

Giant epidermoid cyst of the spleen with carbohydrate and cancer antigen production managed laparoscopically



Ann. Ital. Chir., 2006; 77: 443-446

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True cysts of the spleen are rare; in a few cases, high serum levels of carbohydrate and cancer antigen have been reported. In such instances, cyst resection or splenectomy is indicated to rule out malignant lesions and to remove the cancer antigen producing epithelium. We report the case of a young woman with a symptomatic giant epidermoid cyst of the spleen. Due to the secreting epithelium lining the cyst, serum levels of CA 19-9 and CA 125 were elevated. Laparoscopic splenectomy achieved symptoms relief and long-term normalization of serum tumor markers.

KEY WORDS: Laparoscopy, Splenectomy, True splenic cyst, Tumor markers.

Introduction

True primitive cysts of the spleen are uncommon. Unlike false cysts of parasitic and traumatic origin, the inner surface of the cystic wall is lined with epithelium that may produce carcinoembryonic antigen (CEA), carbohydrate antigen (CA19.9), and cancer antigens (CA125, CA50) ¹⁻⁹. These markers are found both in the serum of the patient and in the cystic fluid.

Epidermoid cysts of the spleen that are symptomatic, become complicated, or are associated with an abnormal raise in serum markers levels, are best managed by surgery.

Case report

A 28-year-old woman was referred to our Department with the chief complaint of acute-onset abdominal pain in the upper left quadrant. History revealed neither significant disease nor trauma in the past. Physical examination of the abdomen was negative, pulse and blood pressure within the normal range, and routine chemistry did not result in any abnormal value.

The patient underwent an ultrasound scan of the abdomen that showed a huge thin-walled cyst of the spleen with a small amount of free fluid in the peritoneal cavity. The abdominal CT scan detailed the lesion as a roundish, giant, splenic cyst measuring 15x13x10 cm, with a thin wall containing some small calcium deposits, and low-density, not-contrast enhanced fluid content (Fig. 1).



Fig. 1: Abdominal CT scan showing a huge splenic cyst.

Pervenuto in Redazione Gennaio 2006. Accettato per la pubblicazione Giugno 2006.

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Free peritoneal fluid was also confirmed. There were no other abdominal pathological findings.

A work-up was started to better define the nature of the cyst. Anti-echinococcus antibodies were absent, but serum analysis showed some significant abnormalities, including 3283.9 ng/ml for CA19-9 (normal value = 0-37), and 161.7 U/ml for CA125 (normal value = 0-35). Carcinoembryonic antigen and Interleukine-10 levels were within the normal range (Table I).

With a consistent diagnosis of a giant true cyst of the spleen, the patient underwent laparoscopic surgery. The spleen was approached anteriorly, using two 12-mm ports and two 5-mm ports, with the patient positioned at 45 degree on her right decubitus. As much as 500 ml of brownish, watery, free peritoneal fluid was evacuated. The left colonic flexure was taken down and the greater sac opened. Attempts to peel-off the cystic wall from the spleen were not successful due to excessive bleeding from the splenic pulp. Thus, a formal laparoscopic splenectomy was performed by means of a harmonic scalpel dissection and stapled blood vessels suture/resection (Fig. 2). The bagged specimen was delivered through a small Pfannestiel incision. The course was uneventful and the patient was discharged on day five, after having received anti-pneumococcal immunization.

The pathologist's report identified a 5 mm thick cystic wall, with an outer layer made of fibrous tissue with calcifications, and an inner layer consisting of pluri-stratified, well-differentiated, squamous epithelium. Immunohistochemistry assays on the cyst wall for epithelial cell antigens resulted negative. (Fig. 3)

Determination of antigens were made in the peritoneal and in the cystic fluid (Table I), and serially, on the patient's serum. The patient became serologically normal five months after surgery, (Table II), and so she has remained after two years of follow-up.

