

Case Report

Primary Splenic Hydatid Cyst: a Case Report with Characteristic Imaging Appearance

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Abstract

A middle-aged lady presented with pain, tenderness and swelling in the left hypochondrium since one month. She had a history of contact with dogs and grazing animals. Sonography and computed tomography showed the pathognomonic signs of hydatid disease. The patient refused surgical treatment. She was discharged on Albendazole therapy and did not return for a follow up.

Introduction

Hydatid disease is caused by the cestode tapeworm *Echinococcus granulosus*. It is predominantly prevalent in live-stock rearing areas. Liver (75%) followed by the lungs (15%) are the most common sites of involvement in adults.^{1,2} Splenic involvement is rare and accounts for 0.9% to 8.0% of all hydatid disease.^{3,4} The diagnosis relies on a careful history along with radiological and serological investigations.

Case Report

A 55 year old diabetic lady presented with mild pain, tenderness and swelling in the left hypochondrial region of one month duration. She had a history of previous hospitalisation due to uncontrolled diabetes. Physical examination revealed a slightly pale, overweight lady. The abdomen was soft and a mass was detected in the left hypochondrium. Rest of the physical examination was unremarkable. Sonography showed a well defined round mass; eight cm in diameter of mixed echogenicity with undulating membranes arising from the medial aspect of the spleen

(Figure 1). Computed tomography (CT) scan revealed a well defined circular mass in the posterior part of the spleen with curvilinear calcification inside, producing a whirl shaped appearance (Figure 2). No enhancement was noted after injection of intravenous contrast.

Figure 1. Ultrasound of upper abdomen demonstrating a degenerated splenic hydatid cyst. The characteristic undulating membranes giving a twirled appearance can be appreciated. This is the spin or whirl sign.

Differential diagnosis at this stage was an old cyst with calcification or a splenic neoplasm. Routine laboratory investigations were normal. *Echinococcus* antibody titres were 1:64 (Normal 1:32). At this stage, history of contact with dogs or grazing animals was obtained which was positive. A splenectomy was planned but the patient refused

to have surgery. She was discharged on Albendazole therapy and did not return again for follow-up.

may occur as a result of rupture of a hepatic hydatid cyst with abdominal and pelvic dissemination.

Most hydatid cysts are asymptomatic and diagnosed incidentally. Clinical manifestations are non-specific and include abdominal pain, enlarged spleen and fever.^{4,5} Severe pain may occur secondary to infection or rupture. The cysts grow slowly and development takes 5-20 years.^{4,5} Associated liver and/or lung involvement may be seen in 20-30% of cases.^{4,5} Splenic cysts are usually solitary.

Diagnosis relies on radiology and serology. Sonography is helpful, especially in early stages of the disease when the lesion is cystic, in detecting the daughter cysts, cystic membranes, septa and hydatid sand.^{3,6} When the fluid pressure in the cyst becomes too high, dissections may occur resulting in detachment of the parasitic membranes. At this stage, these undulating pathognomonic membranes can be seen on sonography and CT. This is known as the snake or serpent sign.⁶ At a more advanced stage of collapse, the membranes appear as twirled and twisted on imaging, which is known as the spin or whirl sign.⁶ Calcification is best detected with a CT scan. Magnetic resonance imaging (MRI) helps in detecting central nervous system involvement of the disease. CT scan typically shows a homogenous fluid content with water attenuation values.¹ These may be confused with an abscess.

The presence of mural calcification and/or daughter cysts rules out other cystic lesions of the spleen like: epidermoid cyst, abscesses, haematoma, hemangiomas, pseudocysts, metastases and cystic neoplasms. Calcification is usually curvilinear or ring-like and involves the pericyst.⁷

Although there are a variety of pathognomonic signs of hydatid disease on imaging, they are not always present. Imaging when combined with serological tests like enzyme-linked immunosorbent assay (ELISA), indirect hemagglutination and immuno-electrophoresis can lead to a successful diagnosis of splenic hydatid in 90% of cases.⁸

In instances where modern imaging facilities are not available, plain X-rays may be used. Typically they will show an elevated left hemi-diaphragm, left hypochondrial soft-tissue mass with calcification, or the displacement of stomach and/or left colonic flexure.⁵ Eosinophilia is a non-specific finding in hydatid disease.

While surgery is the modality of choice for the treatment of splenic hydatid disease, there is disagreement over the technique to be used. Some authors prefer total splenectomy while others prefer conservative surgical techniques like partial splenectomy, cyst enucleation, de-roofing with omentoplasty and others. Those in favour of total splenectomy argue that there are lower risks of recurrence and perioperative hemorrhage.^{4,9} Those for conservative surgery believe that total splenectomy predisposes to sepsis related

Figure 2. CT abdomen shows a splenic hydatid cyst. Detached calcified membranes produce a characteristic whirl pattern. Curvilinear calcification can be seen in the pericyst.

Discussion

A hydatid cyst should be included in the differential diagnosis of a splenic mass. This is particularly true for endemic areas. Our case serves to highlight the features associated with splenic hydatid disease with particular reference to its imaging.

Adult *E. granulosus* worms live in the intestinal tract of the definitive host; usually a dog. The excreted eggs may be swallowed by intermediate hosts like sheep and cattle where they develop into small cysts. Humans may get infected either by direct contact with a dog or by ingestion of foods and fluids contaminated by the eggs.

Involvement of sites other than the liver and lung is rare but no site is immune. Bones may be involved in 0.5%-4% of the cases.² Generally vascularized bones like vertebrae, long bones and epiphysis are involved. Renal involvement, primarily of the cortex, may be seen in 2-3% of the cases.² Hydatid disease of the brain constitutes 2% of all intra-cranial space-occupying lesions in endemic regions.² Cardiac hydatid disease occurs in 0.02%-2.0% of cases and most commonly involves the left ventricle.² Other sites that may be involved include soft-tissues, breast, ovaries, pancreas, scrotum, inguinal canal and the chest wall. Hydatid disease in such uncommon sites may cause a significant problem in diagnosis.

Primary splenic involvement is rare. This is because the cyst embryos are trapped in the liver and/or the lungs after ingestion and therefore do not reach the systemic circulation to infect the spleen. Secondary splenic involvement

predisposes to sepsis related deaths and thus should be avoided especially in children.⁵ Conservative techniques may be used for superficial cysts, cysts confined to one pole and with cysts with extensive adhesions. If left untreated there is a high risk of rupture. Post-operative Albendazole therapy may help prevent recurrence.⁹

This case demonstrates an unusual but pathognomonic appearance produced by calcifications in the pericyst and collapsed membranes in the case of a degenerated hydatid cyst. We conclude that sonography and CT can provide pathognomonic evidence of splenic hydatid disease.

References

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