	0	1
Pre-school (1 < age ≤ 8)	4	7	2
School (8 < age ≤ 15)	2	1	0
Gender			
Male	5	6	1
Female	5	2	4
Symptom & sign			
Abdominal mass	5 (1 with anemia)	3	1 with anemia
Abdominal pain	1	4	0
Abdominal distension	0	1	2
Prenatal diagnosis	3	0	2
Incidentally	1	0	0
Duration of symptoms (day), median (range)	10.5 (1-30)	7 (2-365)	21 (7-180)
Cyst size (cm)			
≤ 10	5	0	1
11 < size ≤ 20	4	5	2
> 20	1	3	2
Contents			
Serous	6	7	3
Blood	2	1	1
Necrotic	1	0	0
Chyle	1	0	1
Pathology			
Lymphangioma	7	8	5
Pseudocyst	3	0	0
Treatment			
Complete excision	7	8	5 (including bowel R&A)
Partial excision & OK-432 injection	1	0	0
Biopsy only	2	0	0

R & A, resection & anastomosis.

patients presented with one or two clinical symptoms, such as palpable intra-abdominal mass in 9 patients, abdominal pain in 5 patients, and abdominal distension in 3 patients. Two patients had severe anemia. The median duration of symptoms was 7 days (range, 1 to 365 days). In one patient, the intra-abdominal mass was incidentally detected during the evaluation of congenital heart disease (Table 1).

For imaging diagnosis, ultrasonography or computed tomography was performed. Lymphangiography was performed in one patient and magnetic resonance imaging was checked in one patient, respectively. However, despite imaging studies, the preoperative localization of cysts was possible only in 3 patients. Based on operative findings, 10 cysts were located in retroperitoneum, 8 in omentum, and 5 in mesentery. Among 10 retroperitoneal cysts, 2 involved the mesentery and 1 extended to the buttock. Among 8 omental cysts, 4 extended to the lesser sac. Among 5 mesenteric cysts, 3 were originated in jejunum, 1 in ileum and 1 in sigmoid.

The median size of cysts was 13 cm (range, 3 to 30 cm) in largest diameter. Six were smaller than 10 cm, and 6 were larger than 20 cm. The nature of cyst fluid was serous in 16 cysts, serousanguinous in 2, chyle in 2, and necrotic material in 1. The other 2 cysts contained a lot of blood, which caused significant anemia. Twenty cysts were completely removed. Simultaneous bowel resection was needed in 5 mesenteric cysts. Three of retroperitoneal cysts were impossible to complete excision because of its location and extensiveness of the lesions. OK-432 was injected into the remaining cyst of buttock after partial excision of retroperitoneal cyst in one patient and biopsy alone without excision in 2 for multiple and extensive mesenteric invasion. The pathologic findings showed that 20 masses were lymphangioma (16 cystic, 4 cavernous type) and 3 were pseudocyst (Table 1).

The surgical complications after operation occurred in 2 patients. One was adhesive ileus that arose 6 months after omental cyst excision. One was post-operative mortality, an infant born at 30 weeks of gestational age with fetal hydrops. He had a 7 cm sized cystic mass at retroperitoneum and mesentery. After biopsy of the mass at age of 54 days after birth, he suffered from chronic lung disease, and post-surgical intra-abdominal infection, and died of sepsis 2 weeks after surgery.

During the follow-up period, all but the one death are doing well with no evidence of recurrence.

DISCUSSION

The intra-abdominal cystic masses were classified by location and histology. Besides the solid organs, retroperitoneum, mesentery, and omentum are the possible origin of the cystic mass. It is difficult to define the location of the cyst topographically. Many authors suggested that retroperitoneal, mesenteric, and omental cysts should be grouped together because on the basis of shared embryologic structures [1-3].

Walker and Putnam [4] stated that the size of the lesion rather than the location appears to be a more important factor in determining clinical symptomology. The intra-abdominal cystic mass could present the three ways: asymptomatic, chronic abdominal pain, acute abdomen [1]. Our series showed that most of cysts present with abdominal mass, pain, and distension. 21.7% of cysts were detected prenatally.

Retroperitoneal, mesenteric, and omental cysts could cause acute abdomen from cyst rupture, infection, hemorrhage, volvulus and extrinsic compression [1]. In general, these acute symptoms are more common in children than adults [1]. Patients under 10 years of age are different from the older group, having a shorter duration of symptoms, a higher number of emergency surgeries and lower recurrences [3]. Particularly in children less than 10 years of age, the average duration of symptoms prior to treatment is 2.2 months, compared to 9.8 months in patients older than 10 years [3]. The reason for earlier detection in children may be the smaller body habitus and abdominal cavity [5]. Our study revealed that the median age at operation was 46 months and the median duration of symptoms was 7 days. We observed that most of the retroperitoneal, mesenteric, and omental cysts occurred in young patients in our study. The median diameter of cyst was 13 cm. The largest one was up to 30 cm. It suggests that the peritoneal cyst in children could become enlarged to the entire abdomen result-

ing in rapid severe symptoms.

