



# Reports of Symptoms, Diagnosis and Management of Eight Cases of Primary and Isolated Splenic Hydatid Cyst

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## Abstract

**Introduction:** Primary and isolated hydatid cysts of the spleen are very rare seen even in endemic country. We report here our experience with primary and isolated splenic hydatid cysts.

**Methods:** We retrospectively analyzed the record of eight cases of primary and isolated spleen hydatid cyst that treated in Arya and Razi Hospital, Iran, Rasht.

**Results:** Among eight patients, who underwent treatment for primary and isolated splenic hydatid cyst disease, five patients were male and three patients were female. The mean average cyst diameter was 8 cm (4-15cm). Four of patients underwent splenectomy as the surgical procedure and three of patients underwent cystotomy, partial cystectomy, and omentoplasty processes as a spleen preservative surgery. One case underwent medical treatment with albendazole because the size of cyst was 4 cm. Three patients out of eight could not undergo splenectomy because in two of them the cyst was localized in the lower pole of the spleen and in another patient it was localized in upper pole of the spleen.

**Conclusion:** Primary and isolated splenic hydatid cyst is very rare. A hydatid cyst must be included in the differential diagnosis of other cystic lesions of the spleen. The best toll for diagnosis is the abdominal tomography and U&S. A splenic hydatid cyst should be treated surgically due to the high risk of a rupture, and the ideal procedure in adulthood is standard splenectomy. The outcome of treatment is good without recurrences.

**Keywords:** Isolated Splenic Hydatid Cyst, Splenectomy, Omentoplasty, Splenomegaly

## Introduction

Hydatid cyst (HC) is a zoonotic infection caused by *Echinococcus granulosus* or rarely by *E. multilocularis* [1,2]. Hydatid Cyst (HC) is a neglected tropical disease in few developing countries in the world and especially in the endemic affected areas. However, HC remains a serious public health problem in multiple Mediterranean countries [1,2]. The dog or other carnivore is the definitive host [1,2]. The intermediate host are herbivores in as (sheep, cows and camel) humans are incidentally infected [3,4]. HC is endemic in central Asia, Mediterranean regions, Middle East, Australia, New Zealand, South America [2,3]. HC involves liver in 65 to 75%, lung in 27%, kidney in 3%, and brain in 1 to 2% [1]. The disease occurs less frequently in others

organs as spleen, pancreas, heart, adrenal and muscles, soft tissue rib and mediastinum [1,5]. Bone and spleen involvement of hydatid cyst is rare and is very uncommon of Echinococcosis disease in humans [3,4,6]. Involvement of hydatid cyst in the spleen is rare, especially when the spleen is the primary and isolated organ affected and infected by *Echinococcus* disease [7-9]. This occurs in about 2% of cases of cystic echinococcosis and occurs when the parasite avoids hepatic and pulmonary filters [7-9]. The goal of standard treatment of HC is to eliminate the germinal layer, daughter cyst, scolex and fluid of the hydatid cyst [1,2]. For treatment of splenic hydatid cyst, usually three options are available: surgery, medical treatment, and percutaneous drainage (PAIR) [10,11]. Surgery was the only treatment option in the past [10,11]. Although radical excision of the cyst is recommended whenever possible, conservative surgery may be needed in a selective group of cases [8,10]. Preoperative prophylaxis with benzimidazoles is commonly advised, although there are no published data establishing its efficacy [5,8]. A percutaneous treatment known as PAIR (Puncture, Aspiration, Injection, Reaspiration) is minimally invasive and very effective treatment for hydatidosis [10,11]. As compared with surgery, PAIR is a simpler less-invasive procedure with low complication, recurrence, and mortality rates [9,12]. In our country we do not use PAIR for treatment of hydatid cysts. The aim of present case series is to show our experiences in the treatment of eight cases of splenic hydatid cyst.

## Materials and Methods

We retrospectively reviewed the case records of all patients

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with hydatid cyst which treated by our team in Razi-teaching and Arya-private hospitals in Rasht-Iran during 10 years. We find 8 cases of splenic hydatid cyst. Three patients with coexisting cysts in any other organ were excluded (liver and lung). All non-hydatid-cyst such as simple cyst, traumatic cyst, dermoid cyst and neoplastic cyst was excluded. Details of clinical findings, diagnostic methods, therapeutic method, and histological data were retrospectively analyzed for all eight patients. These patients form were the basis of this report. Abdominal computed tomography (CT) and abdominal ultrasound (US) were used as radiological diagnostic tools in all the patients. After diagnosis, seven patients underwent surgical resection of the splenic hydatid cyst, including elective splenectomy or spleen preserving surgery (cystotomy, partial cystectomy, and omentoplasty) and medical treatment, according to the number, location, and diameter of the cysts in each patient. All patients were routinely used with 20% saline solution as a scolicidal agent into the cysts during operation. Each specimen of the resected spleen and the cysts was histopathologically analyzed.

