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Splenic Hydatid Cyst - A Rare Entity

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ABSTRACT:
Hydatid disease is endemic in farming areas but occurs worldwide. Although liver is the most commonly affected organ almost any anatomical location can involved. Splenic hydatid disease constitutes upto 4% of cases of abdominal hydatid disease [1] and worldwide incidence is 0.5 - 4% [2] . The rarity of splenic hydatid cyst may pose a diagnostic challenge for clinicians especially in non-endemic areas . Splenic hydatid, being a rare entity, can occur primarily or in association with hepatic, pulmonary or multi-organ hydatidosis. In this case report we present a 60-year old female patient with concomitant splenic and liver hydatid cysts. We report this case because of its rarity.

INTRODUCTION

Hydatid disease (hydatidosis, echinococcal disease or echinococcosis) is a zoonotic infection caused by tapeworms of genus Echinococcus; among these, Echinococcus granulosus is commonly involved in humans. Berlot first described Splenic hydatid cyst as an autopsy finding in 1790. Humans become accidental intermediate hosts when contaminated water or food containing the parasitic larvae is ingested. Most hydatid cysts present 5-20 years later before it is diagnosed. The rate of growth of the cyst remains indolent yet unremittent by character and is dependent on immunologic relationship between the parasite [3]. As a rough estimate hydatid cysts increase in diameter by about 2-3cms every year.

CASE REPORT

A 60 year old female came to Opd with dull aching left upper abdomen pain since 8 months which was mild in intensity, had no relation with food and was on and off. Patient complained of malaise and weight loss since 5 months. Also there was intermittent fever since last one month. Upon clinical examination there was mild tenderness in Left upper quadrant with no palpable mass, no lymphadenopathy. Other systemic examination were normal.

Laboratory examination showed no abnormalities except the anti-echinococcal IgG antibody which was positive 2.10 (>1.1 s/o positive report).

Plain radiograph of the abdomen and chest radiograph were normal. Abdominal usg revealed a well defined multi cystic lesion measuring 5.3*4.6*4.9 cms with thick separations in spleen and 6.7*6.5*5.5 cms in liver. CECT abdomen revealed a 5.6*5.6*5.8 cms located cyst with many septa originating from the lower pole of spleen with central and peripheral calcification, Medially it is seen abutting the right dome of diaphragm. A second cyst of size 6.7*6.5*5.5 cms from segment viii of right lobe of liver is noted. HRCT chest was normal.

In view of elective splenectomy the patient has been vaccinated to counter capsulated organisms (Haemophilus influenza,Meningococcal and Pneumococcal) prophylactically one week prior to prevent OPSI. The patient was started on tablet albendazole 400mg BD one week prior to reduce tension, stabilise cyst and decrease chances of rupture during surgery. Exploratory laparotomy with

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deroofing and partial cystectomy was done for liver hydatid cyst. The cyst contents were aspirated after injecting hypertonic saline without spillage of the contents, followed by removal of inner germinal layer. Splenic hydatid cyst was resected en bloc with spleen.

Microscopy revealed cyst wall composed of acellular laminated membrane along with degenerated and calcified material.

The postoperative period was uneventful and was discharged with Tab.Albendazole 400mg BD for 8 weeks. The clinical and ultrasound followup showed no evidence of recurrence at 6 months.

DISCUSSION

Hydatid disease caused by echinococcus granulosus exists in endemic sheep and cattle raising countries worldwide.The human infections are caused by four different species of echinococcus , of which Echinococcus granulosus and Echinococcus multilocularis are the most common. The other two species namely Echinococcus oligarthus and Echinococcus vogeli , are seldom affiliated with human infection

It commonly affects liver (50%-70%), followed by lungs (5%-25%). Primary involvement of spleen is rare with worldwide incidence of 0.5-4% [2], but in India it is higher amounting to 2.5% [4].

Dogs and wolves are the primary hosts. Sheep, cattle and deer are the intermediate hosts. Humans enter the cycle through infected canine faeces and become intermediate hosts [5]. Involvement of spleen is rare because only 15% of the cyst embryos enters the systemic circulation with the rest being trapped in liver and lungs. Primary infestation of spleen occurs when the eggs of parasite escape the liver-lung barrier through the arterial route [6]. The other route necessitating the spread of parasite is via retrograde pattern through the splenic and portal venous system bypassing liver and lung. Secondary splenic hydatid disease is caused by the rupture of hepatic hydatid cyst leading to systemic dissemination or intraperitoneal spread.

The presentation of splenic hydatid disease can greatly vary. They are mostly asymptomatic, solitary, slow growing and diagnosed incidentally. The common symptoms are abdominal discomfort, pain, palpable mass in the left upper quadrant. Sometimes it can present with vague symptoms like fever, burning micturition and vomiting without Spleenomegally [7].

There are various serological tests for diagnosis, screening and post op follow up for recurrence. Serum immunoelectrophoresis is the most reliable test with 95% sensitivity. Indirect haemagglutination is 85% sensitive. Western blot and ELISA have also been used in addition to eosinophilia, high IgE and IgM.

Untreated hydatid cyst can lead to inflammation, secondary infection, intraabdominjal rupture, anaphylactic shock, acute abdomen, compression of other viscera and fistula formation. Rupture of the cyst may cause peritoneal irritation, urticaria, anaphylaxis and death.

