

CASE REPORT

Huge splenic epidermoid cyst with elevation of serum CA19-9 level

Sayo Matsumoto^{1,2}, Toshifumi Mori¹, Jinsei Miyoshi², Yoshitaka Imoto¹, Hirohiko Shinomiya¹, Satoshi Wada¹, Toshihiro Nakao³, Hisamitsu Shinohara³, Sadahiro Yoshida³, Keisuke Izumi⁴, Jun Okazaki², Naoki Muguruma², and Tetsuji Takayama²

¹ Department of Gastroenterology, Oe Kyodo Hospital, ² Department of Gastroenterology and Oncology, Institute of Health Biosciences, the University of Tokushima Graduate School, ³ Department of Surgery, Oe Kyodo Hospital, ⁴ Molecular and Environmental Pathology, Institute of Health Biosciences, the University of Tokushima Graduate School, Tokushima, Japan

Abstract : A 30-year-old female was referred to our hospital for further examination of liver dysfunction. A huge, soft mass was noted in her left upper quadrant on physical examination. Abdominal ultrasonography and computed tomography revealed a huge cystic tumor of 20 cm in the hilus of the spleen. Serum CA19-9 was 491 U/ml, and splenectomy was performed under suspicion of a malignant cystic tumor. The inner surface of the cyst was lined by squamous epithelial cells that were immunohistochemically positive for CA19-9. Serum CA19-9 level was normalized after the surgery. Our case of a very rare, huge epidermoid cyst of the spleen suggests that measurement of the serum CA19-9 level is useful for evaluating therapeutic efficacy of a splenic epidermoid cyst. *J. Med. Invest.* 62 : 89-92, February, 2015

Keywords : splenic cyst, CA19-9, splenectomy

INTRODUCTION

Splenic cyst lesions are very rare although they are detected more frequently after development of modern imaging technology. They are classified into true cyst (primary, 25%) and pseudocyst (secondary, 75%), according to the presence of an epithelial cellular lining inside the cyst (1-4). Splenic epidermoid cyst, a true cyst, constitutes approximately 10% of total cysts and is more commonly encountered in individuals under 40 years of age and in females (2, 5). There have been several case worldwide reports of splenic true cysts that showed elevated serum carbohydrate antigen 19-9 (CA19-9) levels (6, 7). Here we report on a case of huge splenic epidermoid cyst, a rare type of splenic cyst, with an elevated serum CA19-9 level, and discuss the significance of CA19-9 for splenic epidermoid cysts.

CASE REPORT

A 30-year-old female was referred to our hospital for further examination of liver dysfunction discovered in a health checkup. She was asymptomatic and had no significant past medical, surgical, or traumatic history. There was no history of alcohol drinking. A huge soft mass was noted in her left upper quadrant on physical examination. A hematological examination showed slight liver injury (GOT 36 U/L, GPT 44 U/L) and the level of the serum tumor marker CA19-9 was elevated to 491 U/ml (normal : 0-37 U/ml) (Table 1). Abdominal ultrasonography revealed a huge cystic tumor of 20 cm with wall calcifications in the hilus of the spleen (Figure 1). The spleen was compressed by the cyst which was markedly swollen but the splenic vein seemed intact. No masses were found in the pancreas, liver, kidney, and gastrointestinal tract.

Received for publication August 12, 2014 ; accepted September 19, 2014.

Address correspondence and reprint requests to Naoki Muguruma, Department of Gastroenterology and Oncology, Institute of Health Biosciences, the University of Tokushima Graduate School, 3-18-15, Kuramoto-cho, Tokushima city, 770-8503, Japan and Fax : +81-88-633-9235.

