Giant splenic cyst: A case report and consolidated review of literature with radiographic features

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ABSTRACT

The spleen is a relative stranger to the world of cysts and tumors, which when discovered, are often incidental findings in otherwise asymptomatic individuals. Splenic cysts are far more common than solid lesions and can include congenital, inflammatory, vascular, post-traumatic, neoplastic, and parasitic types. Parasitic cysts in the spleen are usually caused by the larval forms of *Echinococcus*, a cyclophyllid tapeworm. However rare, it is important to recognize the splenic presentation of this uncommon entity to prevent life-threatening complications like anaphylaxis induced by traumatic or spontaneous cyst rupture. We report here a case of a giant dumbbell-shaped splenic cyst. Hydatid disease of the spleen should be considered in the differential of every patient presenting with a cystic lesion of the spleen in the endemic areas until proven otherwise.

Key words: *Cystic splenic lesions, Echinococcus, Hydatid disease, Spleen*

Hydatid disease (HD) is a zoonotic health problem endemic in cattle rearing areas of South America, Africa, Middle East, Southern Europe, India, and Australia. It is caused by the larval form of the tapeworm *Echinococcus granulosus*, *Echinococcus multilocularis*, *Echinococcus vogeli*, or *Echinococcus oligarthrus*. Among these organisms, *E. granulosus* and *E. multilocularis* are the most common organisms involved in human hydatidosis, causing cystic Echinococcosis and alveolar Echinococcosis, respectively. The involvement of the spleen in HD is rare, and isolated splenic involvement is even less common [1]. Worldwide incidence of splenic hydatid is 0.5–4% [2]. We present the case of a giant dumbbell-shaped splenic cyst in a 58-year-old coming in with the chief complaint of pain abdomen and discuss some clinical correlates of this presentation.

CASE REPORT

A 58-year-old female was referred to our radiology clinic with a chief complaint of abdominal pain and decreased appetite lasting more than 4 weeks. On clinical examination, the patient was afebrile, but her abdomen was found to be enlarged, indicating splenomegaly.

Routine laboratory tests including hemoglobin, hematocrit, red blood cell (RBC) counts, lactate dehydrogenase (LDH), bilirubin, and liver and kidney function tests, showed no significant abnormality. Since RBC count and LDH were in the normal range, hereditary anemias and hypersplenism were no longer considered. Erythrocyte sedimentation rate was relatively normal for patient parameters, but the white blood cell count was slightly elevated concerning for a myeloproliferative disease, splenic lymphoma, or an infectious etiology of malaria - which is indigenous to our region. The patient's peripheral blood film was further evaluated by pathology to rule out all of the above. Following which, the patient was brought to the radiology department, where we performed abdominal ultrasonography (USG) and revealed a cystic lesion in the spleen with daughter cysts.

Since the patient complained of sudden onset dyspnea soon afterward, along with persistent abdominal pain disproportionate to the examination findings, a contrast-enhanced abdominal computed tomography (CECT) was performed to rule out a contained perimembranous, peritoneal, or pleural rupture of the cyst, apart from pre-operative planning. A chest X-ray was also obtained to rule out pulmonary involvement. In USG, we observed a well-defined, multivesicular cystic mass in the location of splenic hilus, measuring 12 cm×11 cm. The left kidney was compressed and displaced to the inferolateral position.

On CECT scan with oral and IV contrast agent, we confirmed the finding of a non-enhancing, hypoattenuating cyst, almost entirely replacing the splenic parenchyma (Figs. 1 and 2). It had well-defined smooth borders and contained multiple and round daughter cysts in the periphery of the lesion (Fig. 3).

With these imaging findings, the final diagnosis was made as HD of the spleen, and the patient underwent elective splenectomy. The lesion was proved to be hydatid cyst pathologically.

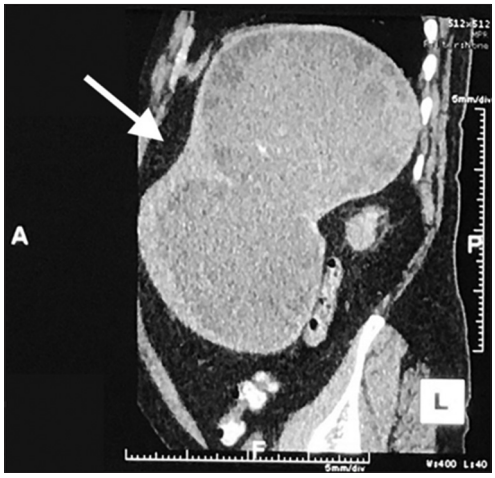

Figure 1: Computed tomography image reformatted in the sagittal window. Arrow shows the extent and size of cyst, almost completely replacing the splenic parenchyma

