



National Journal of Medical and Allied Sciences

[ISSN Online: 2319 – 6335, Print: 2393 – 9192|Case report |Open Access]

Website:-www.njmsonline.org

PRIMARY SOLITARY MULTILOCLAR GIANT SPLENIC HYDATID: A CASE REPORT

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Abstract

Primary Splenic Hydatidosis is a rare clinical occurrence having a worldwide prevalence of just 0.5% - 4%. We report here a case of a 39 year old homemaker hailing from Maharashtra (India), who presented to the surgical outpatient department of ESIPGIMSR & Model Hospital (Andheri), with chronic abdominal pain associated with heaviness in the left hypochondrium. A diagnostic triad of Ultrasonography, Contrast-Enhanced Computed Tomography and Enzyme Linked Immunosorbent Assay for Hydatid antibodies confirmed the presence of a primary solitary multilocular giant Splenic Hydatid. Open Total Splenectomy was performed as a safe and definitive treatment for Primary solitary multilocular giant Splenic Hydatid.

Key words: Echinococcosis; Primary Splenic Hydatosis; Splenectomy

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Introduction:

Echinococcosis (Hydatidosis) is a frequently encountered zoonotic parasitic infestation caused by the larval stage of *Echinococcus granulosus*. Known to be endemic in the sheep rearing countries like Greece, Turkey, the Middle East, Northern China, South America, Australia, New Zealand and South Africa, Echinococcosis has percolated into the developed countries due to urbanisation and extensive international tourism. Echinococcosis is therefore a growing public health concern across the globe.^[1] In India, Hydatidosis is encountered, occasionally in the farming areas of Andhra Pradesh and Tamil Nadu and the sheep and cattle rearing areas of Bihar and Uttar Pradesh.^[2] Nagpur has been reported to have the highest incidence of

Hydatid disease in India.^[3] As compared to the rest of the world, the incidence of Hydatid cysts occurring at unusual sites is higher in India.^[4] Hydatidosis can involve any organ system in the body, but liver (60% - 70%) followed by lungs (20%) are the two commonest sites for Hydatidosis.^[5] Splenic Hydatidosis is an uncommon entity in the general population with a reported incidence of 0.5-6.0% within abdominal Hydatidosis and less than 2% to 3.5% of all human Echinococcosis.^[6] Primary Splenic Hydatidosis i.e. Hydatid disease involving exclusively the spleen, has a worldwide incidence of only 0.5-4%, the highest being in Iran (4%).^[7] Here we report a case of Primary Splenic Hydatidosis in a 39 year old

homemaker presenting with chronic abdominal pain and fullness in the left upper abdomen. An abdominal Ultrasound followed by a Contrast-Enhanced Computed Tomography (CT) of the abdomen revealed the presence of a giant multilocular Hydatid cyst in the spleen. The Enzyme Linked Immunosorbent Assay (ELISA) for Hydatid antibodies was positive. The patient underwent a curative Open Total Splenectomy.

Case report

A 39 years old homemaker hailing from Maharashtra (India) presented to the surgical outpatient department of ESIPGIMSR & Model Hospital (Andheri), with chronic abdominal pain associated with heaviness in the left upper abdomen for duration of two years. She denied any history of jaundice, cough or respiratory distress, bleeding diathesis, hematemesis, melena, abdominal trauma and anorexia or weight loss. Her past history did not reveal anything significant and she harboured no other medical comorbidities. The patient also denied having any prolonged contact with farm animals or having travelled to endemic areas. General examination revealed no abnormalities. Per abdominally, massive nodular splenomegaly with the spleen extending beyond the umbilicus was the only significant finding. Systemic examinations revealed no abnormalities. Haematological investigations including complete blood count, coagulation profile, renal function tests and liver function tests were within normal limits. The Chest Skiagram was normal. Real-time Ultrasound of the abdomen revealed a large intraparenchymal multilocular cystic lesion in the spleen suggesting Splenic Hydatidosis, with the spleen measuring about 18cm x15 cm. The rest of the abdominal Ultrasound was normal. Abdominal CT scan reported a giant spleen measuring 18cm x 15 cm, harbouring multiple, complex, cystic hypodense lesions. The cysts contained multiple hypodense

daughter cysts within them and a rim of wall calcification was seen along inferior aspects of the larger cysts. The multilocular hydatid cyst was located centrally and intraparenchymally within the spleen, with a very thin rim of overlying splenic tissue. The spleen was reported to be adherent to the lateral abdominal wall and the diaphragm. The CT scan confirmed the absence of Hydatid disease in any other abdominal viscera, thereby establishing the diagnosis of Primary Splenic Hydatidosis. Serological Enzyme Linked Immunosorbent Assay (ELISA) for Hydatid antibodies was positive. (Echinococcus -Ig G serum = 6.9).