Table 1. Laboratory data

〈Hematology〉		〈Blood Chemistry〉		〈Serological test〉	
WBC	7930 /mm ³	GOT	36 IU/l	CRP	0.05 mg/dl
RBC	462×10 ⁴ /mm ³	GPT	44 IU/l	HBsAg	(-)
Hb	12.9 g/dl	LDH	194 IU/l	HBcAb	(-)
Ht	40.2 %	ALP	181 IU/l	HCVAb	(-)
Plt	24.2×10 ⁴ /mm ³	γ-GTP	30 IU/l		
		T-Bil	0.58 mg/dl	〈Tumor Marker〉	
〈Coagulation〉		BUN	11.8 mg/dl	CEA	3.1 ng/ml
PT	109.3 %	Cr	0.54 mg/dl	CA19-9	491 U/ml
HPT	93.0 %	Na	139 mEq/l		
		K	3.7 mEq/l	〈Immunological Test〉	
		Cl	105 mEq/l	ANA	(-)
		TP	7.6 g/dl	LKM-1	(-)
		Alb	4.5 g/dl	AMA	(-)
		Amy	76 IU/l	AMA2	(-)
		CPK	63 IU/l	P-ANCA	(-)
		UA	5.8 mg/dl		
		CHE	404 IU/l		
		T-cho	161 mg/dl		
		FBS	92 mg/dl		

Abdominal plain and enhanced CT also showed a huge cystic lesion as well as diffuse severe low density area in the liver suggesting fatty deposition (Figure 2a-c). Because of her high body-mass index (BMI) and the ultrasonographic finding, she was considered as having NAFLD (Non-alcoholic fatty liver disease), the possible reason for her liver injury. Abdominal MRI revealed high-intensity areas in the huge mass with both T1 and T2 weighted images, suggesting intracystic hemorrhage (Figure 3a, b). Diffusion-weighted-image (DWI) of the cyst did not show hyperintensity. Splenectomy was performed due to the huge size of the cyst and because of suspicion of a malignant cystic tumor with a high level of serum CA19-9.



Figure 1 Abdominal ultrasonography revealed a huge cystic tumor sized 20 cm with wall calcifications in the hilus of the spleen. The spleen was compressed by the cyst which was markedly swollen but the splenic vein seemed intact.

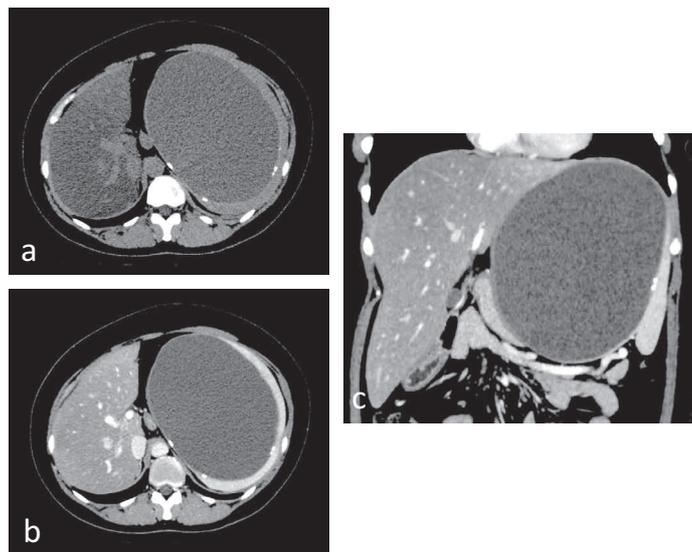


Figure 2 (a) : Abdominal CT also showed a huge cystic lesion in the hilus of the spleen and severe low density area was diffusely demonstrated in the liver. (b) : Enhanced image showed no nodule in the wall. (c) : Coronal view.

