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Citation	日本外科宝函 (1987), 56(1): 77-84
Issue Date	1987-01-01
URL	http://hdl.handle.net/2433/204003
Right	
Type	Departmental Bulletin Paper
Textversion	publisher

Kyoto University

Arch Jpn Chir 56(1), 77～84, Nov., 1987

A Completely Resected Case of Giant Congenital Liver Cyst

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Received for Publication, Sept. 8, 1986.

Many classifications have been proposed for cystic lesions of the liver. Becker[2] classified congenital liver cysts into four groups: 1. solitary cysts, 2. congenital polycystic dieseases, 3. congenital hepatic fibrosis, and 4. congenital dilatation of the intrahepatic bile ducts. The former two were called primary parenchymal cysts by Wellwood et al.[21] and constituted the majority of congenital liver cysts. Among the cases of solitary cysts, a large primary cyst associated with some small daughter cysts is occasionally found. Koyama et al.[9] proposed the following classification of liver cysts for clinical application: 1. localized type; relatively small number of cysts limited to the unilateral lobe of the liver, 2. scattered type; relatively small number of cysts scattered in bilateral lobes of the liver, and 3. diffuse type; so-called polycystic liver with multiple cysts occupying the bilateral lobes of the liver, and frequently associated with a polycystic kidney.

As to the operative procedures for congenital primary cysts of the liver, deroofing[3] or fenestration[11] is chosen besides cystectomy or hepatectomy. In the cases of giant cysts occasionally seen among localized and scattered types, surgical removal of the whole cyst is controversial.

Case Report

A 63-year-old female complaining of a large, painless mass in the abdomen and general malaise was admitted on May 4, 1985. Although she had palpated the mass for 3 years, rapid development of the mass had started one month before admittance. Her brother had hypertension and her sister had died of cardiac insufficiency. The patient had undergone appendectomy at the age of 40, had been admitted to a hospital for one month because of pyelonephritis, and had rheumatoid arthritis since the age of 59. Physical examination showed a relatively healthy woman of 150 cm in height, weighing 51 kg, blood pressure 106/74 mmHg, regular heart rate of 54/min, and with regular respiration. She had no jaundice, the conjunctivae were not anemic, no abnormal breath sounds or heart murmurs were audible, and lymphnode enlargement was not

Key words: Congenital liver cysts, Giant liver cysts, Total cystectomy.
索引語：先天性肝嚢胞，巨大肝嚢胞，嚢胞全摘出術.
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observed over the entire body surface. On abdominal palpation, a smooth, soft, elastic mass occupying the entire right abdomen and the left upper quadrant of the abdomen, with some respiratory wandering, was found. The kidneys and spleen were not palpable. No edema was found in the legs.

Laboratory tests including hematology, urinalysis, liver chemistry studies, renal function, serum amylase, CEA and alpha-fetoprotein assays showed no abnormal results. Gastrointestinal radiography revealed marked ptotic displacement of the stomach. The stomach had a normal mucosal pattern. Drip intravenous pyelogram showed the right renal pelvis dislocated medially and inferiorly and the right ureter kinked, but with a normal diameter. Drip intravenous cholangiography showed the gallbladder descending into the pelvic cavity. A unilocular cyst occupying the entire right lobe of the liver appeared on an ultrasonogram (US) of the abdomen. No intracystic debris or any irregularity of the cyst wall was identified. In addition to the giant cyst, another small cyst was found in the left lobe. Computed tomography (CT) of the abdomen revealed a lage mass with homogeneous low density taking the place of the right hepatic lobe (Fig. 1). Marked displacement of the hepatic vessels to the left due to the avascular cystic mass was found by selective celiac angiography. (Fig. 2)

With a tentative diagnosis of a giant congenital liver cyst in the right lobe, abdominal exploration was carried out on June 12, 1985. There were no ascites. A yellow-reddish liver cyst of the right lobe with some thin capsules was found. In consideration of the results of intraoperative US careful dissection of the cyst wall from the liver parenchyma was performed using an ultrasonic scalpel. Structures suggesting fine vessels or bile ducts were meticulously ligated and divided. Nothing was done to the small cyst in the left lobe during the surgery. The re-