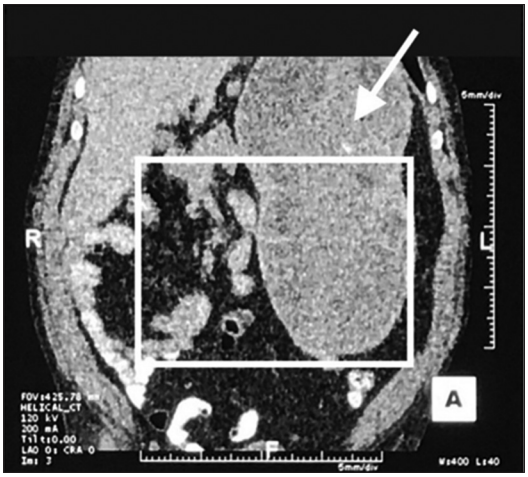

Figure 2: Computed tomography image reformatted in the coronal section. Arrow demonstrates the expansive encystment of the spleen

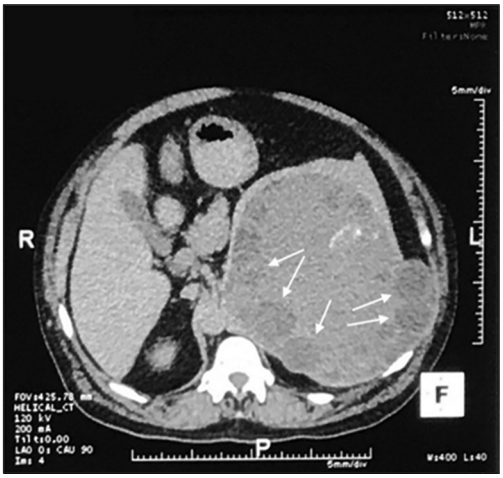

Figure 3: Axial computed tomography with arrows pointing to multiple, round, and peripheral daughter cysts

Besides surgery, the patient was also on medical treatment with albendazole and was in good health with no recurrence 6-month postsurgery.

DISCUSSION

Rare as it is, splenic hydatidosis has been reported since Berlot first described it as an autopsy finding back in 1790 [2]. Splenic involvement in HD is usually uncommon, representing <2% of all human infestations by *Echinococcus* [3], with isolated HD of the spleen being even rarer [4]. In India, the recorded prevalence of the splenic hydatid cyst is 2.5%, with the highest incidence reported in the central parts of the country [5]. For echinococcal encystations, the most commonly involved organ is the liver (75%), followed by the lung (15.4%) and the spleen (5.1%) [6]. Other commonly involved sites include kidney, bones, and brain [7]. Possible routes of primary hydatid of spleen include arterial route after passing through the liver and lung [2]. Arterial spread of the egg is feasible if it escapes the liver and lung filters [8]. Secondary hydatid spleen usually follows systemic dissemination or intraperitoneal spread following ruptured hepatic hydatid cyst [2,9] or occurs after operations involving hydatidosis in other regions [10]. These cysts are generally slow growing and therefore have the potential to reach an enormous size and still be asymptomatic [11].

The first clinical indication of the presence of splenic HD is usually an incidentally discovered mass in the abdomen. The differential diagnosis of such lesions includes large solitary abscess or hematoma, intrasplenic pancreatic pseudocyst, and cystic neoplasm of the spleen.

USG and CT remain the most valuable tools for the diagnosis of a splenic cyst [12]. Plain radiograph may demonstrate an eggshell-like calcification in the splenic area suggestive of splenic hydatidosis [2]. USG has a sensitivity of approximately 90–95% [13]. The most common appearance on USG is that of an anechoic smooth, round cyst, and when daughter cysts are present, characteristic internal septations are seen. USG images in case of *E. multilocularis* infection show the typical hailstorm pattern, characterized by multiple echogenic nodules with irregular and indistinct margins [14]. As CT is the best modality for determining the number, size, and anatomic location of the cysts, it is preferred over ultrasound to detect extrahepatic encystments. Multivesicular hydatid cysts contain low-density peripheral daughter cysts attached to the higher density central mother cyst to create a difference in density, which results in a characteristic CT image of *E. granulosus*. Magnetic resonance imaging (MRI) is recommended in patients with negative serology and indeterminate USG and CT investigations. MRI can differentiate parasitic, non-parasitic, or traumatic cysts by demonstrating a "low-signal intensity rim" - the RIM SIGN - which has been described as characteristic of hydatidosis, best seen in the T2-weighted sequence. However, reliable, MRI should not be used as the first imaging method in patients with proven or suspected HD [2].

A combination of medical and surgical therapy is used, as evidence suggests that drugs principally act as parasitostatic agents and not as parasiticides, implying a low cure rate [10]. Albendazole is an effective adjuvant therapy as there is less risk of recurrence in patients who receive it [11].

In our case, multiple daughter cysts in the periphery; surrounding the central mother cyst, evident on both USG and CT, clinched the diagnosis of splenic HD. Since there was no evidence of rupture or free fluid in the peritoneum on CT, the patient was scheduled for an elective splenectomy for the following week. The diagnosis of hydatidosis caused by *E. multilocularis* was eventually confirmed on histopathology. The post-operative course was uneventful with 3 weeks of albendazole treatment. USG follow-up did not show any evidence of recurrence at 6 months.

CONCLUSION

We report this case due to the relative rarity of primary splenic hydatidosis, especially given the size in our patient. To sum-up, it is important to consider HD in the differential for splenic cysts, especially in endemic areas. Radiological investigations play a major role in the diagnosis of this clinical entity and significantly decrease morbidity, as our case also showed.

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