Following all the investigations, we planned an open Total Splenectomy under general anaesthesia for the patient as a definitive cure for the disease. The patient received Pneumococcal and H. Influenza b and N. Meningitidis vaccines and was started on Albendazole, 400 mg orally twice a day with meals 2 weeks prior to the planned surgery. The large size of the splenic hydatid, thin overlying splenic parenchyma and dense perisplenic adhesions prevented us from attempting laparoscopy or hand assisted laparoscopy in this patient for the fear of fatal anaphylaxis and recurrence following an intraperitoneal spillage of the protoscolex rich fluid due to accidental rupture of the cyst. Thus an open total Splenectomy was performed through an upper midline abdominal incision under general anaesthesia. The spleen was densely adherent to the diaphragm, splenic flexure, omentum and the lateral abdominal wall. The rest of the abdominal organs including the liver were normal. The spleen was isolated using sponges soaked with the scolicidal solution of 1.5% cetrimide-0.15% chlorhexidine (10% Savlon) to counter any inadvertent rupture of the cyst and spillage. (Fig.1) Hypertonic saline (15%NaCl) was instilled into the cyst before commencing any splenic dissection in order to kill the daughter cysts. Meticulous dissection was carried out to excise the

spleen en -bloc. (Fig.2) Post Splenectomy a thorough peritoneal lavage with hypertonic saline solution (15%NaCl) was given to eliminate any potential contamination. Postoperative antihelminthic therapy was not indicated in this patient because the total splenectomy was uneventful and there was no evidence of residual disease elsewhere. Following an uneventful post operative course, the patient was discharged on the tenth postoperative day after suture removal. A follow-up after six months did not reveal any recurrence clinically and on Ultrasonography.

The gross histopathological examination revealed a giant spleen weighing 1060 grams and measuring 17cms x 13cms x 5cms. The external surface was nodular and mottled with greyish white areas. The cut surface revealed several multiloculated cysts containing multiple, variable sized daughter cysts. (Fig.3) On microscopy, an outer laminated acellular chitinous ectocyst, inner germinal endocyst with numerous scolices were seen. The acellular laminated membrane of the Echinococcus cyst stained strongly for Periodic-acid Schiff (PAS) stain. In view of the rarity of this clinical entity, a formal consent for the publication of this case report was obtained from the patient and her family.



Fig.2: En bloc Splenectomy



Fig.3: Splenic Hydatidosis

Discussion

Hydatid disease, known to be endemic in the sheep rearing Mediterranean countries, commonly involves the Liver (75 %) and the lungs (15%).^[1,5] In 10% cases, the disease involves uncommon sites like brain, cavernous sinus, sub-mandibular gland, thyroid gland, heart, pleura, chest wall, retro-crural tissue, kidney, spleen, pancreas, peritoneal cavity and inguinal canal, breast, bone and soft tissue.^[8] The incidence of Hydatidosis occurring at such unusual sites is higher in India as compared to the rest of the world.^[4] Isolated Primary Splenic Hydatosis is a rare clinical entity with a worldwide incidence of only 0.5-4%.^[6] The published prevalence of the Splenic Hydatidosis in India, is about 4.3%, with the highest incidence being reported from central India.^[9] Human beings, who are the definitive hosts in the life cycle of



