CASE REPORT

A Rare Primary Malignant Hydatid Cyst of Spleen : A Case Report

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ABSTRACT

Malignant hydatid disease of spleen is a misnomer as it is a benign condition caused by Echinococcus multilocularis. The entire infected organ is replaced by multiple small cysts over a period of time.It is difficult to treat and mimics clinically and prognosis wise to malignancy-hence the name.Invasion of isolated spleen is rare in Hydatid disease.

Here we present a case in 68 years old elderly lady who initially came with mass per vagina since 15 yrs with CT abdomen showin a complete malignant hydatid cyst totally replacing the spleen. Splenectomy was performed and massive spleen measuring 34cm X 30 cm x 26cm removed.Histopathological examination confirming the thin wall of spleen with entire parenchyma replaced by daughter cyst.

Keywords - Malignant Echinococcus cyst, hydatid multilocularis ; Splenectomy

INTRODUCTION

The incidence of splenic involvement by hydatid cysts in relation to the rest of the abdominal viscera is very low[1], extremely rare even in endemic population 0.5 to 4% of all cases of hydatidosis.[2-3].Man is an accidental intermediate host, as entry of the larval forms into humans represents an end stage in its life cycle. Alveolar Echinococosis (AE) is a severe helminthic disease that is highly lethal in humans caused by the larval form of the parasitic tapeworm Echinococcus multilocularis.

A classification system has been designated as the "PNM" system (P = parasitic mass, N = involvement of neighboring organs, M = metastasis). the system takes into consideration the localization of the parasite , the extent of lesion involvement, regional involvement, and metastasis.

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 dissemination or intraperitoneal spread following ruptured hepatic hydatid cyst [4-7].The splenic hydatid cysts may suppurate,fistulise to adjacent organs or rupture into the peritoneal cavity [8].

We report a case in a 68 years female who was admitted for prolapse of the uterus and incidental finding of giant hydatid cyst totally replacing the spleen was noticed on ultrasound abdomen.

CASE REPORT

A 68 years elderly lady presented to out patient department with mass per vagina since 15 yrs