There are several classifications of intra-abdominal cysts based on histopathologic features: 1) cysts of lymphatic origin-lymphatic (hilar cysts) and lymphangiomas; 2) cysts of mesothelial origin-benign or malignant mesothelial cysts; 3) enteric cysts; 4) cysts of urogenital origin; 5) dermoid cysts; and 6) pseudocysts-infectious or traumatic etiology [6]. In our series, the lymphangiomas were overwhelming [7]. Histologically, cystic lymphangiomas are lined by flat endothelial epithelium with abnormally dilated lymphatics, abundant lymphoid tissues and smooth muscle present in the cyst wall [5,8,9]. D2-40 is a monoclonal antibody to lymphatic endothelial cells and serves as a more specific marker of lymphatic lineage [10]. Lymphangiomas may occur at all ages with a peak incidence in third and sixth decades of life [11], and 95% of cases occur in the neck or axilla [12]. The intra-abdominal lymphangiomas in retroperitoneum, omentum, and mesentery are very rare, and account for 3 to 9.2% of all lymphangiomas, with sites of 59 to 68% in mesentery, 20 to 27% in omentum, and 12 to 14% in retroperitoneum [13-17].

During the study period, authors reviewed 408 cases of lymphangioma. The prevalence of lymphangioma in retroperitoneal, omental, and mesentery was 4.9%. We observed the different distribution among them. The lymphangioma of omentum was 40%, retroperitoneum was 35% and mesentery was 25%. Abdominal lymphangiomas are usually asymptomatic and should be differentiated from other asymptomatic abdominal cystic masses such as cystic teratomas, mucinous cystadenomas, bronchogenic cysts, ovarian cysts, nonpancreatic pseudocysts, and complicated ascites [18,19]. In our study, the patients almost had a tumor-related symptom. However, it doesn't mean that all of abdominal lymphangioma cause the acute symptoms, because we included only the patients who underwent the operation. Unfortunately, there are no specific findings differentiating the origin or nature of the intra-abdominal cysts in imaging studies. Factors that help with differential diagnosis include presence of septa, thickness of the wall, and presence of calcification of the wall or displacement of the bowel [4,8]. Lymphangioma usually shows a multiloculated cyst without discernible wall on computed tomography (CT) [8]. Non-pancreatic pseudocyst shows a unilocular or multilocular cyst with abundant debris and enhancing wall, located in either mesentery or omentum. Mesothelial and enteric cyst show as an anechoic, thin walled cyst. Enteric duplication cyst are found as a unilocular cyst with an enhancing wall on CT. Cystic mesothelioma and lymphangioma will have a similar radiologic finding because they have thin walls even though mesothelial cyst wall lined by mesothelial cell without smooth muscle. They occur most commonly in adult females and tend to recur, requiring multiple operations [8,20]. In our study, the preoperative localizations were possible only in 13.0%, because imaging studies showed huge cystic masses that could not differentiated from lymphangioma, pseudocyst and duplication cyst. The treatment of choice is complete surgical resection. It is recommended as soon as possible after the diagnosis is established, because the complications such as infection, hemorrhage, and bowel obstruction can occur. As time passes, the growth of the cyst might prevent complete removal [12,13]. The other treatment modalities such as aspiration, drainage, and irradiation give poor results [7,13,21,22]. Injection of OK-432 for the unresectable lymphangioma is recommended [23,24]. Generally, patients with retroperitoneal cysts were more likely to have incomplete excision of the cyst and therefore had a higher rate of recurrence. They also required marsupialization more often [3]. In this study, we found out that it is impossible to complete excision in those patients with extensive retroperitoneal lymphangioma and mesentery lymphangioma. The risk of short bowel syndrome and possibility of damage of vital organs must be balanced with the gain of aggressive excision. In that case, OK-432 injection for lymphangioma is the easy and feasible treatment. Even in neonates, OK-432 injection is known as a safe treatment modality in head and neck as well as in intra-abdominal organ [23,24].

Overall the main complication is recurrence, which has been demonstrated to occur in 9.5% of patients, often when resection was incomplete [7]. Hebra et al. [16], who followed 13 patients with mesenteric lymphangiomas and 6 with omental lymphangiomas from 1 month to 17 years, reported 3 (16%) recurrent mesenteric lymphangiomas.

During follow-up period of our study, we had no recurrences, even in incomplete excised cysts. One death was caused from intra-abdominal infection combined with chronic lung disease in a premature baby.