Differential diagnosis of splenic hydatidosis are cystic lesions of spleen such as splenic abscess, epidermoid cyst, hematoma, pseudocyst and neoplasm [8]. Pre operative diagnosis may be challenging due to resemblance of the presenting symptoms and the radiological findings.

Abdominal radiograph shows marginal or crumpled egg shell like calcifications in spleen suggestive of hydatidosis [9]. Other features seen are elevated left hemidiaphragm, reactive left pleural effusion, displaced stomach bubble and left colon . USG is the primary investigation and is useful in detecting daughter cysts. A classical sign (Water lily sign) is due to the detachment of endocyst results in floating membranes within the pericyst . CT has higher sensitivity than USG and is the investigation of choice. CT attenuation depends upon intracystic contents.

Hydatid cyst consists of three layers. The pericyst (outermost layer) is made up of fibrous tissue, ectocyst (middle layer) contains laminated, hyaline and acellular membrane [laminated membrane produces new generation of parasites]. endocyst (innermost layer) is the germinating layer consisting of daughter cyst and brood capsules with scolices which secretes the hydatid fluid.

Gross - Cyst wall consists of multiple grey yellow membranous tissue, surface is smooth, glistening with areas

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of congestion. Microscopy - Cyst wall shows eosinophilic laminated cuticle along with cholesterol clefts, brood capsules, scolex, and mixed inflammatory cell infiltrate containing neutrophils and lymphocytes. Coagulative type of necrosis and haemorrhage are also seen.

A combination of medical and surgical line of management is required. The drugs used in medical line of management have parasitostatic action and not parasiticidal action, resulting in low cure rate. These drugs interfere with mechanisms of glucose absorption through the wall of the parasite. Medical treatment is used in preoperative period to reduce the size and in postoperative period to reduce recurrence. Medical treatment remains the mainstay in postoperative followup period. Tablet Albendazole 400mg BD one month prior or four days preoperatively and 2 months postoperatively is recommended.

Both open and laproscopic surgeries are done. When the size of the cyst is small and located superficially without any complication, laproscopic surgery is preferred. When there are multiple cysts, large in size, located deep in the organ, infected or ruptured, open surgery is preferred[10]. In cases of isolated splenic hydatid cyst surgery is preferred.

Owing to the risk of spontaneous or traumatic rupture causing anaphylaxis, splenectomy is the treatment of choice especially with large hydatid cyst since splenic parenchyma is reduced notably due to pressure atrophy. Partial splenectomy carries a risk of poor vascular control and carries a risk of postoperative infection, hence total splenectomy is the treatment of choice. Patients are vaccinated before splenectomy when posted electively to decrease the chances of OPSI.

Due to spillage secondary infection occurs in 2-21% of cases. Recurrence is not seen after complete resection of cyst [10]. Post surgery the patients are kept followup period for at least 3 years and patients who are managed with medical line of treatment with USG every 6 months [11]

CONCLUSION

In conclusion the rarity of the splenic hydatid disease makes it a formidable early diagnostic challenge particularly in non-endemic areas because it needs treatment before the cyst wall ruptures and patient presents with shock and anaphylaxis. Hydatid disease should be taken into consideration in the differential diagnosis of all the cystic masses of spleen/liver. CT is the investigation of choice for diagnosis. The splenic hydatid cyst may turn out to be a daring surgical problem, hence the anatomical relations of the cyst must be established before surgery on account of varied presentations. The treatment of choice in adults is total splenectomy as it provides complete cure offering low morbidity and mortality rates. In children spleen preserving surgery should be considered to prevent OPSI. Preoperative and postoperative medical prophylaxis should be considered to ensure complete healing.

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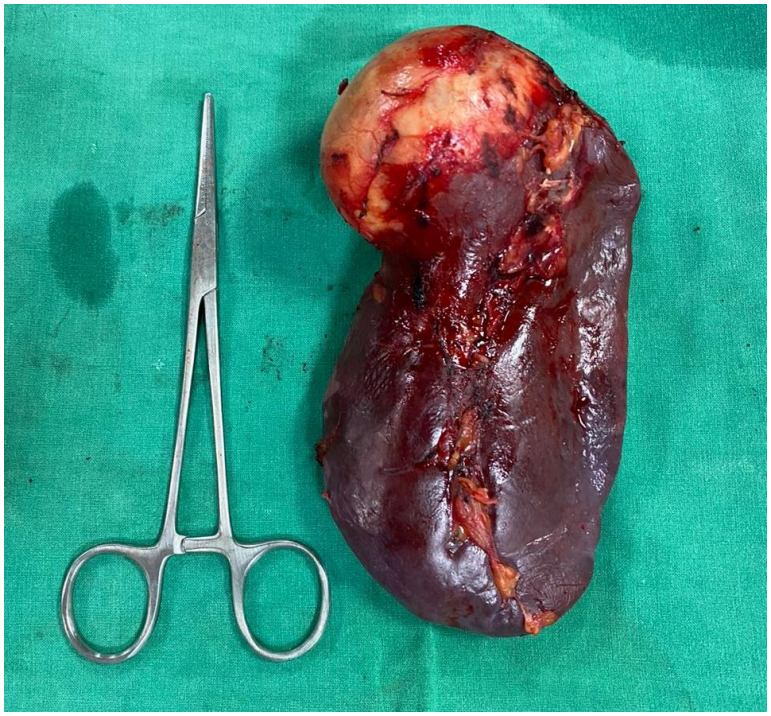
IMAGES



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