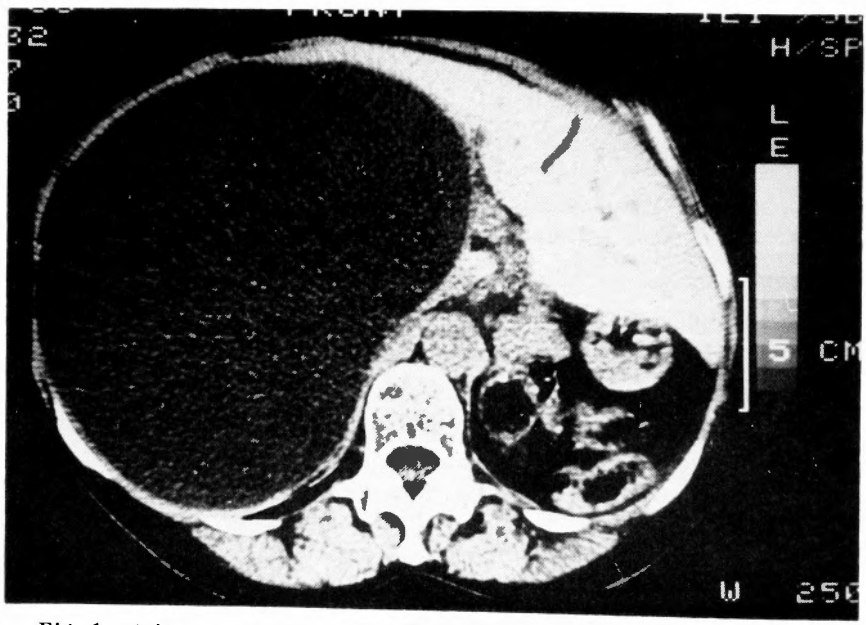

Fig. 1. A large cystic mass taking the place of the right hepatic lobe on CT

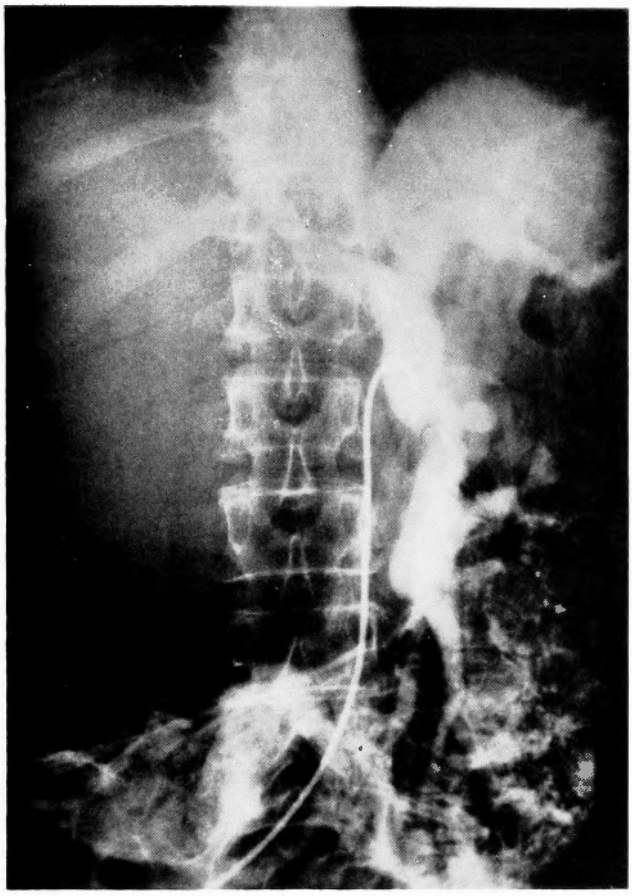

Fig. 2. Marked displacement of the vessels to the left due to an avascular mass on celiac angiogram

sected specimen was a 2,850 g, 23×22×6 cm cyst on the flat plate. In cross section it was revealed that as a unilocular cyst with a wall 2 to 3 mm thick, containing clear yellowish fluid whose cytology was Pap. II, CEA 5 ng/ml, with negative relults of microbe cultures. (Fig. 3) Photomicroscopic sections revealed that the lining of the cyst wall was one layer, with small cuboidal epithelia; the wall was partially hyalinized connective tissue. (Fig. 4) The final diagnosis was a congenital liver cyst.

