Hydatidosis of the spleen (SH) is an extremally rare condition.

The aim of the study was to present our experience and discuss diagnostic methods and therapeutic options in cases of spleen hydatidosis.

Material and methods. Between 1993 and 2008, 8 patients were diagnosed with SH, 6 of whom were operated on (4 males and 2 females). Their average age was 44.5 years (the age range 30-59 years). Four patients presented isolated SH, one had a spleen and liver hydatidosis and there was also a case of one spleen, liver and peritoneal hydatidosis.

Results. The main symptom of the condition was abdominal pain and hydatidosis serology was always positive. The average hydatidosis cyst size was 13.3 cm (range: 7-18 cm). Splenectomy was performed in 5 cases and a partial cystectomy in one case. A left lateral sectionectomy was required in 1 case, a segment III subsegmentectomy was delivered in 1 case and multiple cystectomies in the case of the patient with disseminated hydatidosis. In the follow-up period of the above mentioned surgical procedures no mortality among the treated patients was reported. The majority of patients did not present any symptoms of morbidity (4 patients). We registered one wound infection and one cavity abscess solved with percutaneous drainage in the patient following partial cystectomy. The hospitalisation period averaged to 5 days (within the range of 5 to 12 days). The patients' follow-up was 98 months on average (range:19-190 months) without any traced relapse.

Conclusions. Total splenectomy is the treatment of choice of SH. Other surgical techniques could be employed in special cases.

Key words: hydatidosis, spleen, review

Hydatidosis is a disease caused by an infestation of Echinococcus granulosus larvae. Its incidence in Spain has declined in recent years, but a growing immigration from other ethnic group zones (Romania and Maghreb) and new endemic cases appearing have reversed this trend.

Splenic hydatidosis (SH) is uncommon, especially in its single presentation form, and, according to distinct series of data, it only occurs in between 0.5% and 8% of hydatidosis patients (1-8). As a primary cause, SH is developed when the larva crosses hepatic or lung filters and reaches the spleen, where it gives rise to a cyst. As a secondary cause, either the rupture of an intra-abdominal cyst – usually located in the liver – or its invasion on the neighboring tissues leads to this disease (1-8). We describe here our experience with 8 cases and present diagnostic methods, differential diagnosis concerning other spleen cyst lesions and current therapeutic options.

The aim of the study was to present our experience with 8 cases of splenic hydatidosis.

MATERIAL AND METHODS

Eight SH cases were assessed at our Department between 1993 and 2008. Two patients did not undergo surgery due to the decline of any procedure on the part of the patient. One of them, an 83-year-old male patient exhibited a disseminated and scarcely symptomatic hydatid process (fig.1). The other pa-
Patient, a 79-year-old male, was diagnosed with a single 5 cm SH in concomitance with an unresectable gallbladder cancer stage IV. The remaining group of patients consisted of 4 males and 2 females – their average age was 44.5 years (age range 30-59). It was decided to carry out a retrospective study of clinical, analytical and serological data, diagnostic methods, therapeutical measures undertaken, and histological data from all patients. All of them underwent abdominal ultrasonography and CT scans. Four patients exhibited splenic affection only (one case partially due to a hepatic hydatidosis previously operated). One female patient exhibited splenic and hepatic hydatidosis, and the remaining patient presented hepatic, splenic and multiple peritoneal hydatidosis. Anti-pneumococcal vaccines were administered to all patients pre-operatively. Morbidity was assessed according to the Clavien Classification during hospital stay and follow-up.

RESULTS

Five patients sought medical advice due to abdominal pain. The other case was diagnosed by an imaging test performed during a periodic control of a previous hepatic hydatidosis surgery – also in this case the patient complained of abdominal pain. Serology for hydatidosis tested positive for all patients. Blood tests showed eosinophilia and alteration of the hepatic profile only in the patient with multiple hepatic hydatidosis, while for the others it showed normal levels. Both ultrasonography and abdominal CT (fig. 2) yielded a conclusive diagnosis without exception, and detected cystic calcification in one patient only. All patients presented with a single splenic cyst except for the patient with peritoneal dissemination. The averaged size of cysts was 13.3 cm (range: 7-18 cm)

Surgery procedures comprised splenectomy in five patients, and a partial cystectomy performed on a patient exhibiting a 16-cm cyst involving diaphragm, stomach and retroperitoneum. Additionally, other procedures included a left lateral sectionectomy on the female patient with hepatosplenic hydatidosis (fig. 3), a sub-segmentectomy of segment III on the patient exhibiting an 18-cm cyst as it was too closely attached to the liver, and partial cystectomies (Lagrot) on multiple hepatic

Fig. 1. CT. Disseminated hydatidosis with splenic involvement

Fig. 2. CT. Giant splenic hydatidosis cyst. C – cyst, RV – renal vein, P – pancreas
and peritoneal cysts occurring in the patient with disseminated hydatidosis. All cysts were multivesicular, and the histological study confirmed the hydatidosis diagnosis.

