Reminder of important clinical lesson

Primary epithelial splenic cyst with micro-rupture and raised carbohydrate antigen CA 19-9: a paradigm of management

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Summary

Epithelial splenic cysts are rare entities that surgeons might not have previously encountered and their management is based on historical studies.

A 21-year-old female presented with a cyst of the spleen that produced a high serum concentration of carbohydrate antigen CA 19-9. A partial splenectomy with the removal of the entire cyst and preservation of splenic parenchyma by laparotomy was performed and the patient made an uneventful recovery. A microscopic rupture of the wall of the cyst and degradation of blood in the fluid of the cyst were confirmed. Eighteen months following surgery a functional splenic parenchyma was documented. There was no episode of infection during this follow-up time.

Partial surgical splenectomy with resection of the entire cyst prevents recurrence and preserves splenic function.

BACKGROUND

Primary epithelial splenic cysts are rare disorders encountered more commonly in females,¹ with 60% of instances in individuals under 40 years.² A paradigm of management is herein presented.

CASE PRESENTATION

A 21-year-old female was admitted with a 1-day history of acute left upper abdominal quadrant pain. Physical examination revealed a tender left upper quadrant but no other symptoms or signs were evident. Her medical history including history for trauma was unremarkable.

INVESTIGATIONS

Blood examinations on admission showed a haemoglobin of 10.6 g/dl, white blood cell (WBC) raised to $14 \times 10^9/l$, a platelet count of $244 \times 10^9/l$ and normal concentration of C reactive protein of 3.79 mg/l.

An ultrasound (US) scan revealed a cyst of the spleen. A CT scan showed that the cyst was $12.2 \times 11 \times 10.2 \times 11$ cm in size with a few fine incomplete septa. The cyst occupied a central position, compressing the splenic parenchyma to its periphery and was filled with liquid and debris suggesting the presence of blood (figure 1). MRI offered no additional findings.

DIFFERENTIAL DIAGNOSIS

The absence of relevant symptoms in association with a normal peripheral blood film limited the probability of a splenomegaly related to a haematology disorder. The absence of trauma history reduced the probability of a post-traumatic cyst. The lack of dense rim calcification and diaphragms confirmed by US, CT and MRI scans in association with normal serology limited the probability of the most common parasitic infection by *Echinococcus granulosus*. The

serum carbohydrate antigen (CA 19-9) was high at 478.2 U/ml (<37 U/ml), whereas the carcinoembryonic antigen (CEA) and cancer antigen 125 (CA 125) were within normal limits.

TREATMENT

Scheduled laparotomy during the same admission was performed by a left subcostal incision. The lesion was composed of strong whitish fibrous tissue partially covered by splenic parenchyma (figure 2). Its dissection was difficult because of firm adhesions to the diaphragm. A relatively bloodless dissection plan was created between the splenic parenchyma and the cystic wall, and a $7 \times 3 \times 2$ cm splenic remnant around the lower splenic pole was preserved.

Antimicrobial prophylaxis by ampicillin and sulbactam was started on the induction of anaesthesia and continued for 3 months. Vaccinations for *pneumococci*, *Haemophilus influenzae B* and *meningococci* were administered on the 15th postoperative day.

The formalin-fixed specimen weighed 895 g and was filled with brownish fluid because of the degradation of blood (figure 3). The interior surface of the cyst was glistening, white in colour and smooth, but interrupted by variable sized trabeculae which were randomly directed.

Microscopically, the wall of the cyst consisted of thick connective tissue with hyalinisation and was lined with stratified squamous epithelium (figure 4). A microscopic rupture of the wall of the cyst into the splenic parenchyma was observed. The histological diagnosis was that of a primary epithelial cyst. The patient made an uneventful recovery and was discharged on the sixth postoperative day.

OUTCOME AND FOLLOW-UP

On the sixth week following surgery, the serum CA 19-9 was normal (2.28 U/ml). Doppler US indicated that the

Figure 1 CT scan.

Figure 2 Perioperative photograph demonstrating the dissected spleen with the cyst.

splenic remnant was vascularised, and the isotope combined liver/spleen scintigraphy performed by colloid technetium Tc^{99m} demonstrated a functioning splenic remnant (figure 5).