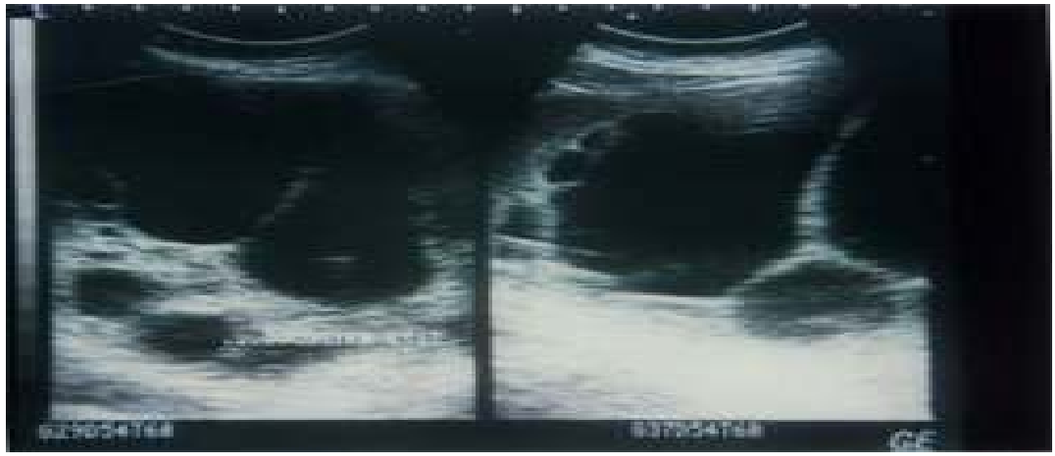

Figure :1 – USG Abdomen Showing Large Hypoechoic Multi Cystic Lesion

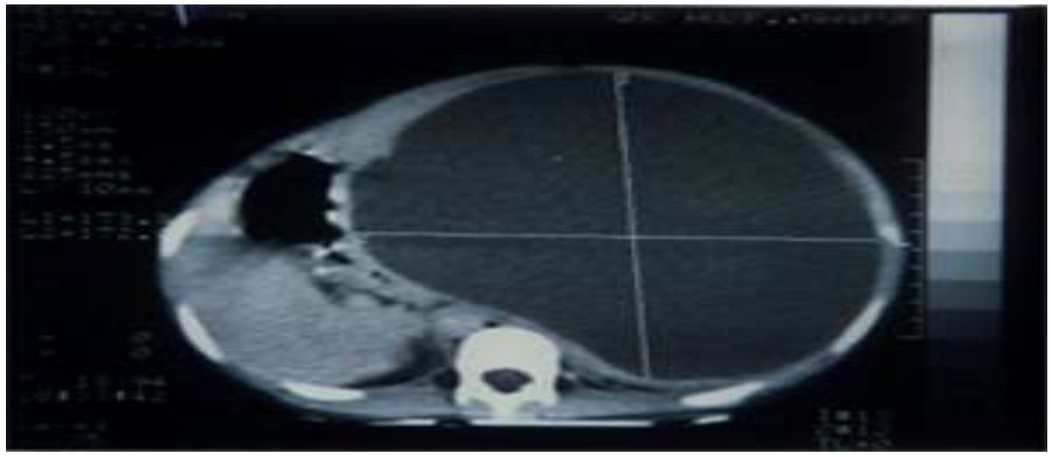

Figure :2 – CT Scan Abdomen Showing Grossly Enlarged Spleen Compressing Stomach

and pain abdomen since last six months. She was not a diabetic but hypertensive. There was no significant past or family history. On examination she was moderately built and nourished ,but appeared pale. Vital signs were within normal limits.Per abdominal examination revealed painless massive splenomegaly palpable upto the umbilicus. Bowel sounds were normal.Per speculum examination revealed 3rd degree uterovaginal prolapse with cystocele, rectocele and enterocele.

Hematological examination showed a haemoglobin level of 10g/dl, a hematocrit of 29.4% and total leucocyte count of 4500 cells/ cumm, with neutrophils 77 % ,lymphocytes 20% and eosinophils 3 % on differential count. Platelet count was 100,000/cu.mm Peripheral blood smear showed a dimorphic blood picture . Biochemical parameters were within normal limits. Serological tests for HIV, HbsAg and VDRL were negative. A preliminary diagnosis of Hypersplenism was made.

USG abdomen (Fig-1) revealed a large thick wall non enhancing hypodense cystic lesion with multiple septations measuring 30 x 26 x 24 cm showing well enhancement and calcific specks causing mass effect and displacement of adjacent

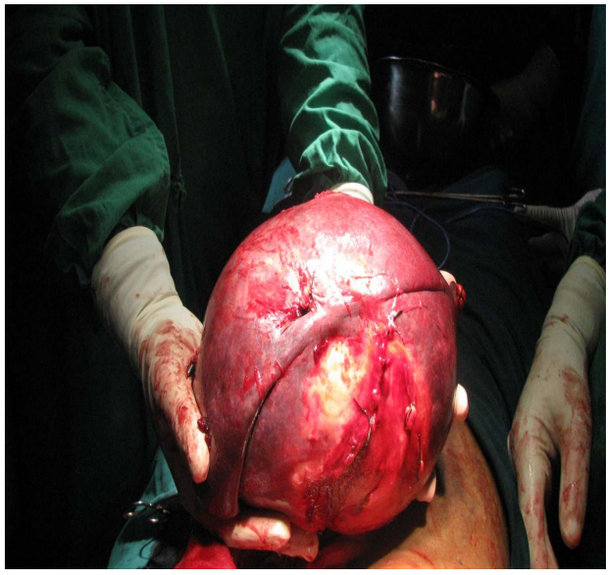

Figure - 3 : Spleen Measuring 34 cm X 30 cm X 26 cm

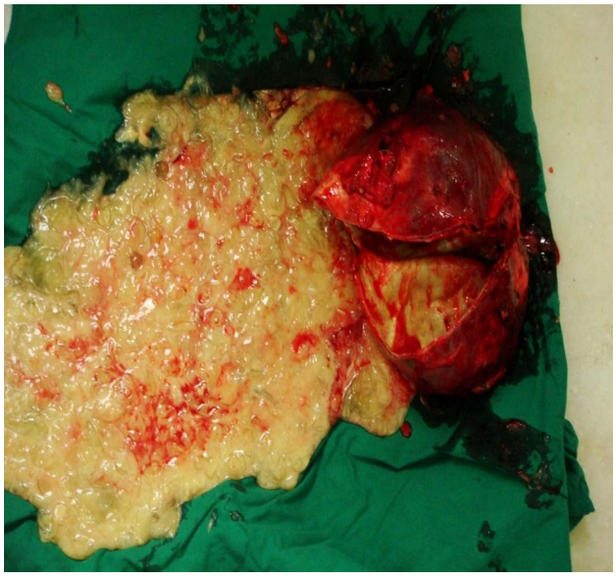

Figure - 4 : Cut section of spleen showing extensive replacement by multiple daughter cysts

organs. Computerised tomography (Fig-2) abdomen suspected to be as hydatid cyst or pseudopancreatic cyst. Endoscopy for dyspeptic symptoms was done which revealed severe erosive gastritis and extraluminal compression

Pre operative antihelmenthic regimen consisting of Albendazole was given to decrease the chance of anaphylaxis and tension in the cyst wall.