The postoperative course was uneventful. The patient was discharged on the 24th postoperative day, and has been well. CT and US of the abdomen one year after the surgery revealed that the resected space was occupied by intestines, the right kidney had returned to the normal position, and there was no enlargement of the small cyst in the left hepatic lobe.

Discussion

Cases of cystic lesions of the liver are being reported in an increasing number recently. Diseases assumed to be congenital parenchymal liver cysts were recorded in Japan in a total of 635

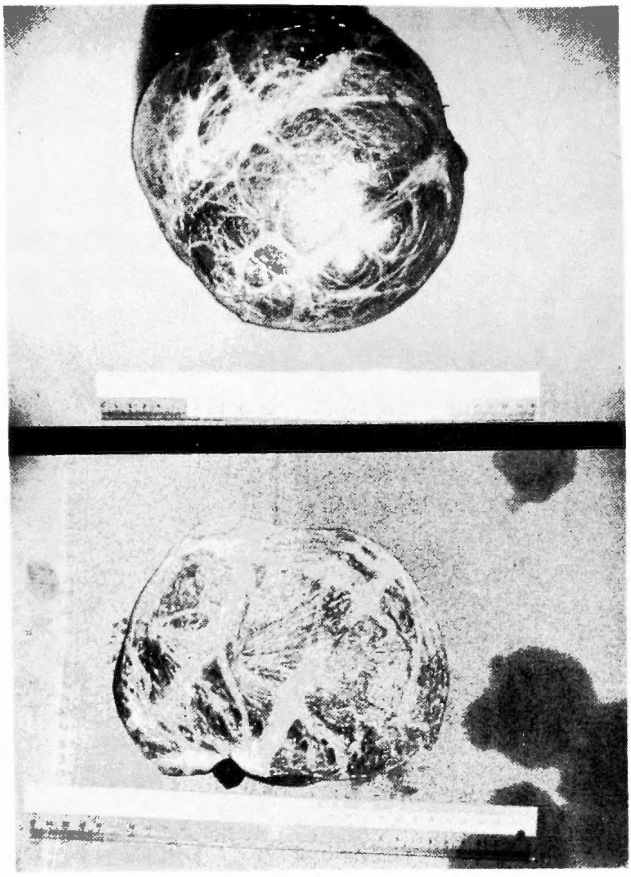

Fig. 3. A resected cyst (upper) and internal surface of the cyst (lower)

cases in an 84-year period from 1896 to 1979, according to TONODA et al.[20] and FUJITA et al.[5], but in the past six years since 1980, the statistics amounted to 318 cases. In these 318 cases, the male-to-female ratio was 1 : 3.1, and the largest age group were patients in the 60s. As classified by KOYAMA et al.[9], the 277 recorded cases consisted of 156 cases of the localized type, 23 cases of the scattered type, and 98 cases of the diffuse type.

Congenital liver cysts are mostly asymptomatic, and whether solitary or multiple, 55 per cent of the cysts are palpable as liver swelling or abdominal mass, or 35 per cent occur as abdominal pain.[17] In the present survey, in 236 cases in which symptoms were recorded, the abdominal mass was palpable in 106 cases, or 45%, and abdominal pain was noted in 87 cases, or 37%, while jaundice, abdominal distention, and geneal malaise were observed in some cases, but none of these symptoms were pathognomonic. However, liver cysts, larger than 10 cm in diameter, are reported to be often accompanied by some notable symptoms.[10] In almost all recent reports, liver cysts are being accurately diagnosed by imaging techniques, such as US and CT. Since it is now possible to observe the process easily by these imaging modalities, surgical application to liver cysts which are benign and less painful diseases should be strictly differentiated.

