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Isolated primary splenic hydatidosis: Series of four cases radiological and pathological evaluation

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Abstract

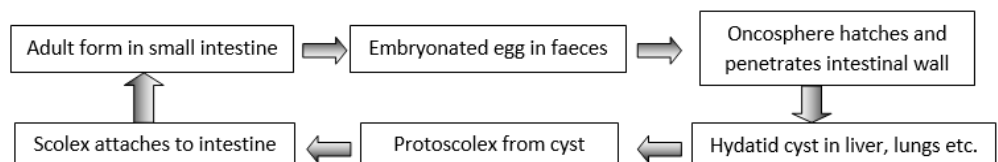
Primary extrahepatic hydatid cysts are rare, and primary splenic hydatid cysts are even more rare, constituting 2% to 4% of all hydatid cysts [1, 2]. Liver is the commonest site of involvement by the hydatid disease followed by lungs. Other rare sites include kidney, spleen, bone, thyroid, breast, pancreas, heart and muscles. The larval form of the genus *Echinococcus granulosus* most commonly causes hydatid disease. The infestation of the spleen usually occurs through arterial route after the parasite has passed through hepatic and pulmonary filters. The retrograde venous route is also considered. In cases of cystic splenic lesions, hydatid disease should be always be considered in the differential diagnosis.

Keywords: Isolated primary hydatidosis, extrahepatic hydatid, hydatid cysts, hydatid disease, echinococcus, splenectomy, rare hydatid

Introduction

Hydatid cysts of spleen may occur as a part of disseminated disease or may be isolated. Hydatid disease involving only the spleen, in the absence of disease in other organs is known as Isolated primary splenic hydatidosis. In India, the recorded prevalence of the splenic hydatid cyst is 2.5%, more prevailing in the central parts of the country [3]. Berlot first described splenic hydatid cyst as an autopsy finding in 1790 [4]. The diagnosis depends on a careful history along with radiological and serological investigations. Humans are the accidental intermediate host in the development cycle of hydatid disease. The growth rate of hydatid cysts likely depends upon the immunologic relationship between the parasites and humans and also on the resistance offered by the enveloping and surrounding structures. Secondary infection, fistulization to adjacent organs and rupture into the peritoneal cavity are some of the complications of hydatid disease. Traumatic or spontaneous rupture of a hydatid cyst may cause a life-threatening complication (systemic anaphylactic reaction). In general, the cystic lesions of the spleen are rare and usually include simple (benign) cysts, parasitic cysts or neoplastic cystic lesions such as lymphangiomas, cavernous hemangiomas or dermoid cysts, and non-neoplastic cysts (pseudocysts) secondarily due to hemorrhage or infarction [5-8]. Among parasitic cysts, hydatid cysts are the most common. Splenic hydatid cysts are usually asymptomatic and they are diagnosed incidentally while evaluating such patients for other reasons (as was with one of our patients). When the cyst attains a considerable size the patient becomes symptomatic and mostly presents with painful left upper abdominal mass [9, 10].

Life cycle of echinococcus: *Echinococcus granulosus*, also commonly known as the hydatid worm or dog tapeworm or hyper tapeworm is cyclophyllid cestode that parasitizes the small intestine of canids as an adult, and has important intermediate hosts such as livestock and humans, where it causes cystic echinococcosis, also known as hydatid disease.



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Definitive host (dogs and other carnivores) = ingestion of cysts (in organs).

Intermediate host (sheep, goat, swine etc) = ingestion of eggs (in faeces).

Four species of echinococcus 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.oligarthus* cause polycystic echinococcosis and are less frequently associated with human infection. Primary infestation of the spleen by the parasite is a rare phenomenon.