Eighteen months following surgery the patient remained free from any episode of infection. Haemoglobin was

12.4 g/dl, WBC $5.33 \times 10^9/l$ and platelet count was $322 \times 10^9/l$. Howell-Jolly bodies were undetected in the erythrocytes of the peripheral blood film. The size of the splenic remnant assessed by US scan increased to $7 \times 3.15 \times 3.5$ cm and was homogenous and vascularised.

Figure 3 The opened formalin-fixed specimen.

Figure 4 Wall of the cyst with epithelial lining. H&E $\times 20$.

DISCUSSION

About 650 patients with non-parasitic cysts of the spleen were reported until 1978³ and more than 1000 by 2002.¹ Twenty per cent of splenic cysts remain asymptomatic. Left upper abdominal quadrant pain and tenderness are the most common clinical findings.² A variety of radiological signs,

because of adjacent organ displacement, have been reported.⁴ Details on the position, size, rim calcification, cystic component, presence of diaphragms, vascularisation and coexistence of other pathologies can nowadays be obtained by US and CT scans, which contribute to preoperative differential diagnosis.

Figure 5 Scintiscan: posterior view.

Earlier classifications of splenic cysts were based on the cyst lining.^{5 6} A recent review combining the mechanisms of pathogenesis, clinical, macroscopic and microscopic characteristics classified cysts into parasitic and non-parasitic. The latter were further subdivided into primary, including congenital and neoplastic, and secondary including traumatic and necrotic.⁷ A glistening white inner surface and trabeculation are considered features of primary epithelial cysts.⁷ The variability of the lining epithelium has been attributed to secondary alterations,⁸ and it has been proposed that when the lining epithelium is mesothelial, transitional or epidermoid the cysts can be considered primary and congenital in origin.^{1 7}

Stomata-like pores that connect the lumen of the cysts with the capillaries of the wall of the cyst or the adjacent splenic sinuses have been demonstrated by electronic microscopy.⁸ The influx of blood cells and fluid through these stomata has been implicated in the contribution of the growth of cysts and in their symptomatology.⁸ The observed degradation of blood in the fluid of the cyst and the microscopic rupture of the cyst of the present case report are indications of preceding increased tension on the wall of the cyst. Increasing tension may also explain the observed abdominal pain and tenderness.

High concentrations of CA 19-9 have been found in the serum and fluid of cysts in association with the immunoreactivity to CA 19-9 of the epithelium of primary spleen cysts.⁹ High concentrations of CEA and cancer CA 125 have also been found in the serum and fluid of primary epithelial spleen cysts.¹⁰ Tumour markers have been found despite the non-cancerous nature of the cysts.

The primary goal of surgical treatment is to remove the wall of the cyst and its lining without leaving cystic remnants in situ¹ because recurrences have been reported fol-

lowing incomplete removal.¹¹ The *en block* excision of the spleen and intact cyst achieves this goal.

The second consideration is preserving splenic parenchyma to preserve splenic function. A partial splenectomy with complete removal of the entire cyst fulfils both of the above mentioned principles. However, a partial splenectomy that leaves in situ the wall of the cyst in juxtaposition to the splenic parenchyma, often referred to as 'decapsulation', carries the risk of cyst recurrence.

Excision can be performed by open laparotomy or laparoscopic techniques.^{1 11–13} Immunisation against *pneumococci*, *H. influenzae B* and *meningococci* as well as antimicrobial prophylaxis is recommended.^{14 15}

Learning points

- ▶ Abdominal pain and tenderness were the main clinical findings.
- ▶ Degradation of blood in the fluid of the cyst and a microscopic rupture are indications of preceding increased tension.
- ▶ Raised serum CA 19-9 found in the absence of malignancy.
- ▶ Dissection plan created between the splenic parenchyma and wall of the cyst proved safe.
- ▶ Excision without leaving cystic remnants prevented recurrence.
- ▶ Preservation of splenic parenchyma preserved splenic function.

Competing interests None.

Patient consent Obtained.

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Papadopoulos IN, Davatzikos A, Kasabalis G, Manti C, Konstantoudakis G. Primary epithelial splenic cyst with micro-rupture and raised carbohydrate antigen CA 19-9: a paradigm of management. *BMJ Case Reports* 2010;10.1136/bcr.01.2010.2636, date of publication

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