CASE REPORT

Calcified Splenic Hydatidosis: A Rare Presentation

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Key words: Hydatid disease, spleen, Splenic Hydatid cyst, splenic hydatidosis, splenic echinococciasis.

INTRODUCTION

Hydatid disease is known since ancient times and mostly caused by a cestode named as echinococcus granulosis. It's worldwide in distribution but common in endemic areas of Middle East, India, newzeland and Australia. It affects all age groups and both sexes with equal frequency¹.

Liver and lungs are the most commonly involved visceras. Splenic Hydatid is a rare manifestation of the disease (2-3.5%). Speculated route of spread is either arterial or retrograde venous which avoids the liver and lungs².

We are presenting a case of isolated splenic hydatidosis which was having atypical presentation, sorted out with the help of radiological and operative detail correlation. Case was managed by splenectomy.

CASE REPORT

A 65 year old hypertensive male presented in outpatient department with swelling in the left hypochondrial region for one year duration and pain for 15 days. The patient reported that mass had started enlarging over the last four months. He had a history of previous hospitalization due to uncontrolled hypertension. Physical examination revealed a slightly pale and lean male. The abdomen was soft and a hard mass was detected in the left hypochondrium. Rest of the physical examination and labs were unremarkable. Plain x-ray abdomen gave suspicion calcification in left abdomen. Sonography showed poorly defined mass in the spleen. Computed tomography (CT) scan revealed a poorly defined mass of 45x32x22 cm in the spleen with calcification but no inside detail attained. No enhancement was noted after injection of intravenous contrast.

Patient explored through midline, it was a huge calcified Hydatid cyst which was displacing the stomach and intestine to the right side of the abdomen. It was not possible to aspirate due to calcification.



Plain x-ray abdomen showing calcified lesion in left hypochondrium

Total splenectomy was done.

The patient had an uneventful post-operative recovery and was discharged on the 12th post-operative day with a regimen of albendazole.

Histopathology of the specimen confirmed a Hydatid cyst of the spleen.

DISCUSSION

Hydatid disease is endemic in Eastern Europe, Middle East, South America, North Africa, Indian subcontinent, Australia and New Zealand¹. Due to the poor sanitation and unhygienic environment peoples of indopak region are at particular risk.It affects all age groups and both genders. We encounter the male patient in sixties of his life.

This zoonotic is ascribed to cestode name echinococcus. Of the 4 known species of Echinococcus, three are of medical importance in humans. These are Echinococcus granulosis, causing cystic echinococciasis (CE); Echinococcus multilocularis, causing alveolar echinococciasis (AE); and Echinococcus vogeli. E granulosis is the most common of the three. E multilocularis is rare but is the most virulent, and E vogeli is the rarest.

Echinococcus granulosis is the commonest organism involved², with dogs as the definitive host and human beings acting as an accidental intermediate host. After ingestion, the eggs hatch and the resultant oncospheres penetrate the intestinal mucosa of humans and enter the circulation. They primarily lodge either in the liver, lungs or kidneys

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which are the organs acting as filters of the circulation of the body. In the organs where these organisms lodge, they slowly develop into a cavity.

The outermost layer is the adventitia (pseudocyst), consisting of fibrous tissue as a result of the granulomatous reaction of the viscera to the cyst, from which it is inseparable. The middle layer is the laminated membrane (ectocyst) and the innermost layer is the germinal epithelium (endocyst), being the only living part of the Hydatid cyst.

Segond and Potherat divided hydatids into three groups: (a) those containing much fluid but few daughter cysts, (b) those packed with vesicles and (c) suppurating cysts. Our reported case got calcified wall with presence of daughter cysts inside.

Hydatid cyst grows at a rate of about 1 centimeter a year. The largest cyst ever reported was in Australia and it contained 57 liters' of fluid. We encountered the cyst of 45x34x22cm size³.