Four patients did not show morbidity, although one female patient exhibited infection due to the surgical wound (Clavien I); the partial-cystectomy patient developed an abscess in the residual cavity that required percutaneous drainage (Clavien IIIa). There was no mortality in the series, and the mean hospital stay was 8 days (range: 5-12).

The follow up, which lasted 98 months on average (range: 19-190), did not report any SH relapse, except for the patient with disseminated disease who had a liver relapse and underwent a left hepatectomy.

**DISCUSSION**

Hydatidosis is a disease caused by the larva belonging to the genus Echinococcus, of which the Echinococcus granulosus is the commonest (1, 5). Although its cases can be detected in any point of the world as a result of an increased immigration and a higher incidence of intercontinental migration (3, 9), there are areas where its incidence is typically and significantly higher, and it is regarded as an endemic disease in many Mediterranean countries, the Near and the Middle East, South America, Australia, New Zealand and East Africa (3, 7, 9). Hydatidosis prevails in zones where cattle are reared close to dogs, which are the definitive hosts. Dogs excrete the eggs with the feces and humans become accidental intermediate hosts when they ingest the eggs via contaminated food (7, 10).

Embryos are released from the eggs by the action of gastric juices and reach the liver where most of them end up destroyed; although some of them develop their larval state to become a hepatic hydatid cyst. Sometimes they cross the hepatic filter and reach a secondary filter in the lungs. Those embryos crossing this last filter may eventually turn into a hydatid cyst within any body organ (10). Through this route, the most frequent localization of a hydatid cyst is the liver (60-80%), followed by the lungs (15-20%) and, to a lesser extent, serous membranes (6%), kidneys (2.9%), spleen (0.5%), multi-organ infection or any other organ (1-8, 10, 11).

There are two forms of SH. Primary SH of the spleen, whether it is isolated or affects other organs, in which case it develops through hematic route after crossing the hepatic and pulmonary filters, or retrogradely from the portal vein in patients with inverted portal flow (2, 8). Secondary SH results from larva propagation occurring at peritoneal level when an intra-abdominal cyst, usually hepatic or infiltrated from neighboring organs, breaks open (6, 9). Historically speaking, it has always been affirmed that SH occurs more frequently in combination with other cysts among organs (60-75%). But recently published data series reported isolated SH rates from 70 to 80% (1, 2, 12). As for the age of occurrence and gender distribution, values differ widely among the distinct published series, a factor that does not allow reaching conclusive results. In our study group, five patients presented a primary SH, the remaining three cases can be classified as secondary hydatidosis two as a result of larva propagation at peritoneal level and another one due to infiltration from left liver segments.

Clinical manifestations of SH are unspecific (1, 2). As the growth of hydatid cysts is slow (5 to 20 years), they are asymptomatic, and it is not rare that they can be revealed by coincidence, during the process of studying some other pathology (3). When there are symptoms present, they are due to the mass effect they provoke on adjacent organs or to the onset of complications (1, 2). Abdominal pain, splenomegaly and fever are the most common symptoms (1, 2, 4, 5, 9). As for our
patients, all of them complained of abdominal pain. SH complication comprise: infection of the cyst; rupture of the cyst and dissemination to peritoneal cavity, neighbouring organs – pleura or lungs; intra-cyst hemorrhage, anaphylactic shock and cutaneous fistula (1-4, 8-10).

SH diagnosis is achieved through the combination of imaging techniques and immunological tests (2, 5, 10). The most frequently applied tests include: hemagglutination, immunofluorescence test, and enzyme-immunoassay (EIA), and it has to be remembered that their sensitivity values fluctuate from 60 to 90% (1-3, 8, 10). Negative serologic results do not rule out SH as many cyst carriers do not develop detectable antibodies and antigens circulating in the serum of hydatidosis patients are not always detected either (10). In general, cysts involving lungs, brain and spleen are associated with low serodiagnosis reactivity (10). All out patients exhibited positive serology.

