Primary Hydatid Cyst of Spleen: A Rare Entity
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Abstract
Hydatid disease, a zoonosis, occurs worldwide but its prevalence is high in those countries where sheep and cattle raising constitute an important industry due to close association between man, sheep and dog. Most common sites of involvement by hydatid disease are liver followed by lung. Splenic involvement by hydatid disease is very rare accounting for only 0.9% to 8.0% of all cases. Other rare sites include heart, pancreas and muscles. We report a case of histopathologically confirmed primary splenic hydatid cyst in an 18 year old male, which is a rare entity. Hydatid disease should be considered in the differential diagnosis of cystic lesions of spleen.

Keywords: Hydatid disease; Spleen

Introduction
Splenic involvement in hydatid disease is uncommon, representing less than 2% of all human infestations by Echinococcus [1]. It is an endemic disease in the sheep and cattle raising countries Middle East, North Africa, New Zealand, Australia, and South America [2]. In India, the recorded prevalence of the splenic hydatid cyst is 2.5%, with the highest incidence reported in the central parts [3]. In areas of endemic hydatid disease, most of these cysts are due to the larval form of this parasite, whereas non-parasitic cysts comprise the vast majority in Western countries [4]. The first case of a splenic hydatid cyst was reported by Berlot in 1790 from an autopsy [5]. Herein, we report a case of splenic hydatid disease in a 18 year old male from Haryana (India). We report this case because of its rarity.

Case Report
An 18 year old male presented to surgery outpatient department with complaint of pain in left hypochondrium for one month. Patient was asymptomatic otherwise. Clinical examination revealed mild splenomegaly. Patient’s haematological and serological parameters were within normal limits. Contrast Enhanced CT scan (CECT) revealed a large cystic lesion in distal portion of spleen with imperceptible wall of size approximately 9.8x10x8.4 cm. All other abdominal and pelvic organs were unremarkable on ultrasonography. Partial splenectomy with cyst removal was performed. Histopathological examination showed brood capsules with hooklets along with the cyst wall having an outer acellular laminated layer and inner germinal layer characteristic of a hydatid cyst (Figure 1-3). A final diagnosis of splenic hydatid cyst was made. After one week patient was discharged with instructions to continue

Figure 1: H & E section showing hydatid cyst along with portion of spleen (100X).

Figure 2: H & E section showing hydatid cyst along with portion of spleen (200X).

Figure 3: H & E section showing laminated cyst wall along with scolices (400X).

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Discussion

Hydatid disease (Echinococcosis) is a zoonotic infection caused by the larval form of parasites of tapeworm, Echinococcus granulosus.

Humans are the accidental intermediate host in the development cycle of hydatid disease [2]. 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. oligarthrus cause polycystic echinococcosis and are less frequently associated with human infection [6].

Although hydatid disease affects any organ or soft tissue, it most frequently found in liver (60–70%), lungs (30%), and rarely encountered in the kidney, spleen, bone, thyroid, breast and pancreas [2]. Parasitic cysts of the spleen are almost exclusively hydatid cysts. In endemic areas, 50-80% of splenic cysts are echinococcal [7]. Splenic echinococcal cysts may be primary or secondary to ruptured liver cysts [8]. Possible routes of primary hydatid of spleen include arterial route after passing through liver (first filter) and lung (second filter). Another route is the venous route through portal circulation bypassing liver and lung. Secondary hydatid spleen usually follows systemic dissemination or intraperitoneal spread following ruptured hepatic hydatid cyst [9].

Hydatid cysts are usually slow growing and in some organs, especially in the spleen, they can reach an enormous size and be asymptomatic [1]. Splenic cysts present with local or referred pain (often postural) or signs and symptoms relating to splenomegaly, abdominal distension and compression of nearby structures [4]. However, secondary infection, cyst rupture and anaphylactic shock may develop [1].

The differential diagnosis includes cystic lesions of adjacent organs, e.g. Pancreas, liver and omentum, intrasplenic aneurysm and benign and malignant splenic tumors [4].

At present sonography and CT, are the most valuable imaging techniques for the diagnosis and evaluation of focal splenic diseases [1] the intradermal (Casoni) test is a valuable diagnostic procedure [9]. Serological tests are highly sensitive and specific for Echinococcosis [2]. More recently, immuno electrophoresis has improved diagnostic accuracy in up to 95% of cases [8]. In our case, imaging studies were inconclusive and histopathological examination confirmed the diagnosis. On histopathology, the hydatid cyst consists of three layers. The outermost adventitia (pseudo cyst) is formed of compressed splenic tissue, a middle layer laminated membrane of friable ectocyst and an innermost germinal layer, endocyst [2].

Total splenectomy, partial splenectomy, cyst enucleation and unroofing with omentoplasty are the various preferred surgical techniques to treat splenic hydatid disease [4]. In our case, partial splenectomy was preferred as the cyst was involving the distal portion and also patient was a young male. However, during surgical treatment extreme caution must be taken to avoid life threatening complications like anaphylactic shock due to spillage of cyst contents. Laparoscopic approach has also been advocated for uncomplicated hydatid cyst of the spleen. Chemotherapy and newer methods, such as Puncture, Aspiration, Injection, and Re-aspiration (PAIR) technique using hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts [2].

Albendazole is an effective adjuvant therapy in the treatment of hydatid cyst. There are less chances of recurrence in patient who received albendazole therapy, as we prescribed in our case [2]. Following treatment, hydatid cysts are best followed sonographically due to the absence of ionizing radiation and ease of access to this modality [7].

Conclusion

Splenic hydatid cyst should be kept in mind in patients presenting with left upper abdominal pain, especially from endemic area. An early diagnosis is recommended to avoid further complications.

References