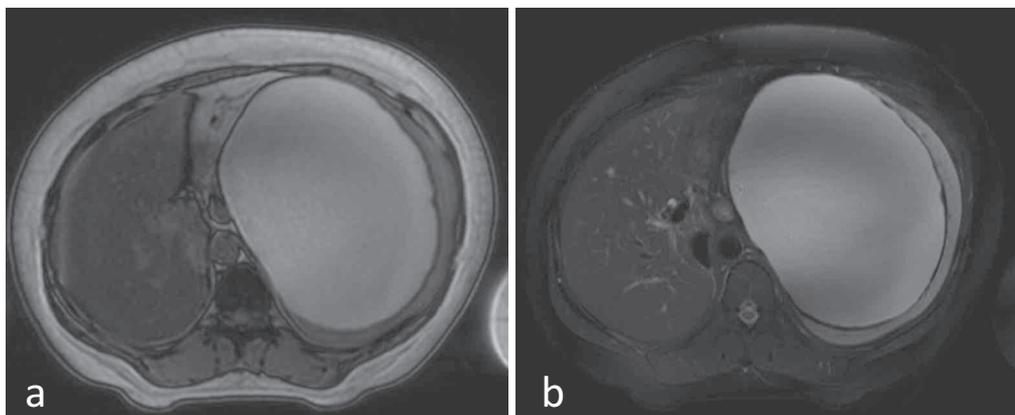


Figure 3 Abdominal MRI revealed high-intensity areas in the huge mass both with T1 (a) and T2 (b) weighted images.

Under general anesthesia, exploration identified a dark reddish strained cystic lesion with a smooth surface approximately 20 cm in size in the hilus of the spleen (Figure 4a). A needle puncture into the cyst was performed and 1,800 mL of serous brown liquid containing bloody contents were drained (Figure 4b). The cytological diagnosis of the liquid was class II. The cyst was not fixed to the stomach or pancreas and was adhesive to the superior part of the spleen. The splenic vein, short gastric vein and posterior gastric vein were intact and there was no need of vessel management. Splenectomy was performed and the shrunk cyst was completely taken out via the abdominal cavity (Figure 4c). The cyst wall was thin and partially calcified without nodularity. Histologically, the inner surface of the cyst was lined by squamous epithelial cells. Skin appendages such as hair roots or sweat glands were not observed. The cells were immunohistochemically positive for CA19-9 (Figure 5a, b), but there were no malignant findings. Thus, the lesion was diagnosed as a benign epidermoid cyst. The patient made an uneventful recovery and the serum CA19-9 level was normalized after the surgery.

DISCUSSION

Splenic true cysts, constituting 25% of splenic cysts, are subclassified into epithelial, endodermoid and parasitic cysts. Of these three types of cysts, epithelial cysts comprise epidermoid and dermoid cysts (8). In the present case, the diagnosis of the epidermoid cyst was made by histological examination of the excised spleen because the inner surface of the cyst was lined by squamous epithelial cells without skin appendages such as hair roots or sweat glands. A splenic epidermoid cyst is generally congenital and is very rare. The size of most splenic epidermoid cysts is less than 15 cm, and only a few splenic epidermoid cysts larger than 20 cm have been reported (2). The origin of epidermoid cells is still unclear, however, they are considered to develop from mesonephric tissue in the developing spleen in early embryonic life (8). In general, small epidermoid cysts are asymptomatic, but left upper abdominal quadrant pain and tenderness are the most common clinical findings. Our case was asymptomatic despite the giant size of the cyst of 20 cm in diameter.

