Review Article

Literature review- Primary splenic hydatidosis

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Abstract

Aim: Cystic lesions in Spleen are common, but a hydatid cyst is uncommon. No doubt, the occurrence of hydatid disease has decreased due to handwashing after contact with canine species, vigorous washing & cooking of vegetables before eating, and stoppage feeding the slaughtered animal’s guts to dogs. Its incidence at unusual sites is about 8-10%. These uncommon sites are Spleen, psosas muscle, pelvic cavity, peritoneum, mesentery, brain, kidneys, bones, muscles and soft tissues. Primary extrahepatic hydatid cysts are uncommon, and primary splenic hydatid cyst (SHC) is even more so. Hydatid disease of the Spleen is known since 1790 as endorsed by Berlot and is caused by Echinococcus granulosus.

Materials and Methods: Discussed below is a literature review focusing on the epidemiology, pathology, clinical presentation, investigations, treatment and follow up modalities of splenic-hydatidosis.

Results: Our literature review examines dissimilar articles published on this topic and sum up the data obtained.

Conclusion: The incidence of splenic involvement by hydatid cysts in relation to the rest of the abdominal viscera is very low and is about 0.5 to 4% of all cases of hydatidosis. Man is an accidental intermediate and end-stage host in its life cycle. Splenectomy (open and laparoscopic) was the gold standard treatment for splenic hydatidosis as medical therapy seems ineffective. However, the last two decades have shown a leaning towards splenic conservative surgery in suitable cases to reduce opportunistic post-splenectomy infection (OPSI).

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1. Introduction

Hydatid disease is a systemic zoonosis and has been known since ancient times. It has a worldwide distribution in the Middle East and Mediterranean region. Berlot described the first case of splenic Hydatid cyst in 1790. Humans are the accidental intermediate host in the development cycle of hydatid disease. Four Echinococcus species cause infection in humans; Echinococcus granulosus and Echinococcus multilocularis are the most common, causing Cystic Echinococcosis and Alveolar Echinococcosis, respectively. The two other species, E. vogeli and E. oligarchs, cause polycystic echinococcosis and are less frequently associated with human infection. Primarily it infects the liver (50-77%) and lung (18-35%) and occasionally in other organs such as Spleen (0.5-8%). A splenic hydatid cyst (SHC) is an extremely rare disease, even in endemic areas. Because of cyst’s slow growth, a latent period of five to twenty years occurs before the diagnosis of hydatid cysts is made as most of these infections are acquired in childhood. The growth rate depends upon resistance offered by affected organs and its immunologic relation with the human host. As a very rough guess, hydatid cysts increase in size by about two to three centimetres in diameter each year.

2. Materials and Methods

A search of the MEDLINE and Cochrane databases was conducted to identify reports describing splenic hydatid cyst. A total of 32 articles were chosen and assessed. Manual
Echinococcus granulosus is a part of the order Cestoda (flatworm) and Taenia family. The adult form of Echinococcus is a 5 mm long hermaphrodite that infests the small intestine of the definite host (carnivorous animals), typically dogs, foxes and coyotes. Eggs are shed in these animals’ faeces and are incidentally consumed by grazing animals such as cows, sheep, buffalo, zebras, and moose. Enzymatic digestion of these eggs’ shells in the duodenum of these intermediate hosts free the embryonic forms. These travel through the intestinal wall to enter the portal circulation, moving towards the liver or lung. These are filtered here and transformed into the microscopic larval stage, the protoscolex or scolex. These larval forms are capable of asexual multiplication within the affected organ. When the host dies or is slaughtered, the ingestion of the infected organs completes the cycle. The larval forms grow-up into adult parasites within the small intestine of the carnivore.

Man is an accidental intermediate host, as the entry of the larval forms into humans represents an end-stage in its life cycle. Contaminated vegetable consumption or meat, which are not washed or cleaned free of eggs, also exposes man to the larval forms. An alternate entry mode is direct contact with dogs whose fur has the eggs sticking onto it.

Once in man’s intestine, these embryonic forms enter the portal circulation and spread to various organs, including the liver, lungs, pancreas, Spleen, etc. About 10-15% of embryos escape from the liver and lung and filters into general circulation.