Discussion

Classic hydatid cysts are caused by echinococcus granulosus. These grow very slowly, at the rate of 1–2 cm per year. Splenic hydatid cysts occur at all ages and in both sexes. Diagnosis is mainly based on serological investigations, abdominal ultrasound and computed tomography. Treatment varies from spleen preservation to splenectomy. Approximately 25-30% of splenic cysts are asymptomatic. The symptoms are usually non-specific [11], and include an abdominal lump, dull dragging ache, dyspepsia, and dyspnea due to pushing up of the left hemidiaphragm. Some patients may present with complications such as infection of the cyst, rupture of the cyst into the peritoneal cavity, fistula formation into hollow viscera, bronchopleural fistula, sympathetic pleural effusion, calcification, hypersplenism, and severe anaphylactic reaction. Herein, we report our experience with isolated primary splenic hydatid cysts in four adult patients.

Primary splenic involvement by hydatid cyst is rare. The cyst embryos are captured in the liver and/or the lungs after ingestion and hence do not reach the systemic circulation to infect the spleen. Secondary splenic involvement may occur as a result of rupture of a hepatic hydatid cyst with

abdominal and pelvic dissemination. Bones may be involved in 0.5% - 4% of the cases. Renal involvement, primarily of the cortex, may be seen in 2-3% of the cases. Hydatidosis of the brain constitutes 2% of all intra-cranial space-occupying lesions in endemic regions. Other rare sites that may be involved include heart (most commonly the left ventricle) [12], soft-tissues, breast, ovaries, pancreas, scrotum, inguinal canal and the chest wall.

Radiology and serology play a very vital role in the diagnosis of splenic hydatidosis. Ultrasound is helpful in detecting the daughter cysts, cystic membranes, septa and hydatid sand [13]. Snake or Serpent sign refers to the undulating pathognomonic membranes within the cysts that can be seen on ultrasonography and CT. Advancing stages of collapse show the twirled and twisted membranes (spin or whirl sign). The presence of mural calcification and/or daughter cysts helps to rule out other cystic lesions of the spleen such as epidermoid cyst, abscesses, haematoma, haemangiomas, pseudocysts, metastases and cystic neoplasms. Imaging when combined with serological tests like enzyme linked immunosorbent assay (ELISA), indirect haemagglutination and immuno-electrophoresis can lead to a successful diagnosis of splenic hydatid in 90% of cases.

Case discussion: On clinical examination, the vital signs were normal in all our four patients. Chest, cardiovascular, central nervous system and the musculoskeletal system were unremarkable. Routine laboratory investigation complete blood count, coagulation profile, biochemistry, renal function test, liver function test and electrolytes revealed no abnormalities. ESR was normal at the end of hour. Plain chest radiographic appearance revealed mildly elevated left hemi-diaphragm, left hypochondrial soft-tissue mass without calcification, or the displacement of stomach and/or left colonic flexure.

Table 1: Clinical and routine investigation details of our patients with isolated primary splenic hydatid cyst

Age (yrs)/Sex	Signs, symptoms and clinical examination	Hb (g/dl)	Leucocytes count (per cumm)	Vitals	Liver and Renal function tests
51 / M	Asymptomatic	12.4	8, 900	stable	Normal
65 / M	Abdominal discomfort, dull dragging pain, palpable spleen	10.2	11, 200	stable	Normal
45 / F	Mild pain in the left upper abdomen	10.5	10, 300	stable	Normal
32 / M	Asymptomatic	14.2	9, 500	stable	Normal

Abdominal ultrasonography of the abdomen was done in all the four patients that revealed splenomegaly with well defined variable sized cystic lesions within the splenic parenchyma. Splenic hydatid cyst appeared as a solitary unilocular well defined anechoic nearly spherical to oval shaped cystic lesions in our patients with multiple internal smaller cysts. In one of our patients, the cyst was exophytic in nature. No other remarkable abnormalities could be detected in our patients except for Grade II benign prostatic hypertrophy in one elderly patient and intramural uterine fibroid in the female patient.