The liver (70%) and the lung (15%) are the most commonly involved viscera's. Involvement of sites other than the liver and lung is rare but no site is immune. Splenic Hydatid is a rare manifestation of the disease (2-3.5%)⁴. Primary infestation of the spleen usually takes place by the arterial route after the parasite has passed the two filters (hepatic and pulmonary). A retrograde venous route, which bypasses the lung and liver, is also reported. Secondary Splenic Hydatid disease usually follows systemic disseminated or intraperitoneal spread following ruptured hepatic Hydatid cyst. Our case followed the primary infestation route. Other sites that may be involved include heart, nervous system, softtissues, breast, ovaries, pancreas, scrotum, inguinal canal and the chest wall⁵

Splenic Hydatid cysts are generally asymptomatic but can present with either pain in left hypochondrium or mass. Diagnosis is usually established incidentally during investigation of unrelated symptoms. Splenomegaly if appreciated can be the result of either splenic echinococciasis or portal hypertension. Our patient was admitted to hospital for pain and a rapidly enlarging mass in the left upper quadrant of his abdomen.

Other likely manifestations are rupture, primary and secondary infection, compression of biliary tree and systemic anaphylaxis abdominal echinococciasis has sparse data as literature only has individual case reports. Few presented with incidental ultrasound reports of cysts in splenic area which was advised for exclusion of some other issues. Eosinophilia might be present which was not in our patient. Serum immunoelectrophoresis is

currently the most reliable, with a sensitivity of approximately 90%, with one year positivity after the organism has been eradicated Marginal or crumpled eggshell-like calcifications in the splenic area on abdominal or chest radiograph are suggestive of splenic hydatidosis as was evident in our patient. Sonography may reveal a solitary unilocular lesion or rarely multiple well-defined anechoic spherical cystic lesions with hyperechoic marginal calcification in the spleen. We got clue of Splenic mass on ultrasonography.

Computed tomography may show the cystic lesion with or without the daughter cysts within the spleen .Few patients can have characteristic cart wheal which is almost undeniable proof hydatidosis. Wall calcification is more clearly seen with CT of the abdomen without having inside details. Patient under discussion was not having the text appearance. However, the diagnosis of most of the Hydatid Splenic cysts is confirmed on CT scan. The presence of mural calcification and/or daughter cysts rules out other cystic lesions of the spleen like: epidermoid cyst, hematoma, hemangiomas, pseudocyst, metastases and cystic neoplasm that arouse the possibility of Splenic Hydatid to us'. This issue is considered common in our environment but very infrequent when the affected organ is spleen. We finished with report of calcified cyst or mass in spleen on CT.

Due to the risk of the spontaneous rupture, splenectomy remains the main stay of treatment. Diagnosis of calcified Splenic Hydatid cyst was confirmed peroperatively. Splenectomy is the primary treatment in such cases which aims at eradicating the disease while decreasing the chances of recurrence by removing the intact cyst 8. Aspiration is recommended to reduce the intracystic pressure but not performed if total splenectomy is decided^{9,10}. We couldn't perform it due to calcification in pericvst and accordance splenectomy done in to recommendations. Postoperative course augmented by albendazole for one month. After six month follow up, the patient was alright 11.

The didactic interest of this case is based on the final diagnosis of a calcified Splenic Hydatid cyst in a patient who was presenting unusual clinical signs but showing suspicious findings in the first ultrasonography performed being confirmed by abdominal CT scan images. In the beginning these images induced confusion among experimented clinicians due to their great similarity with pathologic abdominal processes like the calcified mass in spleen or pseudocyst. Subsequently, the diagnosis of a large Hydatid Splenic cyst with calcification was confirmed.

CONCLUSION

Splenic hydatidosis is a rare manifestation of Hydatid disease and should be presumed in all calcified cystic lesions of spleen especially in endemic areas. Computed tomography remains the most sensitive investigation for diagnosis especially if calcification is there. Total Splenectomy is the treatment of this rare disease in adults.

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