## Results

Among 8 patients who underwent operations or medical treatment for primary isolated splenic hydatid cyst, five patients were male and three patients were female. The average age was 36 years (22-58 years). Three patients complained of left upper quadrant and epigastric pain, three patients had nonspecific stomach and left flank ache, one patient was presented with pruritus and rash and one patient's cyst was found incidentally. Preoperative CBC, renal and liver function tests were within normal limits. Chest X-rays in six patients were normal, but in two patients, diaphragm was mild elevated. A hydatid cyst serology test was performed on all patients: one showed positive and seven showed negative responses. Abdominal U&S and CT were used as the radiological diagnostic tools in all patients, According to the Gharbi Classification in S&U, six of the cysts were type 2, and two of the cysts were type 1. No hydatid cyst was detected in another organ in eight cases by radiological scanning. None of the patients were operated for hydatid cyst in any organs previously. All patients were administered 10 mg/kg albendazole for prophylaxis seven days in the preoperative period. Seven patients underwent laparotomy. The average spleen size was 12 cm (8-18 cm) in length. Four patients had moderate splenomegaly. As observed in Figure 3, in one case the cyst was huge. The average cyst diameter was 10 cm (4-18 cm). Only one cyst was found in each case. Three patients had a cyst in the upper pole of the spleen, three patients had a cyst in the lower pole of the spleen, two patients had a cyst localized in the central part of the spleen and one patient had a huge cyst involved 80% part of the spleen (Figures 1-3). Four patients underwent splenectomy as the surgical procedure (Figures 4,5). Three patients underwent cystotomy, partial cystectomy, and omentoplasty as a spleen protective surgery which are shown in Figure 5 and Figure 6. The average postoperative hospitalization period was 4, 8 days (3-9 days). All seven patients who underwent elective splenic surgery received pneumococcal and meningococcal vaccines two weeks before the surgery to allow the development of protective antibodies. In this series there was no morbidity and mortality.



Figure 1 Show CT Scan of abdomen and a cyst in the upper pole of spleen.



Figure 2 Show CT Scan of abdomen and a cyst in the lower pole of spleen.

In 30 days, postoperative CBC and platelet level were in normal ranges. We prescribed 10mg/kg albendazole for three 28 days with 14 days interval in the postoperative period [1,2].

## Discussion

Hydatid Cyst (HC) caused by the larva belonging to the genus *Echinococcus*, with *E. granulosus* being the most commonly found [6]. Hydatid cyst has a wide prevalence in the world [6]. HC is a neglected tropical disease in few developing countries across world and especially in the far affected areas [13]. However, HC remains a considerable public health problem in several Mediterranean countries [5,6,13]. The dog or other carnivore is the definitive host The intermediate host in general are sheep, cows and camel ,humans are incidentally infected [3,4,5,13]. HC is endemic in central Asia, the Middle East, Australia, New Zealand, South America, and the Mediterranean regions and Iran [5]. HC involves 65 to 75% in the liver, 27% in the lung, 3% in the kidney, and 1 to 2% in the brain [1,6]. The disease occurs less frequently in the spleen, pancreas, heart, adrenal and muscles, soft tissue, mediastinum and rib [2,6]. HC of bone involvement is rare [2-5]. Although compatible with long term survival, the disease is



**Figure 3** Show CT Scan of abdomen and a cyst in the 80% of spleen.

not easy to eradicate and perhaps impossible to cure [2,5]. The disease is frequently encountered in Iran [1,2,5]. The degree of prevalence of the infection depends on the multitude of herds raised in that region [7,13]. Although hydatid cyst can be found in almost all organs and tissues of the human body [1,2,13], isolated splenic hydatid cysts constituted 5.6 % in some report [13]. The parasite can directly reach the spleen because the portal blood flow turns in the opposite direction in human beings with portal hypertension. Another possibility is that the parasite reaches the spleen by means of the lymphatic or blood flow, and a third is that it reaches the spleen as a result of reflux from the portal vein due to the increase in intra-abdominal pressure [13,14]. Clinically, nearly 30% of the patients with splenic hydatid cysts are asymptomatic [13]. Splenomegaly is the most frequent finding, which is incidentally determined [13,15]. The clinical symptoms caused by splenic hydatid cyst mostly depend on the pressure effect of the cyst on the neighboring organs and the