Fig.1: Spleen isolated with Savlon soaked surgical mops

Echinococcus granulosus, contract Cystic Echinococcosis by accidental ingestion of Echinococcus cysts- containing organs of the infected intermediate hosts like sheep and goats or food contaminated with faecal material containing the tapeworm eggs or following prolonged contact with infected dogs and other carnivores.^[10,11] Berlott (1790) was the first to describe Splenic Hydatidosis as an autopsy finding.^[12] Dieulafoy was the first physician to diagnose Splenic Hydatidosis.^[13] Splenic Hydatidosis is classified as primary (isolated splenic involvement) and secondary (multiple organ involvement). Primary Splenic Hydatidosis occurs either due to antegrade infestation of the spleen via the arterial route after the parasite escapes the first (hepatic) and the second (pulmonary) Lehman's filter or a portal vein retrograde extension from the liver to the spleen (portal hypertension). Secondary Splenic Hydatidosis is a sequel to systemic dissemination following an intraperitoneal rupture of an extra-Splenic Hydatid cyst.^[14] Primary Splenic Hydatidosis is known to occur in both sexes and at all ages. The cyst grows insidiously at a rate of 0.3-1 cm per year gradually obliterating segmentary vessels leading to necrotic areas resulting in parenchymal destruction and extensive pericystic atrophy. The aseptic inflammation of the splenic parenchyma leads to perisplenitis which along with splenomegaly causes chronic abdominal pain. Perisplenitis extends beyond the splenic capsule forming adhesions with contiguous viscera i.e. stomach, colon, omentum and diaphragm.^[4] Thus colonic compression causes bloating and constipation. Diaphragmatic compression causes dyspnoea. Splenomegaly is usually the only sign that is elicited on physical examination. In due course the Splenic Hydatid cyst may undergo complications like infection, spontaneous rupture into the pleural or peritoneal cavity and fistulisation into adjacent organs like colon, stomach, left kidney

and Pancreas.^[15] Splenic Hydatid perforation into the left colon presents with massive gastrointestinal bleed.^[16] A lethal complication associated with Hydatid cyst is the spontaneous intra peritoneal rupture of the cyst causing fatal IgE induced anaphylaxis.^[17] Splenic Hydatidosis merits suspicion when investigating cystic lesions of the spleen, particularly in endemic areas, and has to be differentiated from other cystic lesions of the spleen, such as Cystic Hemangiomas, Lymphangiomas, Dermoid Cysts, Epidermoid Cyst, Pseudocyst, Abscess and Haematoma.^[18] The triad of Abdominal Ultrasound, Contrast Enhanced Computed Tomography (CT) and Enzyme Linked Immunosorbent Assay (ELISA) for Hydatid antibodies clinches the diagnosis of Splenic Hydatidosis. Abdominal Ultrasound is a ubiquitous, non-invasive, sensitive, and economical imaging modality for diagnosing Splenic Hydatidosis. The cyst may present as a solitary unilocular cyst or multiple well defined anechoic spherical cystic lesions or an anechoic spherical cystic lesion with hyperechoic marginal calcification as seen in this case.^[19] Abdominal CT confirms the cystic lesion with or without daughter cysts within the spleen with attenuation value near that of water and non-enhancement after intravenous contrast administration. CT conclusively detects the site, size localization and the number of cysts and the presence of cysts in other organs. CT is also the preferred modality for monitoring postoperative recurrences.^[20] Serological Enzyme Linked Immunosorbent Assay (ELISA) for Hydatid antibodies is currently the most reliable serological test, with a sensitivity of approximately 90%. It stays positive for up to one year after the infection has been eradicated.^[21] Our case tested positive by ELISA for Hydatid antibodies. Splenectomy is deemed as the gold standard for the treatment of Primary Splenic Hydatidosis. Surgical excision aims to inactivate the parasites, evacuate the cyst

cavity, remove the germinal layer, and obliterate the residual cavity thus eradicating the local disease, preventing complications, morbidity, mortality and forestalling postoperative recurrences.^[22] The extent of surgery varies from spleen preservation to Total Splenectomy. A Total Splenectomy is warranted when the spleen harbours either multiple cysts or a single large cyst located centrally or intraparenchymally, compromising more than 75% of the splenic parenchyma, as seen in our case.^[23] Preoperative vaccination minimises the risk of post Splenectomy sepsis.^[24] Pre- and postoperative Albendazole sterilizes the cyst, decreases the tension in the cyst wall, reduces the risk of anaphylaxis and reduces the postoperative recurrence rate. Intra-operatively, hypertonic saline or 0.5% silver nitrate is instilled into the cyst before opening it; this tends to kill the daughter cysts, thereby preventing further spread and anaphylactic reaction.^[25] We have instilled hypertonic saline in the cyst. Amongst spleen preserving surgeries, Partial cystectomy has been attempted for small superficial cysts, marginal cysts, localized polar cysts and densely adherent cysts. Here the splenic cyst is evacuated using a large-gauge sump drain and the residual cavity is filled with a scolicial solution followed by partial cystectomy.^[26] Other Spleen preserving surgeries like Cyst Enucleation, Deroofing of the cyst with Omentoplasty, Internal Drainage with Cystojejunal Anastomosis and external drainage, have been performed in cases where the cysts are inactive.^[27, 28] Spleen sparing surgeries are preferred in children since they are at higher risk of Splenectomy-related sepsis.^[28] Technological advancements have made laparoscopic and robotic Total Splenectomy and spleen-sparing surgeries safe, effective and offering all the advantages of minimal access surgery like reduced incision size, better cosmetic results and especially early return to work.^[29] Laparoscopy and conventional open surgery have similar outcomes