TABLE I – Tumor markers determination in the patient's serum (preoperatively), peritoneal fluid and cystic fluid

Antigens	Patient's serum, pre-surgery	Peritoneal fluid	Cystic fluid
CA19-9 (n.v. 0-37 ng/ml)	3283.9	>5000	>5000
CA 125 (n.v. 0-35 U/ml)	161.7	894.8	>6000
CEA (n.v. 0-5 ng/ml)	normal	5.5	1057,9
Interleukine-10 (n.v. 0-8.9 pg/ml)	10.2	normal	normal

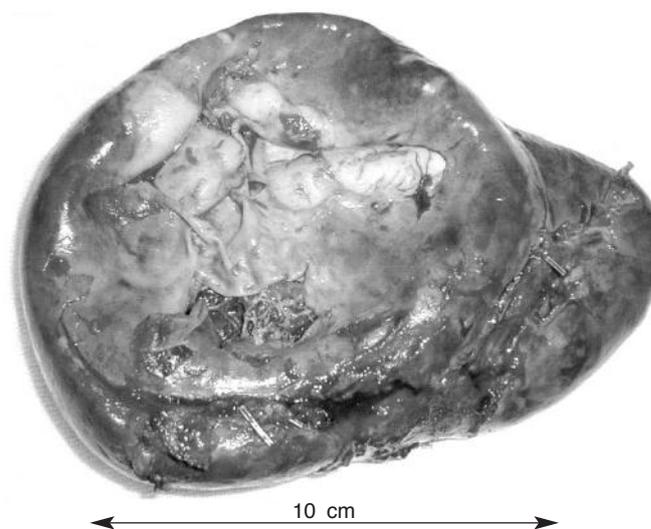
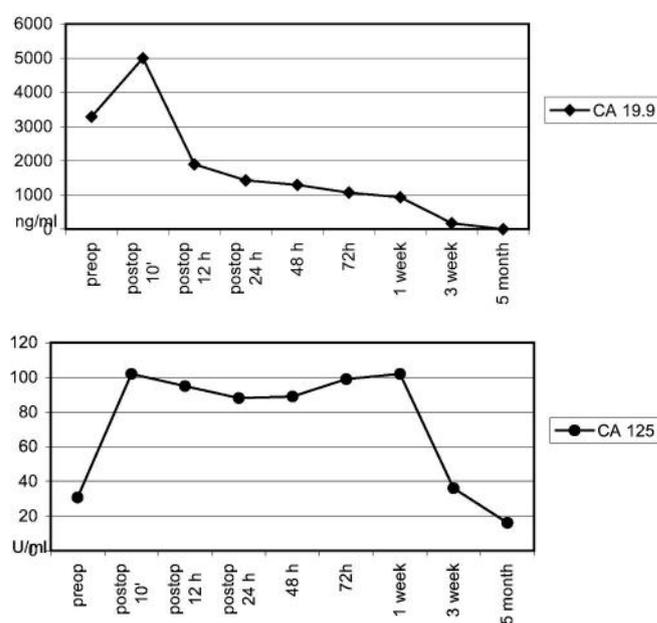


Fig. 2: Operative specimen.

TABLE II – Follow-up serum profiles of CA 19.9 and CA 125 anti-gens



Discussion

True splenic cysts are rare entities, accounting for 10-25 % of all cystic lesions of the spleen^{10,11}. Their prevalence is more common in young females. Usually, when small, they remain asymptomatic and are incidentally detected during an abdominal scan^{4,12}. Progression of the size over 5 cm diameter, may be accompanied by upper abdominal pain and/or evidence of a palpable mass in the left upper quadrant. Complications include rupture, intra-cystic haemorrhage and infection^{13,14,15}.

