Isolated alveolar echinococcosis of the spleen – clinical presentation and management review

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Summary

Alveolar echinococcosis is a zoonosis which infects primarily the liver, and secondary involvement of other organs is common. However, exclusive extrahepatic involvement is exceedingly rare, and isolated splenic involvement even more so. Workup, differential diagnosis and management of isolated splenic alveolar echinococcosis are discussed and literature is reviewed based on a clinical case.

Key words: Alveolar echinococcosis; splenic cyst; splenectomy; zoonosis; helminth infection

Alveolar echinococcosis primarily infects the liver and secondary involvement of other organs is common. However, exclusive extrahepatic involvement is exceedingly rare, and isolated splenic involvement even more so. This is in contrast with hydatid disease, where other organs than the liver are commonly the site of primary infection and splenic cysts are frequent. We would like to take advantage of a rare case of isolated splenic alveolar echinococcosis to discuss the clinical presentation, differential diagnosis and management as well as the current literature concerning this unusual presentation.

A 73 year old male patient with a history of cardio-vascular disease and hypertension was admitted to our hospital with sudden onset of left upper quadrant abdominal pain radiating to the left shoulder. Clinically the abdomen was distended, painful and showing discreet signs of peritonitis. The overall clinical condition of the patient, however, was good. There was no fever and the haemodynamic parameters were normal.

Inferior myocardial infarction was ruled out by ECG and normal cardiac enzymes. The laboratory findings did not suggest severe infection or haemorrhage: only a slight elevation of the CRP at 24 mg/ml and of the WBC at 13.4 G/l with discreet eosinophilia at 5.4% were present and the values for haemoglobin and haematocrit were slightly below normal at 135g/l and 40% respectively. A CT scan showed a large, heterogeneous splenic cyst, 11cm in diameter and a small amount of perisplenic liquid suggesting rupture of the cyst (fig. 1 and 2). Except for colonic diverticulosis there were no other pathological findings.

More in depth history revealed that the patient underwent an abdominal MRI just over a year prior to his current admission, as part of the work-up of his hypertension. The MRI demonstrated a totally asymptomatic, heterogeneous, plurilocular cystic lesion of the spleen, 6 cm in diameter (Fig. 3). At that time, the radiologist considered haemangioma or cystic lymphangioma as the most probable diagnoses. The patient was then referred to a surgeon who also considered post-traumatic, epithelial, dysgenetic and parasitic cysts. Arguing that diagnostic splenectomy seemed too heavy a procedure for a totally asymptomatic patient, the surgeon chose surveillance only and recommended a redo CT scan one year later.

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In the current presentation, rupture of the cyst with haemoperitoneum was suspected. An emergent exploratory laparotomy was performed. When entering the abdomen, there was a large amount of a milky, inodorous intraperitoneal fluid without any signs of peritonitis and no blood at all. Meticulous inspection of all intra-abdominal organs did not reveal any source for this liquid other than the splenic abscess. A splenectomy was done in the usual fashion. The post-operative course was uneventful and the patient recovered well.

Cultures of the intraperitoneal liquid remained sterile. Histopathology of the spleen showed multiple cysts surrounded by eosinophilic and granulomatous necrosis and containing PAS-positive lamellae of chitin which was consistent with an infection by echinococcus alveolaris (fig. 4 and 5).

Post-operative serology was positive for echinococcus alveolaris and confirmed the diagnosis. The patient was placed under treatment of albendazole. A cerebral CT-scan one month after the operation did not show any secondary lesions.

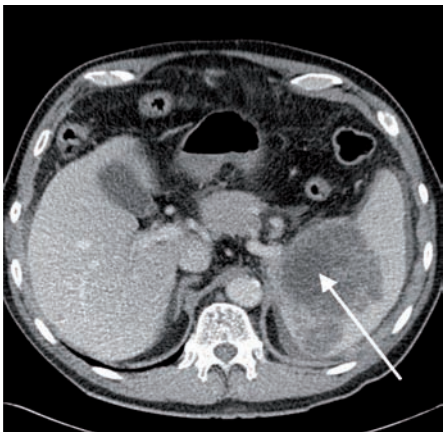

Figure 1

CT scan showing a heterogeneous splenic cyst (arrow) and a small amount of perisplenic liquid.