The radiological aspect of SH varies according to the cyst localization, its age and the accompanying complications (perforation, infection, etc) (5). As a rule, SH manifests as single unilocular cysts that may be calcified, although some cases describe the occurrence of various cysts (1, 7). The study of SH has always been carried through simple radiology and isotopic studies (1, 5, 8). At present, ultrasonography, CT scan, abdominal magnetic resonance imaging provide a high sensitivity and diagnostic specificity, and an exact topography of the SH, and at the same time reveals any possible affection to other organs, all of which allows to apply the most suitable treatment (4, 5, 8).

From all the above mentioned procedures, helical CT scan is the method of choice, although findings obtained through CT scans may not be thoroughly precise (1, 5). Cyst attenuation during CT scan performance will depend on intra-cyst content. Hydatid cysts tend to display a homogeneous liquid content that depicts water-type attenuation values. However, the occurrence of intra-cyst debris, hydatid sand and inflammatory cells may elevate attenuation values, in special when the CT scan is performed without contrast (5). Calcification, when occurs, can be visualised better with a contrast-free CT scan, since intravenous contrast can mask calcification as a result of an increased uptake of the surrounding spleen parenchyma. Ultrasonographic SH findings are not specific either, even in the case of unique and typical anechoic lesions (5). The mixture of membranes, scolices and hydatid sand may yield a hyperechogenic solid pattern. Nuclear magnetic resonance imaging may be useful for the differential diagnosis with other spleen cysts.

Spleen cyst lesions are divided into parasitic and non-parasitic cysts. As to endemic hydatidosis areas, this condition represents between 50% and 80% of spleen cyst lesions. It is advisable though, to differentiate between SH and other cystic lesions that share similar characteristics (7). Differential diagnosis encompasses: dermoid cyst, epidermoid cyst, epithelial cyst, splenic pseudo-cyst (a more frequent lesion of the spleen), a large single abscess, bruise, intra-splenic pancreatic pseudo-cyst, splenic lymphangioma and other cystic neoplasias involving the spleen (1, 5, 8, 9). Diagnosing SH is easier when the occurrence of daughter vesicles is detected by imaging methods or when there are other cysts in other organs such as the liver (1). Three of our patients exhibited cysts in other organs, and a fourth patient had already undergone surgery for hydatidosis, so that only four cases created a relative diagnostic doubt that serology dispelled.

Splenectomy is the treatment of choice more accepted as it is a simple technique that prevents the occurrence of residual cavities and practically rules out relapses, and above all carries a reduced morbidity-mortality rate (1, 2, 4, 7, 8). Relapse rate after splenectomy varies between 0% and 12% (12 and 13). Post-operative complication rate ranges from 0% and 30%, and mortality figures vary between 0% and 7% (2, 3, 7, 11, 12, 13). As for the splenectomy cases that we present here, morbidity involved a single case of surgical wound infection, with a relapse rate of 0%.

The possibility of using surgical techniques to preserve the spleen (partial splenectomy, cyst enucleation, partial cystectomy and omentoplasty) has been put forward (1, 2, 12). These options may be suitable especially for pediatric patients since the rate of complications and relapse is similar to that seen in splenectomy, while it rules out the risk of post-splenectomy sepsis (2, 12); it may also be suitable for patients who have suffered complications (infec-
Hidatidosis of the spleen

It should also be borne in mind that partial splenectomy techniques involve a series of disadvantages: they are more hemorrhagic and laborious procedures, and are not applicable to all cysts, only to polar or peripheral cysts (1, 7).

Also some published isolated case studies describe treatments of SH through laparoscopic splenectomy; but the risk of rupture accompanied by anaphylaxis appears to be greater (3, 14, 15), and besides, it is not applicable to big-sized cysts (7).

There are presently two options in the event of inoperability due to comorbidity -either the patient’s refusal or when cysts are too small. It is possible to carry out an albendazole-based treatment, which always renders inferior results when compared to other therapeutic options (7), or a percutaneous treatment (PAIR), which has only been conducted on a very limited number of patients exhibiting both low morbidity and relapse rates and with the prevailing advantage of preserving splenic functions (3).

CONCLUSIONS

In conclusion, splenic hydatidosis represents a rare condition that can occur isolated or associated with other hydatidosis processes affecting other organs. Abdominal CT is the best imaging diagnostic technique. Total laparotomy splenectomy is currently the treatment of choice, although laparoscopic procedures may be applicable in preselected cases. Preserving techniques are suitable for pediatric patients or in technically complicated situations.

REFERENCES


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