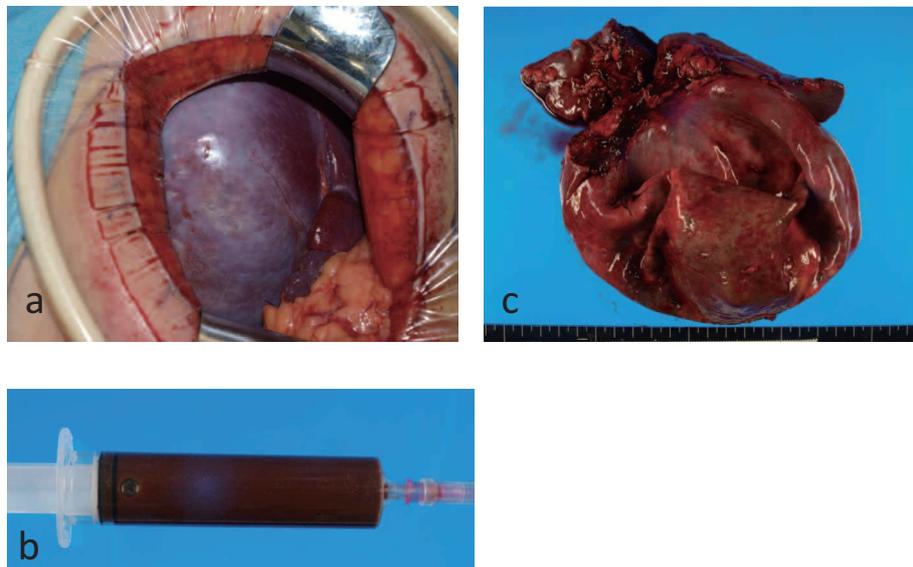


Figure 4 (a) : A dark-reddish strained cystic lesion with a smooth surface approximately 20 cm in size was identified in the hilus of the spleen. (b) : A needle puncture into the cyst revealed serous brown liquid. (c) : Retrieved specimen.

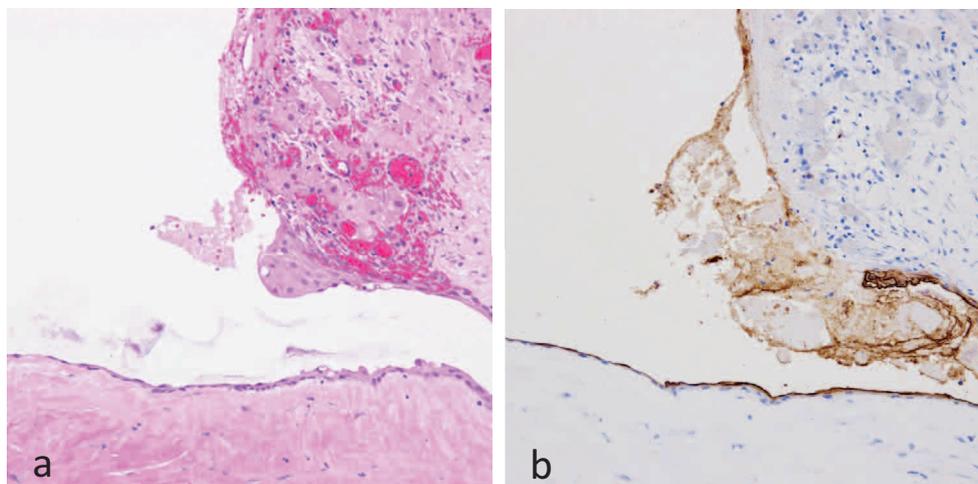


Figure 5 Histologically, the inner surface of the cyst was lined by squamous epithelia (a) and they were immunohistochemically positive for CA19-9 (b).

Total splenectomy is the treatment of choice for larger cysts more than 5 cm in diameter in order to prevent serious complications such as rupture, hemorrhage, or infection of the cysts (8, 9). In particular, cysts sized larger than 10 cm have a high risk of rupture (10), and therefore splenectomy should be performed. Moreover, when a malignant tumor is suspected, total splenectomy should be chosen because there have been some reports of splenic cyst accompanied with malignancy (11, 12). Epidermoid cysts sometimes show hyperintensity by DWI of MRI, which is one of the characteristic features of this cyst (13). In our case, however, DWI of the cyst did not show hyperintensity. We speculate that the reason is due to the fluid content of the cyst which consisted of blood, protein, and mucus. The cystic lesion in the present case was very large (20 cm), and malignancy could not be excluded because of calcification in the cystic wall, intracystic hemorrhage and a highly elevated serum CA19-9 level. Therefore, total splenectomy was performed. The splenic cyst was histologically diagnosed as a benign epidermoid cyst.