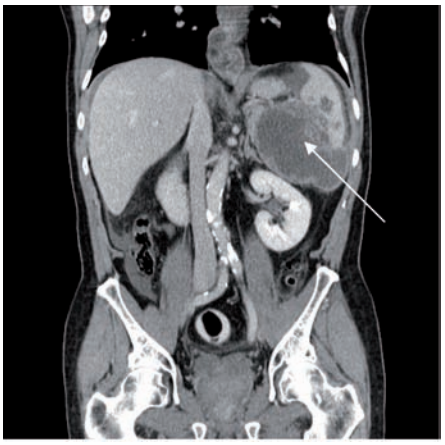

Figure 2

The same splenic lesion (arrow) as in fig. 1 in a frontal view.

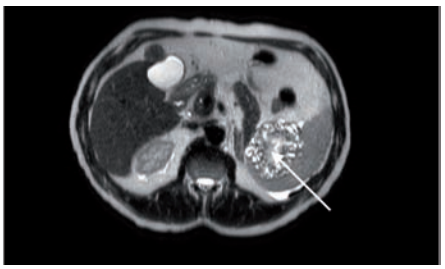

Figure 3

One year earlier a MRI scan shows a heterogeneous, plurilocular cystic lesion of the spleen (arrow).

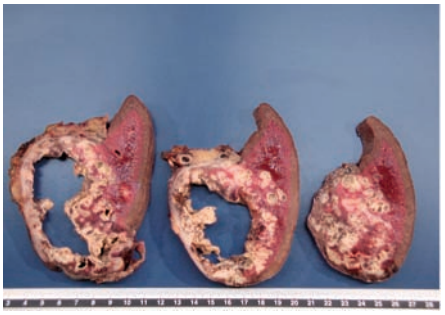

Figure 4

Macroscopic sections after splenectomy demonstrating a large central syst surrounded by multiple smaller cysts. (Photo courtesy to Dr Brigitte Christen, ADMED pathologie Neuchâtel).

Discussion

Splenic lesions are a relatively rare entity and most of them are discovered fortuitously. If the lesion is large, it may cause abdominal discomfort or pain in the left upper quadrant. Sometimes it is a complication such as rupture or infection that leads to its discovery.

Cystic lesions are far more common than solid ones. Two pathologies cover the vast majority of cases:

– Hydatid cyst makes up for 60 to 75% of all splenic cysts. It has a very typical aspect and features at ultrasound and is caused by Echinococcus granulosus.
– Post-traumatic pseudo-cysts which account for 80% of non-parasitic cysts [1].

Other, less frequent, aetiologies to consider in the differential diagnosis of a cystic lesion are congenital cysts, cystic tumours, particularly lymphangiomas and haemangiomas as well as primary tumours of the spleen and metastases with central necrosis (breast, lung, ovary, stomach, melanoma, uterus).

In most instances imaging with ultrasound supplemented by computed tomograhy or magnetic resonance will yield the diagnosis. Blood chemistry is generally of little use. In our case, of course, testing for echinococcosis would have permitted establishing the correct diagnosis much earlier proceeding with splenectomy before rupture. In fact, serological tests for echinococcosis show high sensitivity and specificity (in ranges of about 75 to 100%) and offer a good differentiation rate between E. granulosus and E. multilocularis (up to 95%) [2]. But, as hy-

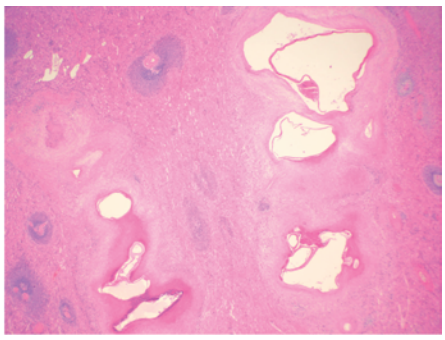

Figure 5

Microscopic aspect. Multiple cysts surrounded by eosinophilic and granulomatous necrosis and containing PAS-positive lamellae of chitin. Photo courtesy of Dr Brigitte Christen.

datid cyst could be discarded on radiological grounds and, even though a parasitic origin was considered, the consultant surgeon thought isolated splenic alveolar echinococcosis (AE) too unlikely a finding to warrant further investigation.