A small fraction of these escaped embryos settles down in the Spleen. The cyst may stay in the interior of the Spleen or at its periphery under the capsule. Infection is typically acquired in childhood, but they are symptoms free. The cyst grows at a rate of 0.3-1 cm per year, and it may take 5-20 years to grow into a sufficient size (3 – 35cms) to cause symptoms of continuous abdominal pain and a visible/palpable swelling in the abdomen.14,15

2.2. Incidence

Spleenic hydatid cyst occurs in 1.5-3.5% of all abdominal echinococcosis cases across all ages and both sexes.2,4–6,8–10 SHCs usually coexist with liver hydatid cysts (secondary); however, in some cases, the Spleen is the primary location.5,11 Simultaneous involvement of two organs is about 5-13% of cases.12 Hydatid disease of the Spleen is extremely rare even in endemic areas (0.5-4 per cent of all cases of echinococcosis). The occurrence of hydatid cysts of Spleen varies widely in sheep raising countries like Iran (4%), Argentina (2.4%) and Australia (2.1%). In India, the splenic hydatid cyst’s recorded prevalence is 2.5%, with the central parts’ highest incidence.15 In areas of endemic hydatid disease, most of these cysts are due to this parasite’s larval form, whereas nonparasitic cysts comprise the vast majority in Western countries.1

2.3. Pathophysiology

SHCs are classified as true cysts (primary) or pseudocysts (secondary) based on an epithelial lining presence. However, it is unclear whether primary cyst or secondary cyst is more common (with wide ranges of incidence rates being reported; primary: 38.1–80.0%, secondary: 16.0–61.9%). It is also unclear whether solitary or multiple lesions are more common (solitary: 41.1–94.7%, multiple: 5.3–33.0%). Unknown geographic, behavioural, environmental, or other many factors affect the hydatid cystic disease characteristics.16

True cysts can be additional subdivided into parasitic (caused by Echinococcus) and nonparasitic.17 Nonparasitic true cysts are congenital or neoplastic. Congenital cysts can be epidermoid, dermoid, or endodermoid, occur at a young age and are usually located in the upper pole of the Spleen. CA 19-9 and CEA levels are elevated in the epidermoid cyst’s contents and the patient’s serum. Pseudocysts are believed to develop after post-traumatic intraparenchymal or subcapsular splenic haematoma and infrequently after splenic infarcts or infections. Secondary cysts account for 75% of all nonparasitic splenic cysts.17 All over the world, over two-thirds of the splenic cysts are parasitic hydatid cysts caused by Taenia Echinococcus. A simple clinically useful classification of splenic cysts is by Martin as per Table-1.18

### Table 1:

<table>
<thead>
<tr>
<th>Martin’s Clinical Classification of Splenic cysts</th>
<th>A</th>
<th>B</th>
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</thead>
<tbody>
<tr>
<td><strong>Primary(true) cysts</strong></td>
<td>1.Parasitic</td>
<td>Secondary(False) Cysts</td>
</tr>
<tr>
<td>2.* Nonparasitic *Congenital *Neoplastic</td>
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Parasitic cyst of the Spleen is a hydatid cyst, and its occurrence is twice than of the nonparasitic variety. The most frequent sites where hydatid cysts develop are the liver (70%), which acts as a first filter and lungs (15%), which is the next filter. The other 15% are found in other organs. Hydatid cyst can occur in all organs from head to toe. Uncommon sites are Spleen, thyroid, omentum, pancreas, gall bladder, central nervous system, kidney, psoas sheet, retroperitoneal region, orbit, cervix, adductor longus muscle, leg, heart and breast. In India, the occurrence of hydatid cysts at unusual sites is higher than in other parts.
of the world and its incidence is highest in the central part of India.

Depending upon splenic hydatid cysts' location, these can be cortical, central, juxtasplenic and abdominothoracic types. Anatomically these cysts can be located anteriorly (gastro-splenic), posteriorly (pancreatico-splenic), superiorly or inferiorly of the Spleen. Depending upon mobility, these cysts are named ascending (immobile) and descending (mobile). Depending upon the number of cysts, further divided into three groups, (a) those be full of much fluid but few daughter cysts, (b) those packed with vesicles, and (c) suppurating cysts.

Splenical infestation usually occurs through an arterial route when the parasite has passed through two other filters: hepatic and pulmonary. SHCs may also occur due to retrograde spread from the liver into the Spleen via the hepatic portal and splenic veins in patients with portal hypertension. The Spleen may also be affected by the rupture of a hydatid cyst into the peritoneal cavity. Other theories about the possible spread to Spleen may direct passage from intestine to inferior vena cava via the lower and middle haemorrhoidal veins without crossing the liver, passage via the chyliferous ducts into superior vena cava and general circulation after penetration of intestinal wall and direct entry into Spleen from stomach and colon across the actual or potential spaces or via lymphatics. The most logic route is the bloodstream, and it seems that 10-15% of embryos escape from the liver and lung, filters into general circulation and a fraction of which settle down in the Spleen.

