

Giant Epidermoid Cyst of the Spleen with Elevated CA 19-9 Production Managed Laparoscopically: Report of a Case

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Abstract

True splenic cysts are uncommon and are associated with elevated serum and intracystic tumor marker CA 19-9 levels. A 33-year-old woman presented to our hospital with a chief complaint of epigastralgia. Computed tomography of the abdomen showed a 10-cm cystic lesion in the spleen. The serum carbohydrate antigen (CA) 19-9 level was 3,347 U/mL (normal, <37 U/mL). Total laparoscopic splenectomy was performed, and the serum level of CA 19-9 had normalized 2 weeks later. Pathological examination showed a benign true epidermal cyst of the spleen with strong immunohistological staining for CA 19-9. Splenic epidermoid cysts most often occur in young women, and laparoscopic surgery to remove cysts of this type is minimally invasive. Thus, laparoscopic surgery should be the method of first choice for most cases of splenic benign true cyst.

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Key words: laparoscopic splenectomy, splenic epidermoid cyst, carbohydrate antigen 19-9

Introduction

Cystic changes in the spleen remain rare, despite being detected more frequently with modern imaging technology¹. Splenic cystic lesions have been classified as parasitic and nonparasitic cysts. Nonparasitic cysts are further classified as primary cysts (true cysts) or secondary cysts (pseudocysts) according to the presence of an epithelial or mesothelial cellular lining inside the cyst¹. Approximately two-thirds of splenic cysts worldwide are parasitic cysts, most often caused by

Echinococcus granulosus. An association between a true splenic cyst and elevated serum levels of carbohydrate antigen 19-9 (CA 19-9) has recently been reported in a few patients². Because the spleen plays an important role in immunomodulation, spleen-conserving procedures, such as partial splenectomy, cystectomy, marsupialization, and cyst decompression, have recently been recommended for the treatment of benign splenic cysts. With recent advances in laparoscopic surgery, all such conservative procedures can now be performed laparoscopically³. Of these laparoscopic procedures, laparoscopic splenectomy is the least invasive and

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has the lowest morbidity rate. Laparoscopic splenectomy also reduces postoperative patient discomfort and requires a shorter hospital stay than do other laparoscopic procedures⁴. Epidermoid cysts of the spleen that are symptomatic and are associated with complications or with an abnormal increase in serum levels of tumor markers are best managed surgically.

Case Report

A 28-year-old woman was admitted to our hospital with a chief complaint of left upper quadrant pain. The physical examination revealed a splenic cyst, and blood tests showed elevated serum levels of tumor marker CA 19-9 (208 U/mL; normal, <37 U/mL). Computed tomography (CT) revealed a large splenic cyst. After the splenic cyst was diagnosed, she became pregnant and gave birth to an infant at term. She had not visited a hospital for the cyst for 5 years. When she had been 33-year-old, she had CT examination and blood test in the presented doctor. She presented with an increase in the size of the splenic cyst and a markedly elevated serum level of CA 19-9 (3,347 U/mL). The patient had a history of atopic dermatitis but no history of trauma or infection. She had no drug, alcohol, or tobacco habits. Her family history was unremarkable. Physical examination of the abdomen revealed no abnormalities other than the cyst, pulse and blood pressure were within normal ranges, and routine biochemistry tests showed no other abnormal values. The results of routine tests (electrocardiogram, X rays, blood test, etc.) were found to be within normal ranges. An ultrasound examination identified a large mass in the left upper quadrant of the abdomen. Abdominal CT showed a 10-cm cystic lesion in the spleen (**Fig. 1**). No masses were found in the pancreas, liver, kidney, or gastrointestinal tract. All symptomatic cysts and any asymptomatic cysts larger than 5 cm should be surgically removed because of the high risk of complications⁵. Therefore, on the basis of findings consistent with a diagnosis of giant true cyst of the spleen, the patient underwent laparoscopic surgery.