Contrast enhanced computed tomography scans of the abdomen, thorax and head were performed in all our patients after ruling out all the possible relative and absolute contraindications. Abdominal CECT images demonstrated confirmed well defined smoothly marginated circular to

oval shaped cystic lesions within the spleen with thin imperceptible wall with multiple daughter cysts having attenuation value near to that of water without enhancement after intravenous contrast administration and with no surrounding abnormality. All the other abdominal and pelvic organs were unremarkable (except for the benign prostatic hypertrophy and degenerative changes of thoracolumbar spine in the elderly male; And intramural uterine fibroid in the female patient). Mild displacement of the adjacent organs and bowel loops was seen in our patients in whom the spleen was significantly enlarged. There were no cysts in any other abdominal viscera. CT scans of the chest did not show any pulmonary or extrapulmonary cystic lesions. CT is more sensitive than ultrasonography in depicting subtle wall calcification. Wall calcification was seen in only one of our patients.

Table 2: Relevant splenic findings on abdominal scans were as follows

	Spleen size	Size of cyst
1 st patient	14.7 x 9.3 cm	72 mm (antero-posterior) x 68 mm (transverse) x 70 mm (craniocaudal)
2 nd patient	13.1 x 7.1cm	18 mm (antero-posterior) x 17 mm (transverse) x 15 mm (craniocaudal)
3 rd patient	18.4 x 10.3 cms	151 mm (antero-posterior) x 172 mm (transverse) x 90 mm (craniocaudal)
4 th patient	15.3 x 8.8 cms	120 mm (antero-posterior) x 106 mm (transverse) x 91 mm (craniocaudal)

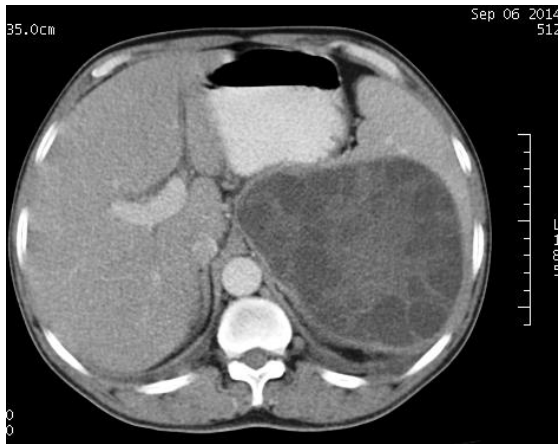


Fig 1



Fig 2

Fig 1: CECT abdomen showing a well defined cystic lesion involving major portion of splenic parenchyma with multiple internal daughter cysts.

Fig 2: Ultrasound abdomen demonstrates a solitary well defined spherical cystic lesion in the spleen.



Fig 3



Fig 4

Fig 3-4: Axial and sagittal images of CECT abdomen showing wall calcification of the splenic cyst.

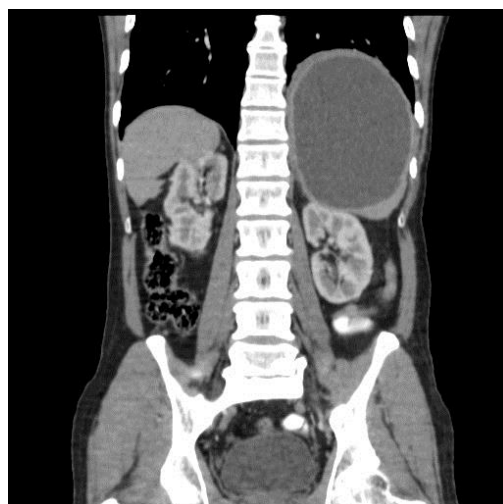


Fig 5

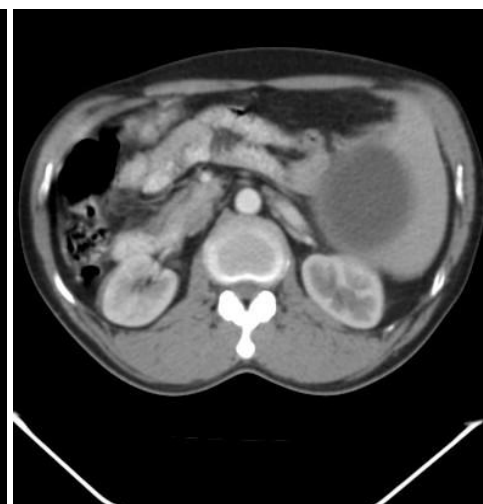


Fig 6

Fig 5-6: Coronal and Axial sections of CECT abdomen demonstrating thin membranous densities within the splenic cysts.