replacement of the neighboring organs [13]. The symptoms are few and non-specific which comprises mainly an abdominal mass that mostly located in the left hypochondrium and less frequently in the epigastrium, and a pain that is usually a dull, dragging ache, dyspepsia, constipation due to pressure on the colon, and dyspnea due to pushing up of the left diaphragm [1,2,8,10]. A pain in the lumbar region constitutes a clinical sign in a few patients [16]. Hypertension induced by renal artery pressure is another clinical symptom that occurs due to the pressure of the cyst on the neighboring organs [15,17]. In our study group, two patients had stomach ache and two had left upper quadrant pain, rash and pruritus, and one case had dyspeptic complaints. U&S and CT scans, alone or in combination, can help a definite diagnosis of splenic hydatid cysts in almost all cases [13]. U&S is the primarily preferred method because it is inexpensive, easy, and available and also has a high diagnostic value [13]. It is diagnostic because it shows the cystic structure of the lesion, the presence

of daughter vesicles, and hydatid sand [18]. CT-scan is usually the next step after an U&S diagnosis has been made [13,18]. The main purpose is to visualize the relation between the hydatid cyst and the surrounding tissue. Although CT scan is more sensitive than abdominal U&S [19], in our study, abdomen CT-scan and U&S was used in all eight cases. The primary nonparasitic cysts, pseudo cysts of the spleen, splenic abscesses, cystic neoplasia, and traumatic spleen cysts should be kept in mind in the differential diagnosis [20]. Several serological tests are specific to hydatidosis and are used to confirm the diagnosis, but we don't use routinely these tests at our clinic [13,14]. Özdoğan and Coworker emphasized that serological tests were not necessary for diagnosing hydatid cyst disease [18].

The treatment of splenic hydatid cysts is made conservatively or surgically. Small and asymptomatic splenic hydatid cysts require close follow-up, although they can be treated by anthelmintic medications [21]. Surgical operations vary from aspiration to total splenectomy [22]. No exact consensus has been reached regarding the selection of an optimal surgical procedure [13]. Symptomatic or large cysts should be treated surgically because they can rupture spontaneously or traumatically [22].



**Figure 4** Show Spleen, Postsplenectomy and hydatid cyst after open the cyst.



**Figure 5** Show laminated membrane after evacuations.



**Figure 6** Show freeing of omentum for filling of cavity with omentoplasty.

In one of our cases, the cyst was minor rupture and the patient was presented with rash and pruritus. Total splenectomy is preferred by most of the surgeons because of its very low or no recurrence rate [23]. It is the preferred approach undertaken in cases with larger, multiple, and symptomatic cysts of central or Hilary location or in cases with simultaneous involvement of other organs [23]. However, sepsis-associated mortality rates of total splenectomy are 4% in children and 1.9% in adults and this is the greatest disadvantage of total splenectomy [13]. For this reason, the number of surgeons in favor of spleen-saving surgery has increased [13]. Spleen-saving approaches are preferred for small and single cysts that are selected in the periphery of the spleen [13]. Spleen-saving approaches include partial splenectomy, enucleating, unroofing with omentoplasty, internal drainage with cystojejunostomy, or external drainage [24,25]. In our study, four underwent splenectomy as the surgical procedure and three patients underwent cystotomy, partial cystectomy, and omentoplasty as a spleen-saving approach. No difference was found between total splenectomy and spleen saving approaches in terms of the recurrence rate of the splenic hydatid cyst disease, postoperative hospitalization period, and complications [24]. Surgical treatment can also be made by laparoscopic or robotic methods [21,26,27]. However, the cyst can be torn during surgical treatments made by laparoscopic or robotic methods and this can result in anaphylactic shock, intra-abdominal dispersion, and recurrence of the cyst [13]. Clinical experience and costs are also included among the significant problems [20,27,28]. If the splenic hydatid cyst is torn during the surgery, either spontaneously or traumatically, anaphylactic shock is a rare but severe condition that can occur [13]. The patient can die if anaphylactic shock is not diagnosed, not immediately treated, or is resistant to treatment [29]. We did not observe any surgical operation-induced anaphylactic reactions in our patients.



## Conclusion

Primary and isolated splenic hydatid cysts are very rare. A hydatid cyst must be included in the differential diagnosis of others cystic lesions of the spleen. The best toll for diagnosis is the abdominal tomography and U&S. A splenic hydatid cyst should be treated surgically or medically due to the high risk of a rupture, and the ideal procedure in adulthood is standard splenectomy. The outcome of treatment is good without recurrences. Although laparoscopic or robotic methods can be used in selected patients, the most prevalent treatment method is total splenectomy by open surgical methods. More spleen protective methods should be preferred, particularly in children.

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