The spleen was approached anteriorly, with two



Fig. 1 Computed tomographic scan of the abdomen showing a 10-cm cystic lesion in the spleen

12-mm trocars and two 5-mm trocars and the patient at an angle of 45 degrees in the right decubitus position. Exploration identified a large cyst at the upper pole of the spleen which was pushing the stomach to the left. There were no other masses or lesions. The cyst was smooth, without nodularity, and blended with the splenic capsule. The general appearance was that of a benign cyst. Thus, laparoscopic splenectomy was performed by means of ultrasonic scalpel dissection, and the resected blood vessels were stapled. To complete the laparoscopic splenectomy, adhesiolysis was performed between the cyst and surrounding tissues and the spleen. To remove the resected spleen, it was guided into a vinyl bag (Endo Catch, Auto Suture Japan, Tokyo, Japan) that had been introduced into the abdominal cavity. The cyst was then drained of 370 mL of yellowish-brown fluid. The cyst was completely excised and taken out via the abdominal cavity with fragmentation. Neither a blood transfusion nor a stay in the intensive care unit was required.

The giant splenic cyst measured 10×8 cm, and there was thinning of the splenic parenchyma (**Fig. 2**). The cyst wall contained dense fibrous tissues and partial hyalinization, and the cortex was covered with squamous epithelium. The final pathological diagnosis was true benign epidermal cyst (**Fig. 3a**), and immunohistological staining was strongly positive for CA 19-9 (**Fig. 3b**). There were no malignant findings. The CA 19-9 level in the cystic fluid was 462,000 U/mL.

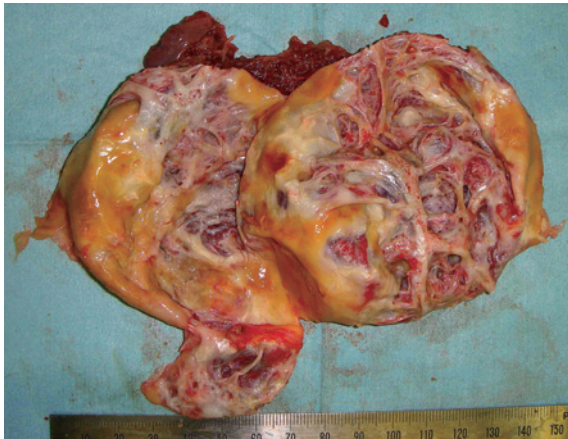


Fig. 2 Operative specimen. The giant splenic cyst measured 10×8 cm, and the splenic parenchyma showed thinning.

The postoperative course was uneventful. The patient was discharged on postoperative day 5. Follow-up CT of the abdomen 6 months later showed no recurrence of the cyst. The CA 19-9 level decreased and had normalized by 2 weeks after surgery. The patient has remained well with normal CA 19-9 levels and no recurrence for 5 years. She has been 38-year-old.

Discussion

Some nonparasitic splenic cysts are classified as either primary or secondary cysts, according to the presence or absence of an epithelial lining of the lumen¹⁶. Most true splenic cysts are epithelial in origin and are thought to contain embryonic inclusions of epithelial cells from adjacent structures⁷. To our knowledge, few cases of cystic malignant tumors of the spleen have been reported⁸⁻¹⁰. True splenic cysts can be associated with high production of tumor antigens, including carcinoembryonic antigen, CA 19-9, CA 125, and CA 50, which can be detected both in the serum and in the cystic contents. Elevated CA 19-9 serum levels have been reported in approximately 32 cases of splenic cyst, most of which were of epithelial origin^{7,11,12,15-20}. Thus, splenic cysts predominate in younger patients. Moreover, 23 of the 32 previously described patients were female. Although open splenectomy for splenic epidermoid cyst has been described, laparoscopic splenectomy has been