E. multilocularis is endemic in many countries of the northern hemisphere including parts of North America, regions of central and eastern Europe as well as most of central and northern Asia. In Switzerland the incidence of newly diagnosed cases of AE varies between 0.02 and 1.4 per 100'000 habitants per year [3].

AE is caused by the fox tapeworm Echinococcus multilocularis. The ova are shed in the faeces of the primary host, dogs and foxes, and are ingested through contaminated food by the intermediate host: sheep, cattle and mice. This can happen accidentally to humans. In the intestine the ova are transformed in oncospheres which penetrate the intestinal wall and enter the portal circulation to access the liver where they then form a small cyst that will grow and build secondary cysts. The lesion exhibit tumourlike growth and can metastasise to adjacent regions (pancreas, spleen, retroperitoneum) or to other organs (lung, CNS) in up to 20% of patients [4]. In contrast to hydatid cyst, the liver is almost invariably involved. Isolated involvement of the spleen, as in our case, or other organs has been reported in only exceptional cases, in fact only two cases have been described in recent years [5, 6].

The mainstay of treatment in alveolar echinococcosis is surgical resection. Due to the invasive growth of AE the resection has to follow oncological principles and should provide free margins of at least 1cm. In some cases, if complete resection cannot be achieved and biliary reconstruction is not possible, liver transplantation may be considered [7]. To reduce recurrence, patients are placed under treatment of benzimidazoles (Albendazol, Mebendazol) 1 to 3 months before surgery and for up to 24 months after surgery. In inoperable cases or when only palliative resection can be achieved chemotherapy should be continued throughout life because benzimidazoles only have parasitostatic action. Nevertheless, as it has been shown that FDG-PET can discriminate active from inactive lesions and attempts at discontinuation of benzimidazoles have been made. But recurrence rates are high and discontinuation cannot be recommended, in general, at this stage [8].

In summary, isolated splenic involvement in AE is exceptional but any cystic lesion of the spleen which cannot be clearly diagnosed otherwise should raise suspicion of AE and prompt echinococcal serologies (table 1).

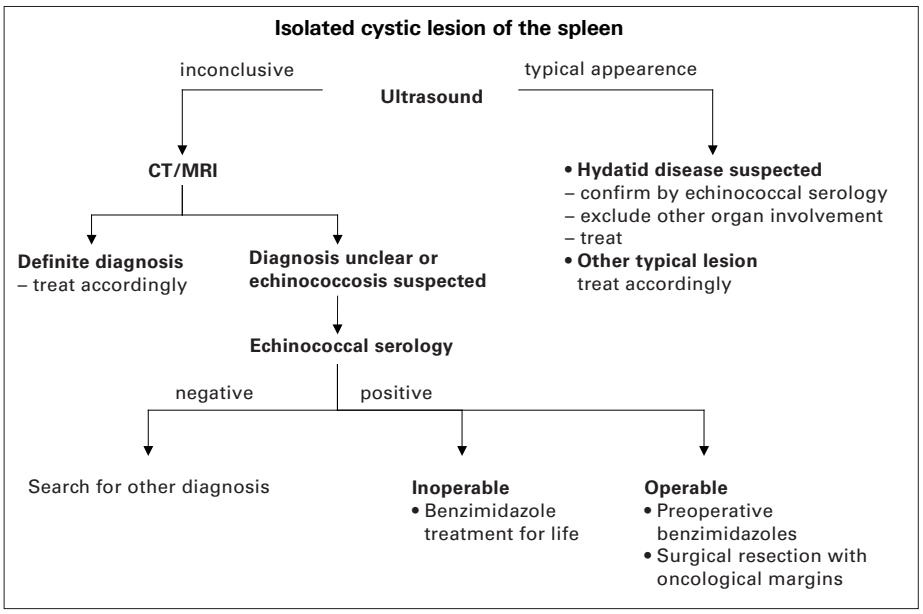

Table 1

Decision diagram for isolated splenic cysts.

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