Primary Hydatid Disease of Spleen: - A Case Report

Authors
Dr Amit Das\textsuperscript{1}, Dr A S Kalra\textsuperscript{2}
\textsuperscript{1}Pathologist, Indian Armed Forces Medical Services
\textsuperscript{2}Surgeon, Indian Armed Forces Medical Services

Abstract
Primary hydatid disease of the spleen alone is very rare, even though spleen is the third most common organ involved in hydatid disease. The rarity of splenic hydatid disease poses a diagnostic challenge for clinicians, particularly in non-endemic areas. Hydatid cyst can present as a simple cyst without having the classic serological and imaging features, and later can lead to life-threatening complications like anaphylaxis. Hydatid disease of spleen should be considered in differential in every patient in endemic areas with cystic lesion of spleen. Hydatid disease, a zoonosis, occurs worldwide but it is endemic in farming and cattle rearing areas. We report a case of histologically confirmed primary hydatid cyst of spleen in a 34 years old male. This case report emphasises that hydatid disease should be considered in the differential diagnoses of all cystic masses in the spleen/abdomen.

Keywords: Hydatid disease, Spleen, Echinococcus.

Introduction
Hydatid disease also known as Hydatidosis and echinococcosis has been recognised since ancient times and has worldwide distribution. It is parasitic infection caused by two species of Echinococcosis namely Echinococcus granulosus & Echinococcus multilocularis\textsuperscript{(1)} Berlot in 1790 accredited with the first description of a splenic hydatid cyst as an autopsy finding\textsuperscript{(2)}. It may be detected incidentally or present with non-specific complaints. It is endemic in cattle-rearing areas of South America, Africa, Middle East, South Europe, India, and Australia. Primary Hydatid disease spleen is extremely rare even in endemic areas (0.5-4\%)\textsuperscript{(3)}. Spleen is the third commonest site for Echinococcus.\textsuperscript{(4)} The most commonly involved organ is the liver (75\%), followed by the lung (15.4\%), and the spleen (5.1\%).\textsuperscript{(4)}

In this article, we report a case of an isolated giant splenic hydatid cyst. The patient underwent splenectomy. The diagnosis of hydatid cyst was confirmed by histopathology. This case suggests that hydatid cyst spleen should be considered as a differential diagnosis in every patient with a cystic mass in left hypochondrium.

Case Report
A 34 years old patient presented to the Surgeon with the complaint of dull aching pain in left hypochondrium for last 1 month. He had no history of fever, wt loss, jaundice, cough, respiratory distress or GI bleed and his past medical history was unremarkable. On examination, vital parameters were normal. Physical examination revealed a large firm, non-tender intra-abdominal mass in left hypochon-
Dr Amit Das et al Volume 05 Issue 09 September 2017

Routine laboratory investigations viz. complete blood counts, coagulation profile, renal function tests, liver function tests & serum electrolytes revealed no abnormalities. ESR was 42mm/ 1st hr (Westergren). Abdominal Ultrasonography showed a round, well defined, cystic lesion of approximately 200x100 mm over pancreas suggesting pseudo pancreatic cyst arising from tail of pancreas. Abdominal Computed Tomography (CT) scan revealed a large homogenous cystic lesion in spleen involving full length of splenic parenchyma measuring 20.5x9x8 cm with an imperceptible wall (Fig 1). All other abdominal and pelvic organs were unremarkable. Patient was taken up for exploratory laparotomy. Intraoperative finding showed a very large splenic cyst with clear fluid & membrane. With due precaution to avoid spillage splenectomy with cyst removal was performed and the specimen was sent for histopathological examination. On gross examination, the spleen along with cyst measured 21x10x8cm and weighed 750gm (Fig. 2). Spleen was replaced by a glistening cyst (fig.3) leaving only minimal original spleen. Post-operative recovery was uneventful. Patient was treated with Albendazole tablet 400 mg twice a day for three months with pneumococcal vaccine. No complication was found during three monthly reviews. Histopathological examination showed scattered brood capsules with hooklets along with the cyst wall having an outer acellular laminated layer and inner germinal layer characteristic of hydatid cyst (Fig.4)

Discussion
Parasitic cysts of the spleen are almost exclusively hydatid cysts. In endemic areas, 50-80% of splenic cysts are echinococcal (4). Four species of Echinococcus causes infection in humans. E. granulosus and E. multilocularis are the most
common, causing cystic echinococcosis and alveolar echinococcosis respectively. The two other species; E. Vogeli and E. Oligarthrus cause polycystic echinococcosis and are less frequently associated with human infection\(^5\). The life cycle of Echinococcus includes a definitive host (usually dogs or related species) and an intermediate host (such as sheep, goats, or swine). Humans are incidental hosts; they do not play a role in the transmission cycle\(^4\). Consumption of contaminated vegetables or meat exposes human to larval form. The eggs hatch and the resultant oncospheres penetrate the intestinal mucosa of humans and enter the portal circulation. They primarily lodge either in the liver, lungs or kidneys which act as filters. Oncospheres slowly develop a cavity lined with germinal epithelium, outside which develops a laminated a cellular area\(^5\). Primary infestation of the spleen usually takes place by the arterial route. A retrograde venous route through portal circulation which bypasses the lungs and liver is also reported. Hydatid cysts are usually grow at a rate of 2 to 3 cm per year\(^6\). The main symptoms associated with the disease are abdominal discomfort, pain and palpable mass in left hypochondrium. Our patient presented with abdominal discomfort & abdominal mass. The complications of splenic hydatid cyst are mainly secondary infection, inflammation, anaphylactic shock, acute abdomen, compression of other viscera or fistulisation to the bowel, mainly colon. The main problem in the diagnosis of splenic hydatidosis is in differentiating it from other splenic cystic lesions. The differential diagnosis of such lesions includes epidermoid cyst, pseudo cyst, large solitary abscess or hematoma, pancreatic pseudo cyst and cystic neoplasms of the spleen\(^7\). CT scan is the most valuable imaging techniques in the diagnosis and evaluation of focal splenic diseases\(^7\). CT has higher overall sensitivity than ultrasound, with sensitivity rates of 95-100%. Serological tests such as ELISA, immunoelectrophoresis or indirect hemagglutination test, combined with imaging, can diagnose splenic hydatid disease in 90% of the cases\(^6\). In our case, imaging studies were suggestive of splenic hydatid disease but the diagnosis was confirmed only after histopathological examination. On histopathology, hydatid cyst consists of three layers. The outer most adventitia, the middle laminated ectocyst and an innermost endocyst from which large number of scolices are produced\(^8\).

Total splenectomy, partial splenectomy, cyst enucleation and unroofing with omentoplasty are the various surgical techniques to treat splenic hydatid disease\(^9\). During surgical treatment extreme caution must be taken to avoid life threatening complications like anaphylactic shock due to spillage of cyst contents. Antihelminthic drug therapy (Albendazole) was given in post-operative period with pneumococcal vaccine. No post splenectomy infection was encountered.

**Conclusion**

Hydatid disease should be considered in the differential diagnosis of all patients presenting with mass over left hypochondrium. Hydatid disease is still a major cause of cystic lesions of spleen in endemic area & causes diagnostic dilemma. Proper evaluation &confirmation of diagnosis is necessary before surgery to avoid further complications.

**References**

1. Koneman’s Textbook of Diagnostic Microbiology. 6\(^th\) ed. Lippincott Williams & Wilkins 2006;
4. Rasheed K, Zargar SA, Telvani AA. Hydatid cyst of spleen: A Diagnostic