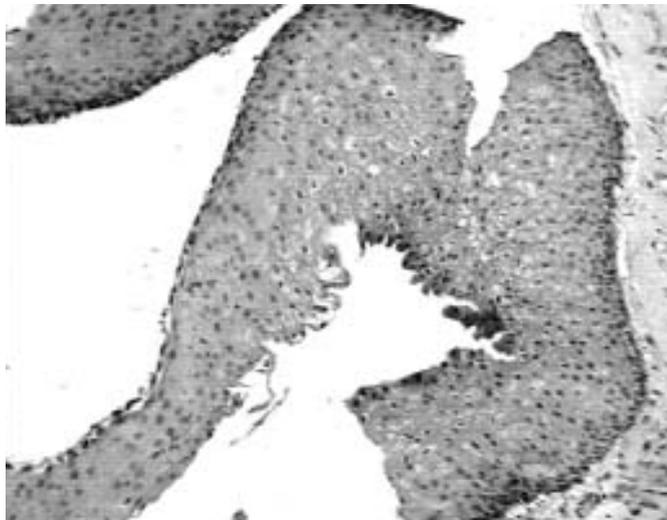


Fig. 3: Details of the multi-stratified epidermoid epithelium lining the inner surface of the splenic cyst (true cyst).

The inner surface of true cysts is lined with epithelium. On the basis of the epithelium characteristics, true cysts may be classified as epidermoid (squamous cells without cheratine production), dermoid (squamous cells with dermal structures), simple (columnar or cubic cells), and endothelial¹⁶.

The pathogenesis of true splenic cysts is still unclear. According to some authors^{9,16,17}, they could be the result of local intususception of the splenic capsula into the splenic parenchyma during embryogenesis. The pluripotent nature of the invaginated mesothelial cells would allow multiple differentiation of the epithelium.

True splenic cysts may be associated with high production of tumour antigens, including CEA, CA19-9, CA125, and CA50, which can be detected both in the serum and in the cystic content. In the latter, the antigen concentration is far higher. In the case we present, this turned out to be true for the CA19-9 and CA125 antigens, whereas CEA was strongly elevated in the cystic fluid but normal in the serum. The rise of serum tumour antigen levels poses a serious diagnostic dilemma concerning the malignancy of the cystic lesion. However, a normal IL-10 serum level in patients with true splenic cysts producing tumour antigens suggests the benign nature of the disease¹⁸.

Surgery should be considered for symptomatic, complicated or antigen producing cysts. Because of the presence of secreting epithelium, the cyst should be completely excised by accomplishing either a peeling cystectomy, partial splenectomy or total splenectomy. There is no room, in our opinion, for less invasive procedures such as drainage and marsupialization that expose the patient to the risk of on-site recurrence, peritoneal fluid and cell dissemination, and persistent high levels of serum markers^{5,12,19}.

Radical cyst surgery may be accomplished by laparoscopy.

Some reports proved the safety and the effectiveness of a peeling cystectomy^{20,21}, partial and total splenectomy²²⁻²⁶, all performed laparoscopically. The choice between these procedures is mostly dependent on the age of the patient, the size of the cyst, the evidence of an appropriate dissection plane, and the achievement of safe vascular control. In all cases, optimal outcome is warranted, along with good cosmetic results.

Conclusion

The lessons learned from this case may be resumed as follows:

- a) serum determination of CEA, CA125, CA19-9 and IL-10 should be obtained before embarking in a surgical procedure for a suspected true cyst of the spleen;
- b) should these values be elevated, then the cyst needs complete excision. This issue may require total splenectomy;
- c) laparoscopy offers a safe and effective method to manage true splenic cysts;
- d) the patient should be provided with a postoperative serum profile follow-up until the return of normal antigen levels.

Riassunto

Le cisti "vere" della milza sono rare. In alcuni casi sono responsabili di elevati livelli sierici di markers tumorali. In questi casi la resezione completa della cisti attuata mediante cistectomia, splenectomia parziale o totale, diventa necessaria per escludere lesioni maligne e per rimuovere l'epitelio cistico secernente. È riportato il caso di una giovane donna affetta da cisti epidermoide gigante sintomatica della milza con elevati livelli sierici di CA 19-9 e CA 125. Dopo splenectomia laparoscopica, si è osservato scomparsa dei sintomi e la persistente normalizzazione dei livelli sierici dei markers tumorali.

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