for patients with single, small-sized, superficially located Splenic Hydatid.^[30,31] For larger cysts however, the greatest deterrent for performing laparoscopy is the possibility of fatal anaphylaxis following intraperitoneal spillage of the protoscolex-rich cyst fluid due to an inadvertent rupture of the cyst during dissection or while extracting the specimen.^[32] The reported incidence of intraoperative fluid spill despite all the precautions, is about 5-10%.^[32,33] About 18% recurrences after surgery have been reported, secondary to incomplete removal, spillage, or growth of small occult cysts that were missed initially.^[33,34] Conventional open surgery is a preferable option when there are multiple cysts, large cysts, infected cysts, cysts in deep organ locations and recurrences.^[35] Here, our case, the giant multilocular Splenic Hydatid cyst with a thin overlying splenic parenchyma and dense perisplenic adhesions, posed a very high risk of rupture, haemorrhage and damage to surrounding viscera if attempted laparoscopically. We therefore opted to perform a conventional open total Splenectomy to minimise the risk of a life threatening anaphylaxis, haemorrhage or visceral damage. Khoury *et al.* advocate Laparoscopy for single, small-sized, superficially located uncomplicated splenic hydatid cysts.^[33] A recently evolved alternative to surgery is PAIR *i.e.* Puncture, Aspiration, Injection, and Reaspiration, a percutaneous drainage technique consisting of pre- and postoperative chemotherapy with Albendazole or Mebendazole, combined with percutaneous drainage of the cyst. PAIR is considered an effective alternative to surgery and chemotherapy especially for patients who refuse surgical therapy or have an unacceptably high risk for anaesthesia.^[36] Adjuvant medicinal therapy with antihelminthic agents is mandatory for cases with multiple cysts involving more than one organ, multiple recurrences, and severe peritoneal Hydatidosis. Recent studies promote the use of

Antihelminthic agents in combination with surgical therapy or PAIR. A prophylactic antihelminthic course of 2 to 4 weeks administered prior to Splenectomy (total or partial) sterilizes the cysts, reduces cyst pressure, and decreases the risk of anaphylactic reactions as a result of cyst manipulation. Postoperative antihelminthic therapy is not necessary in cases of isolated Primary Splenic Hydatosis when an uneventful total Splenectomy is achieved. Postoperative 2- to 6-week course of antihelminthic therapy is advocated for spleen-sparing surgery, recurrences, and extra-Splenic cysts. Albendazole, Mebendazole, and Praziquantel are the commonly used antihelminthic agents that have a good tolerance and safety profiles.^[36]

Conclusion:

Primary Splenic Hydatidosis should be suspected in all patients with cystic lesions of the spleen. Ultrasound and Computerised Tomography are useful imaging modalities for diagnosing and monitoring this disease. Conventional open Total Splenectomy combined with adjuvant antihelminthic therapy is the best curative procedure for large-sized or multiple splenic cysts, as surgery significantly diminishes risk of cyst rupture and its associated complications. Antihelminthics reduce the recurrence rates. Laparoscopy, considered safe and feasible for single, small-sized, superficially located uncomplicated splenic hydatid cysts. Safe guarding against all potentially life threatening complications, conventional Open Total Splenectomy still remains the safest treatment option for primary, solitary, multilocular giant Splenic Hydatid.

Acknowledgement

The authors acknowledge the contribution of the theatre staff for their sincere help and cooperation. They are also thankful to the medical records section, Radiology and Pathology departments at the hospital for their valuable

contribution and necessary support. The authors are grateful to Dr. Meenakshi Mathur, Dean, ESIPGIMSR & Model Hospital, Andheri for her encouragement and support.

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Conflicts of Interest: None Funding: None

Citation: Chaudhari N, Bobde S, Vaidya R, Chaudhari N. Primary Solitary Multilocular Giant Splenic Hydatid: A Case Report. National Journal of Medical and Allied Sciences 2015; 4(1):67-73