A hydatid cyst consists of three layers. The outermost layer is the adventitia (pseudocyst), made of fibrous tissue due to the liver's reaction to the cyst. It is greyish and closely blends with the liver, and cannot be separated. The laminated membrane (ectocyst) is the middle layer and is formed of the parasite itself. It is whitish in colour and elastic full of hydatid fluid. The specific gravity of fluid is 1.005 to 1.009 and it is crystal clear. It contains chlorides, sulphates, phosphates of sodium, sodium, and calcium salts of succinic acid, but no albumin. It is slightly alkaline. When not too old, it also consists of hooklets and scolices. The ectocyst grows very slowly and looks like a child's uncoloured balloon filled with water. It peels very readily from the adventitia but if bacterial infection set in then becomes inseparable. The hydatid fluid is antigenic and too toxic and can cause a fatal anaphylactic reaction in humans. The germinal epithelium (endoctyst) is innermost layer, consisting of a single layer cell lining the cyst.

The living part of the hydatid cyst secretes the hydatid fluid internally and the laminated membrane externally. Brood capsules are formed from the germinal epithelium and attached with pedicles' help to its innermost walls. The scolices develop inside the brood capsules. When the laminated membrane becomes damaged, it disintegrates and frees the brood capsules, which grow into daughter cysts.

Hydatid disease affects all age groups and both sexes with equal frequency.

2.4. Symptoms and Signs

The clinical picture of SHCs depends on location, size, relation to adjacent organs, and complications. The most frequent signs and symptoms are splenomegaly, asymptomatic abdominal mass in left hypochondrium, dragging pain, dyspepsia, constipation due to pressure on the colon, and dyspnea pushing up of the left diaphragm. The symptoms produced by splenic hydatid cyst is mainly due to mechanical displacement and pressure on neighbouring organs. About 30% of splenic cysts are asymptomatic and detected incidentally. Besides, 40% have no detectable physical signs. Known complications are cyst infection, rupture of cyst into the peritoneal or pleural cavity, fistula formation into how organs like colon or stomach; rupture of longstanding cyst into a bronchial tree; splenothoracic fistula, sympathetic pleural effusion, calcification or signs of anaphylactic shock. Anaphylactic reactions and shock due to a hydatid cyst's rupture are well known severe complications in patients with hydatid disease. Pressure effects and perforation to the adjacent organs are more frequently observed in giant hydatid cysts.

2.5. Diagnosis

The diagnosis of SHC is based on historical & geographic backgrounds; a physical examination, radiological tools, serology, fine needle aspiration cytology (FNAC), and histological examination of the excised cyst. Serological tests are useful for diagnosis, screening and follow-up to detect recurrence.

Preoperative diagnosis can be difficult in a few cases, especially if radiological findings are non-specific and serological tests are negative. Even laboratory evaluation of patients with hydatid disease often yields insufficient data. Eosinophilia (above 3%) is not important in endemic areas. A large battery of serological tests is available. These modern imaging modalities like ultrasound (USG), computed tomography (CT), and Magnetic resonance imaging (MRI) are more reliable.

Serological tests of hydatid cysts have a wide range of sensitivity and specificity, and the purity of the antigens decides it. Elevated IgE levels are a non-specific indicator of prior sensitization or active infection with parasitic organisms. In contrast, elevated IgM classes specific to echinococcal organisms may be a sensitive indicator for recurrent disease. Several serological tests are precise to hydatidosis and are done to confirm the diagnosis. Serum immuno-electrophoresis is, at present, the most reliable, with a sensitivity of approximately 90%. However, it stays positive for one year, even after the organism has been...
eradicated. Indirect haemagglutination has a sensitivity of 85%-4 but remains positive for several years, decreasing its usefulness in endemic areas. Complement fixation, enzyme-linked immunosorbent assay (ELISA) and western blot analysis are also done for diagnosis. The Casoni skin test is sensitive but not specific and remains positive for years after eradicating the organism.

The diagnosis of splenic hydatidosis is not possible with conventional radiography alone. The calcified cyst wall suggests hydatidosis. The abdomen’s radiology can reveal a soft tissue shadow with or without calcification, displacement of left diaphragm upwards, stomach to the right and transverse colon with splenic flexure downwards. Intravenous urography may show downward displacement of the kidney.

Ultrasound and Computed tomography alone or in combination, confirm the diagnosis of splenic hydatid cysts in almost all cases. Displacement of adjacent viscera and organs can be recognized on ultrasound and computed tomography. These procedures can recognize daughter cysts and hydatid sand, both of which are specific to echinococcal infestation. Ultrasound is cost-effective and is useful for follow up screening. CT is more precise than ultrasound in localizing and delineating the cyst’s extent but with radiation hazards. CT and USG cannot differentiate between non-calculated benign cysts without daughter cysts and other benign cysts of Spleen. In such cases, histopathology will confirm the parasitic nature of such splenic cyst.

The differential diagnosis for splenic hydatid cyst comprises other splenic cystic lesions such as epidermoid cysts, pseudocysts, splenic abscesses, hematomas and cystic neoplasms of the Spleen tumours of the diaphragm, stomach, colon, left kidney or tail of the pancreas.