A midline vertical incision was taken which was later extended horizontally to facilitate the exposure of spleen.The hilum was displaced upwards towards the diaphragm which was ligated and cut. The cyst was separated from fibrous adhesions and en-bloc excision of spleen was done. Patient did not have any signs of anaphylaxis.

The excised spleen measured 34x30x26cms and was almost entirely replaced by hydatid cyst containing multiple daughter cysts (Fig-3&4). Histopathological examination showed thinned out splenic tissue with thousands of daughter cysts, confirmed as Echinococcus multilocularis.

The postoperative course was uneventful.The patient was given prophylactic vaccination against Streptococcus pneumonia, Haemophilus influenza type B and Neisseria meningitides, with prophylactic penicillin. The patient was discharged on the seventh postoperative day and prescribed antihelminthic therapy. No post splenectomy infection was encountered.

DISCUSSION

Malignant hydatid disease of spleen is a misnomer as it is a benign condition caused by Echinococcus multilocularis. The entire infected organ is replaced by multiple small cysts over a period of time. It is difficult to treat and mimics clinically and prognosis wise to malignancy-hence the name. Invasion of isolated spleen is rare in Hydatid disease.

Hydatid cysts can develop anywhere in human body, liver is the most frequently involved organ (52 –77 %) followed by lungs (10- 40%)[9]. Hydatid cyst is the only parasitic cyst of the spleen and it is said to be twice as common as the non parasitic variety. [10]. In India the incidence of hydatid cysts at unusual sites is higher as compared with other parts of the world.

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However the incidence of isolated splenic hydatid cyst in India is very rare with very few case reported to be infested with Echinococcus multilocularis. Primary infestation of the spleen usually takes place by the arterial route after the parasite has passed two filters (hepatic and pulmonary). The hexacanth embryos will be trapped in splenic capillaries.

Secondary splenic hydatid disease usually follows systemic dissemination or intraperitoneal spread following ruptured hepatic hydatid cyst. As the hydatid cyst grows, it may cause compression of the segmental vessels of the spleen resulting in extensive pericystic splenic atrophy. In addition, the hydatid cyst may entirely replace the splenic parenchyma as seen in this case due to chronic aseptic pericystic.

Inflammation – adhesion to nearby structure develop leading to fistulisation between cyst and adjacent organs, such as stomach, pancreas, left colon,left kidney or bronchus may develop.

Eosinophilia may be a finding which was not seen in our case. Serological tests are available but its importance is diminished with ultrasound, computerised tomography and magnetic resonance imaging.Computerised tomography is more accurate than ultrasound in localising and delineating extent of the cyst.

Histopathological examination should be done to confirm the parasitic nature of splenic cyst. Hydatid immunoelectrophoresis, ELISA,latex agglutination and indirect haemagglutination test are the different serological tests for diagnosis, screening and follow up for recurrence.

The main differential diagnoses of splenic hydatidosis are splenic cystic lesions such as pseudocyst, abscess, haematoma and cystic neoplasm .

The treatment of hydatid cysts is mainly surgical, although controversy exist over the best type of surgical approach. Partial splenectomy is risky because it is difficult to have a vascular control while incising the splenic tissue. However partial splenectomy with omentoplasty can be done for cases with unresectable cyst tightly adherent to other structures.

With pre- and post-operative 1-month course of Albendazole and 2 weeks of Praziquantel sterilizes the cyst, minimizes the chance of anaphylaxis, reduces the tension in the cyst wall (spillage during surgery is less) and the recurrence rate post-operatively.

Intra-operatively, the use of hypertonic saline or 0.5% silver nitrate solutions before opening the cavities tends to kill the daughter cysts and therefore prevent further spread or anaphylactic reaction.

CONCLUSION

The purpose of this case report is to emphasize the fact that Hydatid disease should be suspected in cystic lesions affecting any organ in the body,especially in endemic areas of the world.

Medical treatment should precede and follow the surgical intervention,in this case en-bloc excision of Spleen.

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