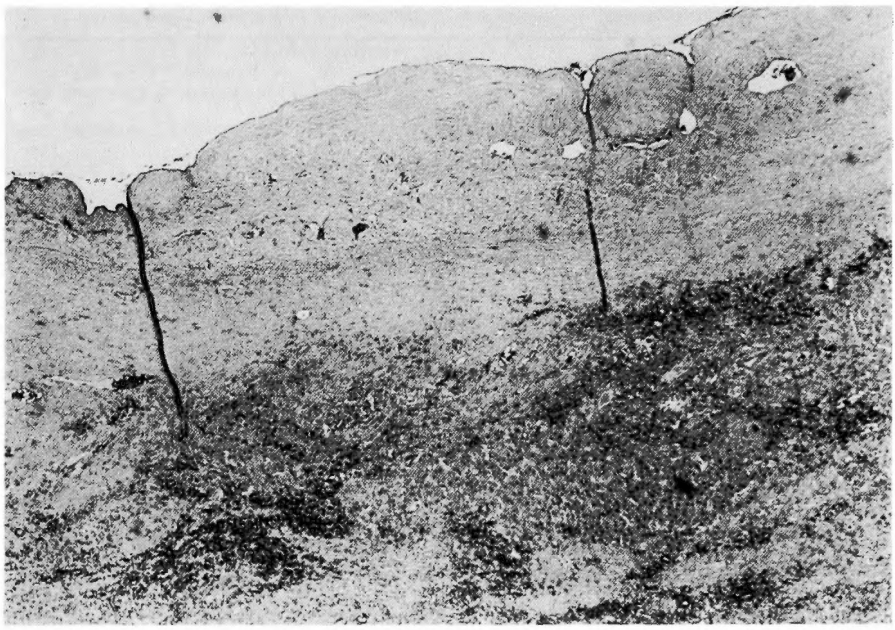

Fig. 4. Photomicroscopic specimen of the cyct wall showing monolayer lining epithelia on the connective tissue (HE ×40)

Absolute surgical indications for congenital liver cysts are rupture, infection, intracystic bleeding, and torsion of the cyst. MORGENSTERN[14] discovered rupture in 4 out of 250 cases, or 1.6%, and reported that the maximum cyst diameter of the ruptured cases ranged from 15 to 35 cm. In the present survey, rupture was noted in 3 out of 318 cases, or 0.9%, and infection was recognized in 8 cases, or 2.5%, and intracystic bleeding in 3 cases, or 0.9%. No case of torsion was found. Other surgical indications may also include progress into cystadenocarcinoma[4,7], but the actual case is very rare. However, should the irregularity of cystic wall, multilocular structure, or abnormal angiographic findings be observed, the possibility of malignancy must be taken into consideration. In addition, oppression of the respiratory or digestive system by cysts, or a suddenly increasing tendency of cyst development should be reagrded as surgical indications.

In the present data gathering, the maximum cyst diameter was recorded in 102 cases, a breakdown of which is given in Table 1. Cysts measuring more than 15 cm in maximum diameter are considered to have the possibility of rupturing, and also considering the possibility of other complications occurring, HADAD et al.[6] recommended to perform surgery when the maximum diameter was over 10 cm, while SEKIGUCHI et al. [18] suggested the maximum diameter

Table 1. Size and types of 102 cases of congenital liver cysts

	Localized	Scattered	Diffuse
More than 20 cm	11	1	1
15-19 cm	25	2	3
10-14 cm	21	2	1
Less than 10 cm	25	4	6

Table 2. Relationship between the size and treatments in 102 cases of congenital liver cysts

	Cystectomy	Hepatectomy	Deroofing or fenestration	Internal or external fistula	Ethanol injection	Only the aspiration	Unrecorded or not operated
More than 20 cm	3	1	6				3
15-19 cm	11	2	12	3			3
10-14 cm	8	4	6		3		3
Less than 10 cm	4	2	10	4	6	5	4

of 8 cm as a surgical indication for the Japanese people. Giant cysts of over 20 cm should be treated, without any question, if asymptomatic. Surgical techniques must be selected in consideration of, aside from cyst size, location and type of cyst, age of patient, and other relevant data.