In summary, retroperitoneal, mesenteric, omental cysts in children are a rare benign disease but present with huge masses and acute symptoms very early in their lives. Preoperative diagnosis and localization are very difficult. Irrespective of the origin, lymphangiomas are overwhelming, pathologically. Complete excision was possible in almost cases despite size, and that brought a favorable outcome. In case of the cyst that invaded a vital organ or much of the mesentery, partial excision with OK-432 injection is a feasible treatment option. The possibilities of this diagnosis should be kept in mind for the patients with acute abdominal symptoms. It could help early diagnosis and lead to treatment with excellent outcome.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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REFERENCES

1. Vanek VW, Phillips AK. Retroperitoneal, mesenteric, and omental cysts. Arch Surg 1984;119:838-42.
2. de Perrot M, Brundler M, Totsch M, Mentha G, Morel P. Mesenteric cysts. Toward less confusion? Dig Surg 2000;17:323-8.
3. Kurtz RJ, Heimann TM, Holt J, Beck AR. Mesenteric and retroperitoneal cysts. Ann Surg 1986;203:109-12.
4. Walker AR, Putnam TC. Omental, mesenteric, and retroperitoneal cysts: a clinical study of 33 new cases. Ann Surg 1973;178:13-9.
5. Goh BK, Tan YM, Ong HS, Chui CH, Ooi LL, Chow PK, et al. Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients. World J Surg 2005;29:837-40.
6. Huis M, Balija M, Lez C, Szerda F, Stulhofer M. Mesenteric cysts. Acta Med Croatica 2002;56:119-24.
7. Steyaert H, Guitard J, Moscovici J, Juricic M, Vaysse P, Juskiewenski S. Abdominal cystic lymphangioma in children: benign lesions that can have a proliferative course. J Pediatr Surg 1996;31:677-80.
8. Ros PR, Olmsted WW, Moser RP Jr, Dachman AH, Hjermstad BH, Sobin LH. Mesenteric and omental cysts: histologic classification with imaging correlation. Radiology 1987;164:327-32.
9. Takiff H, Calabria R, Yin L, Stabile BE. Mesenteric cysts and intra-abdominal cystic lymphangiomas. Arch Surg 1985;120:1266-9.
10. Kalof AN, Cooper K. D2-40 immunohistochemistr: so far! Adv Anat Pathol 2009;16:62-4.
11. Garcia M, Louis LB 4th, Vernon S. Cystic adrenal lymphangioma. Arch Pathol Lab Med 2004;128:713-4.
12. Kosir MA, Sonnino RE, Gauderer MW. Pediatric abdominal lymphangiomas: a plea for early recognition. J Pediatr Surg 1991;26:1309-13.
13. Hancock BJ, St-Vil D, Luks FI, Di Lorenzo M, Blanchard H. Complications of lymphangiomas in children. J Pediatr Surg 1992;27:220-4.
14. Chateil JF, Brun M, Vergnes P, Andrieu de Lewis P, Perel Y, Diard F. Abdominal cystic lymphangiomas in children: presurgical evaluation with imaging. Eur J Pediatr Surg 2002;12:13-8.
15. Ates LE, Kapran Y, Erbil Y, Barbaros U, Dizdaroglu F. Cystic lymphangioma of the right adrenal gland. Pathol Oncol Res 2005;11:242-4.
16. Hebra A, Brown MF, McGeehin KM, Ross AJ 3rd. Mesenteric, omental, and retroperitoneal cysts in children: a clinical study of 22 cases. South Med J 1993;86:173-6.
17. Okur H, Kucukaydin M, Ozokutan BH, Durak AC, Kazez A, Kose O. Mesenteric, omental, and retroperitoneal cysts in children. Eur J Surg 1997;163:673-7.
18. Wilson SR, Bohrer S, Losada R, Price AP. Retroperitoneal lymphangioma: an unusual location and presentation. J Pediatr Surg 2006;41:603-5.
19. Yang DM, Jung DH, Kim H, Kang JH, Kim SH, Kim JH, et al. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. Radiographics 2004;24:1353-65.
20. Carpenter HA, Lancaster JR, Lee RA. Multilocular cysts of the peritoneum. Mayo Clin Proc 1982;57:634-8.
21. Roisman I, Manny J, Fields S, Shiloni E. Intra-abdominal lymphangioma. Br J Surg 1989;76:485-9.
22. Konen O, Rathaus V, Dlugy E, Freud E, Kessler A, Shapiro M, et al. Childhood abdominal cystic lymphangioma. Pediatr Radiol 2002;32:88-94.
23. Oliveira C, Sacher P, Meuli M. Management of prenatally diagnosed abdominal lymphatic malformations. Eur J Pediatr Surg 2010;20:302-6.
24. Luzzatto C, Lo Piccolo R, Fascetti Leon F, Zanon GF, Toffolutti T, Tregnaghi A. Further experience with OK-432 for lymphangiomas. Pediatr Surg Int 2005;21:969-72.