The intradermal (Casoni) test is a valuable diagnostic procedure. It is sensitive but not specific. The haematological and serological parameters were normal in our patients. Routine laboratory investigation CBC, coagulation profile, biochemistry, renal function test, liver function test and electrolytes revealed no abnormalities. ESR was normal at the end of hour. Serological tests are highly sensitive and specific for echinococcosis. More recently, immuno electrophoresis has improved diagnostic accuracy in up to 95% of cases.

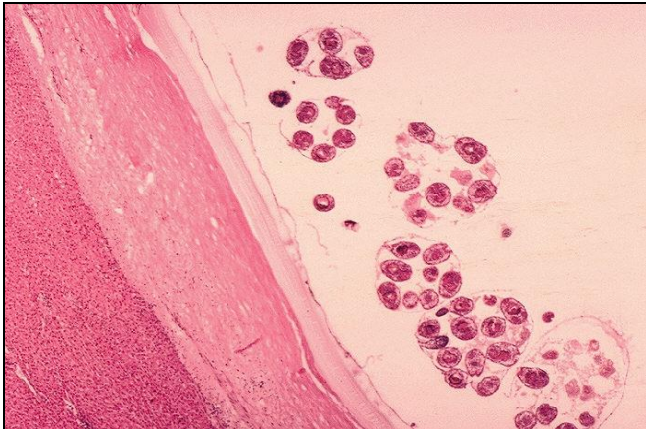


Fig 7: Microphotograph of hydatid cyst

The hydatid cyst consists of three layers : an adventitia made of host tissue, a middle layer of friable ectocyst and an inner germinal layer from which large number of scolices are produced. Daughter cysts are formed if the viability is threatened. Various serological tests are usually carried out for the diagnosis, screening and post-operative follow-up for recurrence. These include the hydatid immunoelectrophoresis, enzyme linked immunoabsorbent assay (ELISA), latex agglutination and indirect haemagglutination test.

The differential diagnosis of splenic hydatidosis includes cystic lesions of adjacent organs, such as pancreas, liver, omentum, intrasplenic aneurysm and benign and malignant splenic tumors. The cystic lesions of the spleen are rare and usually include simple (benign) cysts, parasitic cysts or neoplastic cystic lesions such as lymphangiomas, cavernous hemangiomas or dermoid cysts, and non-neoplastic cysts (pseudocysts) secondarily due to hemorrhage or infarction.

The surgical approach is still accepted as the standard for managing hydatid disease keeping in mind the limited efficacy of drug therapy and owing to the risk of spontaneous or traumatic rupture. Total or partial splenectomy, cyst enucleation and unroofing with omentoplasty are the various preferred surgical techniques to treat splenic hydatid disease. Pre- and postoperative administration of albendazole used to sterilize the cyst, reduce the risk of anaphylaxis, decrease the tension in the cyst wall and to reduce the postoperative recurrence rate. Laparoscopic approach for uncomplicated hydatid cyst of the liver and spleen has been a safe and effective option.

Conclusion

We conclude that although rare, splenic hydatidosis should always be kept in mind while dealing with cystic lesions of the spleen especially in the geographical regions where the disease is endemic. Hydatid disease should be considered in the differential diagnosis of all cystic masses in the spleen.

Computerised tomography scan is the most sensitive investigation for diagnosis. Surgical resection is the best curative procedure.

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