2.6. Treatment

The PAIR (puncture, aspiration, injection, re-aspiration) procedure alternative to surgery is percutaneous drainage and administration of a sclerosing agent, such as 96% alcohol or 3% saline or 10% povidone Iodine under ultrasonography guidance. Biggest problem of PAIR is the increased chance of intraperitoneal spillage and increased recurrence chances. However, the rates of anaphylactic reactions occurring during percutaneous treatment are similar to that of open surgery. A decrease in the cyst’s dimensions, solidification of the cyst substance, and irregularity in cysts walls indicate cure with the PAIR technique.

Surgery is the gold standard treatment for both uncomplicated and complicated SHCs. The main goals of surgery are to eliminate local disease; and minimize morbidity, mortality, and recurrence rates. There is a surgical dilemma whether to do a total splenectomy or a spleen-preserving surgery for SHCs. The majority of surgeons favour total splenectomy to minimize the risk of recurrence. However, splenectomy is associated with sepsis-related deaths in 1.9% of adults and 4% of children due to OPSI. Thus, Spleen sparing surgical procedures like partial splenectomy, enucleation, de-roofing with omentoplasty, internal drainage with cystojejunal anastomosis, or external drainage have been proposed.

Recently, in the minimally invasive surgery era, laparoscopic is possible for benign and parasitic splenic cyst and is superior to open splenectomy. These days partial splenectomy for nonparasitic/parasitic splenic cysts is done laparoscopically. The anaphylactic reaction and recurrence due to spillage were the main concern with laparoscopy surgery. Despite all precautions, the incidence of scolex-rich fluid spillage during surgery is about 5%-10%, which does not necessarily lead to peritoneal spread. Recurrence after surgery was reported in up to 18% of cases, which may be due to incomplete removal, spillage, or growth of small occult cysts initially missed. So, laparoscopic splenectomy (partial or total) has become the gold standard in experienced hands. To prevent total laparoscopic splenectomy complications, partial splenectomy appears to be the best indication, but one should leave behind at least 25% of the splenic tissue with effective hemostasis.

A 10-year randomized clinical trial was carried out in Shiraz (Iran) to investigate spleen-preserving surgery’s long-term outcome versus splenectomy for splenic hydatid cyst. The study consisted of 20 patients (10 for splenectomy and 10 for Spleen preserving surgery), with a mean follow-up of 52 months. There were no differences between the two groups concerning the median hospital stay, postoperative complications and recurrence.

2.7. Medical Therapy

Medical treatment, which comprises Mebendazole (60 mg/kg/day for 6-24 months) or Albendazole (10 mg/kg/day for 6 months), in addition to surgery but cannot replace it. The primary treatment of hydatid cysts is surgical. However, the pre-and postoperative 1-month course of Albendazole and 2 weeks course of Praziquantel is essential to sterilize the cyst, decrease the chance of anaphylaxis, decrease the tension in the cyst wall (thus reducing the risk of spillage during surgery) and to reduce the recurrence rate postoperatively. Albendazole should always be administered preoperatively because the possibility of hydatidosis in other parts of the body could not be eliminated, despite all radiological diagnostic modalities. There is always the possibility of an occult leak during surgery, so medical therapy is mandatory. Mebendazole and Albendazole are benzimidazole derivative and obstruct glucose absorption mechanisms through the wall of the parasite. Albendazole is more effective than Mebendazole, probably because of superior absorption into the intestinal tract.
According to the WHO guidelines for treating hydatid disease, chemotherapy is indicated for a small cyst or for inoperable patients and those with multiple cysts scattered in many organs where surgery can be ineffective or hazardous.  

Splenic hydatid is an uncommon disease, so the definitive treatment guidelines cannot be precisely laid down. The management of splenic hydatid cyst has continued to be a dilemma for treating surgeon.

2.8. Follow Up

Patients with uncomplicated hydatid cysts should be followed with ultrasonography (US) examinations and indirect haemagglutination (IHA) tests starting 6 months postoperatively and every 1-2 years after that. Those with perforated cysts should be followed at shorter intervals.

3. Conclusion

Hydatid cyst of spleen is rare. If a splenic cyst is encountered in an endemic area, always think about hydatid disease. Sometimes preoperative diagnosis becomes challenging as radiological techniques are usually unable to add to the diagnostic accuracy. In such cases, exploration and postoperative decisions are worth practising. As larger cysts are prone to perforation, infection and fistula formation, surgery is the treatment of choice. The management of splenic hydatid cyst has continued to be a dilemma for treating surgeon as there are no definite guidelines.

4. Conflicts of Interest

All contributing authors declare no conflicts of interest.

5. Source of Funding

None.

References


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