There have been several reports published on cases of splenic epidermoid cysts that showed elevated serum levels of CA19-9 (6, 7). In these cases, elevated CA19-9 levels were detected in serum and the cystic fluid, and the epidermoid cells lining the inside of the cyst wall were positive for CA19-9 by immunohistochemistry. However, there was no positive correlation between the cyst size and CA19-9 level. In the present case, the serum CA19-9 level was high (491 U/ml), but CA19-9 level in the intracystic fluid was not measured. The squamous cells lining of the cyst wall were positive for CA19-9 by immunohistochemistry (Figure 5). In addition, the serum CA19-9 level was normalized after splenectomy. The detailed mechanism of CA19-9 production in splenic epidermoid cysts remains unknown. On the other hand, elevation of serum CA19-9 levels in patients with splenic cysts of non-epithelial origin has not been reported. There are potentially many more cases that have not been evaluated for serum CA19-9 level (6) and the diagnostic significance of CA19-9 in splenic epidermoid cyst should be offered based on the sensitivity of serum CA19-9 for them. In this context, although there is a limitation in measurement of serum CA19-9 levels for diagnosing epidermoid cysts in the spleen, it should be useful as therapeutic marker for splenic epithelial (epidermoid/dermoid) cysts once it is elevated.

CONFLICT OF INTEREST

All authors disclose no conflicts of interest associated with the present case report.

REFERENCES

1. McClure RD, Altmeiter WA : Cyst of the spleen. *Ann Surg* 116 : 98-102, 1942
2. Morgenstern L : Nonparasitic splenic cysts : Pathogenesis, classification, and treatment. *J Am Coll Surg* 194 : 306-314, 2002
3. Fowler RH : Nonparasitic benign cystic tumors of the spleen. *Int Abstr Surg* 96 : 209-227, 1953
4. Martin JW : Congenital splenic cysts. *Am J Surg* 96 : 302-308, 1958
5. Qureshi MA, Hafner CD : Clinical manifestations of splenic cysts : study of 75 cases. *Am Surg* 31 : 605-608, 1965
6. Yoh T, Wada S, Kobayashi A, Nakamura Y, Kato T, Nakayama H, Okamura R : Laparoscopic splenectomy for a large multilocular splenic cyst with elevated CA19-9 : Report of a case. *Int J Surg Case Rep* 4 : 319-321, 2013
7. Hoshino A, Nakamura Y, Suzuki H, Mizutani S, Chihara N, Matsunobu T, Maejima K, Miura K, Hanawa H, Nomura S, Toyoda T, Yamagishi S, Nakata R, Muraki A, Uchida E : Giant epidermoid cyst of the spleen with elevated CA 19-9 production managed laparoscopically : report of a case. *Nippon Med Sch* 80 : 470-474, 2013
8. Rana AP, Kaur M, Singh P, Malhotra S, Kuka AS : Splenic epidermoid cyst - a rare entity. *J Clin Diagn Res* 8 : 175-176, 2014
9. Pastore V, Bartoli F : A report of a giant epidermoid splenic cyst. *Afr J Paediatr Surg* 11 : 67-70, 2014
10. Inokuma T, Minami S, Suga K, Kusano Y, Chiba K, Furukawa M : Spontaneously ruptured giant splenic cyst with elevated serum levels of CA 19-9, CA 125 and carcinoembryonic antigen. *Case Rep Gastroenterol* 4 : 191-197, 2010
11. Nimr AT, George WS : Primary reticulum-cell sarcoma of the spleen. *Radiology* 72 : 868-871, 1959
12. Shuman RH, Bouterie RH : Cystadenocarcinoma of the pancreas presenting as a splenic cyst. *Surgery* 80 : 652-654, 1976
13. Piana G, Naggara O, Oppenheim C, Rodrigo S, Meder JF, Marsico R, Page P : Epidermoid-cyst of the conus medullaris : usefulness of DWI. *J Neuroradiol* 35 : 304-305, 2008