Reminder of important clinical lesson

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

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Background

Epithelial splenic cysts are rare entities which surgeons may not have previously encountered. Their management is based on historical studies.

Case presentation

A 21-year-old woman presented with a cyst of the spleen that produced a high serum concentration of carbohydrate antigen CA 19-9. A partial splenectomy with removal of the entire cyst and preservation of the splenic parenchyma by laparotomy was performed and the patient made an uneventful recovery. A microscopic rupture of the wall of the cyst and blood degradation products in the fluid of the cyst were confirmed. A functional splenic parenchyma was documented 18 months after surgery. There was no infection during follow-up.

Conclusion

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 of age.² Their management is described here.

CASE PRESENTATION

A 21-year-old woman 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 past medical history including history of trauma was unremarkable.

INVESTIGATIONS

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

An ultrasound (US) scan revealed a cyst of the spleen. A CT scan showed that the cyst was $12.2 \times 11 \times 10.2$ 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

splenomegaly related to a haematological disorder. The absence of a history of trauma 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 (normal <37 U/ml), whereas carcinoembryonic antigen (CEA) and cancer antigen 125 (CA 125) were within normal limits.

TREATMENT

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

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

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

Figure 1 CT scan.

Figure 2 Intraoperative photograph showing the dissected spleen with the cyst.

Figure 3 The opened formalin-fixed specimen.

Figure 4 Wall of the cyst with epithelial lining (eosin/haematoxylin $\times 20$).

Figure 5 Scintiscan: posterior view.

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 post-operative day.

OUTCOME AND FOLLOW-UP

In the sixth week following surgery, serum CA 199 was normal (2.28 U/ml). Doppler US indicated that the splenic remnant was vascularised, and isotope combined liver/spleen scintigraphy performed with colloid technetium Tc^{99m} demonstrated a functioning spleen remnant (figure 5).

Eighteen months following surgery the patient remained free from any infection. Haemoglobin was 12.4 g/dl, WBC $5.33 \times 10^9/l$ and platelet count $322 \times 10^9/l$. Howell-Jolly bodies were not detected in the erythrocytes of the peripheral blood film. The size of the splenic remnant assessed by US scan had increased to $7 \times 3.15 \times 3.5$ cm and was homogenous and vascularised.

DISCUSSION

About 650 patients with non-parasitic cysts of the spleen were reported to 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 be obtained by US

and CT scans, which help pre-operative differential diagnosis.

Earlier classifications of splenic cysts were based on the cyst lining.⁵⁻⁶ A recent review combining the mechanisms of pathogenesis and clinical, macroscopic and microscopic characteristics classified cysts as parasitic or 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 to be 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 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 in the present case report are indications of preceding increased tension in 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 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 following incomplete removal.¹¹ En block excision of the spleen and intact cyst achieves this goal.

The second consideration is conserving splenic parenchyma to preserve splenic function. A partial splenectomy with complete removal of the entire cyst fulfils both of the above aims. However, a partial splenectomy that leaves the wall of the cyst in situ in juxtaposition with 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, *Haemophilus 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 was found in the absence of malignancy.
- ▶ Dissection between the splenic parenchyma and the wall of the cyst proved safe.
- ▶ Excision without leaving cystic remnants prevented recurrence.
- ▶ Conservation of splenic parenchyma preserved splenic function.

Competing interests None.

Patient consent Obtained.

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