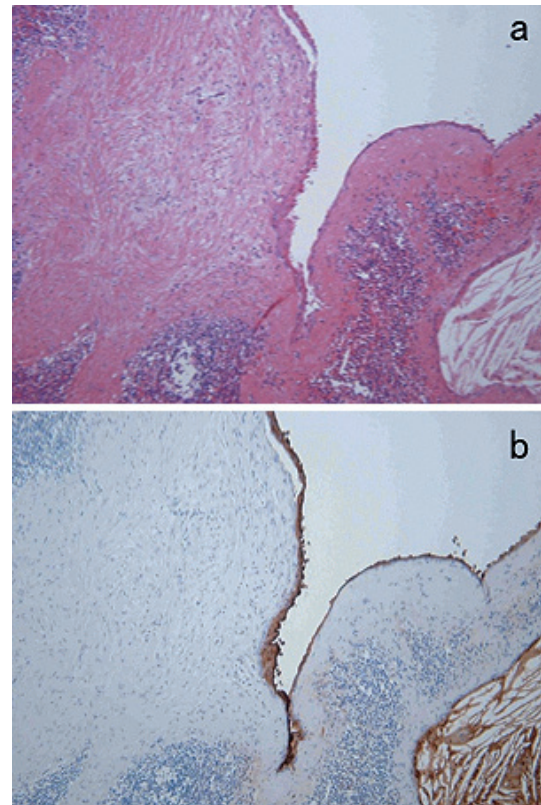


Fig. 3 (a) The cystic wall was covered with stratified squamous epithelium (hematoxylin & eosin, ×100). (b) The stratified squamous epithelium showed immunoreactivity for carbohydrate antigen 19-9 (×100).

performed for at least 4 reported cases.

In the present case, the level of CA 19-9 in serum was elevated, whereas the level of carcinoembryonic antigen was markedly elevated in the cystic fluid but normal in the serum (**Table 1**¹¹⁻¹⁴). However, the serum CA 19-9 level normalized after splenectomy. The splenic cyst contents also showed an abnormally high level of CA 19-9, and immunohistological staining for CA 19-9 was positive in the squamous epithelium of the cyst wall. These findings suggest that CA 19-9 is secreted from the splenic cyst epithelium into the bloodstream^{12,15,16}.

Surgery should be considered for cysts that are symptomatic, complicated, or antigen-producing. Complete removal of the cyst is necessary to prevent recurrence². When such cysts are treated, splenic preservation is, for immunological reasons, a legitimate concern, especially in very young patients^{11,21}. Conservative treatment is not possible, however, when the cyst occupies almost the entire

Table 1 Clinical and biochemical findings of true splenic cysts with high levels of serum tumor markers resected laparoscopically

Case	Author	Age (years)/sex	Chief complaint	Cyst size (cm)	Serum		Cyst fluid	
					CA19-9 (U/mL)	Other tumor markers (U/mL)	CA19-9 (U/mL)	Other tumor markers (U/mL)
1	Sakamoto ¹¹⁾	26/F	Abdominal fullness	13×12	217		925,466	CEA: 2,241
2	Madia ¹²⁾	19/F	Dyspeptic disorders	18×13	273	CEA: 33	2,546	CEA: 1,650
3	Chiarugi ¹³⁾	28/F	Abdominal pain	10	3,283.9	CEA: normal CA 125: 161.7	>5,000	CEA: 1,057.9 CA 125: >6,000
4	Kunishige ¹⁴⁾	28/F	none	6	491	CEA: normal	3,132,600	CEA: 320
5	Present case	33/F	Abdominal pain	10×8	3,347	CEA: normal	462,000	CEA: 440 CA 125: 51,876

CEA: carcinoembryonic antigen; CA: carbohydrate antigen

spleen and necessitates total splenectomy³.

Treatment is recommended when the cyst is symptomatic, infectious, or has ruptured, as well as when the cyst is asymptomatic, but rupture is likely, or malignancy is suspected²². Treatments include percutaneous cyst aspiration and sclerotherapy¹⁷, cyst decapsulation¹⁸, partial splenectomy²³, and total splenectomy¹¹⁻¹⁴. There is no role, in our opinion, for less-invasive procedures, such as drainage and marsupialization, associated with risks of on-site recurrence, peritoneal fluid and cell dissemination, and persistent elevation of serum tumor markers^{4,17}.

Complete removal of the splenic cyst is curative and remains a good, safe treatment for epidermoid splenic cysts²⁴. Peeling cystectomy, partial splenectomy, and total splenectomy, all performed laparoscopically, have been reported to be safe and effective. The selection of a procedure from among these alternatives depends mostly on the age of the patient, the size of the cyst, evidence of an appropriate dissection plane, and the achievement of safe vascular control. Laparoscopic splenectomy is the surgical approach of choice for splenic epidermoid cysts because of its minimal invasiveness, low morbidity, reduced postoperative pain and discomfort, and shortened hospitalization while achieving the same technical success as an open approach^{25,26}.

Conflict of Interest: The authors have no conflict of interest.

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