Table 2 shows surgical techniques in relation to the cyst diameter. LONGMIRE et al.[12] suggested that the best method for a liver cyst was cystectomy, but in difficult-to-remove cases, safer techniques, such as deroofing should be selected. Giant cysts over 20 cm were recorded only in 13 cases in the present survey, of which deroofing was applied in 6 cases and cystectomy in 3 cases. As the maximum diameter became larger, the number of cases of cystectomy tended to decrease. If the extirpation of giant cyst seems to be difficult, the method may be changed to subtotal cystectomy or deroofing. In such cases, it may also be useful to try to fix the remaining cystic epithelium by laser[13,18] or ethanol[13]. Hepatectomy is generally regarded to be an excessive stress for the benign liver cyst, and if hepatectomy is selected as the final choice, every effort should be made to minimize the wound area. Communication between the liver cyst and bile duct was noted only in four cases in the present total, and in these cases, where removal of the cyst was difficult, cyst-enterostomy was selected.

Recently it is being attempted to eliminate or reduce cysts by fixing the lining epithelia of congenital liver cysts by injecting ethanol through an US-guided cyst puncture and puncture catheter[1,19]. Requiring no laparotomy, it is advantageously applied to poor risk patients and for relatively large cysts, ethanol may be injected twice or more[1] Of course, before injecting ethanol, it must be ensured that there is no communication with the bile duct, and the cases must also be proven not to be an acquired hydatid or neoplastic cyst. In multilocular or suspected malignant cysts, adequate follow-up after injection is necessary.

Operative deaths due to congenital liver cysts are not found in the statistics by SANFELIPPO et al.[16], and in the present survey, in the 122 cases in which prognosis was mentioned, death was recorded in 4 cases, or 3.3%. By puncture and suction of the cyst alone, recurrence is said to be inevitable[15]. In the cases undergoing treatments other than puncture and suction, total cystectomy or hepatectomy, only one case presented a complication of persistence of exudate after a construction of external fistula.

KOBAYASHI et al.[8] detected, in 4 our of 7 cases of congenital liver cyst, either a re-collection of cystic fluid in the opened cyst, an increase of the residual daughter cysts, or neoplasty of other cysts by imaging modalities, and warned that a long postoperative follow-up was necessary if they

were clinically asymptomatic. In giant cysts of over 20 cm, if the cyst wall is left over, re-collection of cystic fluid and the accompanying re-proliferation of the cyst is considered highly possible, and in order to avoid such phenomena, it seems wise to perform total cystectomy, as far as possible. In actual extirpation, it is important to make use of US during the operation, and if a malignant cyst is suspected, the use of intraoperative biopsy or a change of the surgical techniques should be considered.

Acknowledgments

We extend our gratitude to Prof. S. Hashimoto and Dr. T. Asakawa of the Second Department of Pathology, Kinki University School of Medicine for pertinent guidance.

A part of this study was reported at the 138th meeting of the Kinki Surgical Society on November 30, 1985 at Sayama-Cho, Osaka, Japan.

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和文抄録

巨大先天性肝嚢胞の1切除例

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園部 鳴海，山田 幸和，久山 健

急速に最近増大の無痛性腹部腫瘤を主訴とした63歳の女性で，右葉全域を占める巨大肝嚢胞が発見された．肝左葉にも小嚢胞の合併がみられた．右葉の嚢胞の完全摘出を施行し，術後経過は良好であった．組織学的に先天性肝嚢胞と診断された．同嚢胞の本邦報告例は1979年までで635例であったが，1980年以降の数年で318例と急増がみられた．1980年以降の例では最大径20 cm をこえる嚢胞は13例のみであり，嚢胞摘出を受けたのは3例のみであった．先天性肝嚢胞の治療については種々の方法が試みられており，それらの点についての若干の考察を加えた．