Case Report

Water-lily sign: A case report

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Abstract

Echinococcosis (or hydatid disease) is a zoonotic disease caused by the larval form of Echinococcus granulosus or Echinococcus alveolaris. It continues to be endemic in developing countries including India. Liver is the most common organ involved but the parasite can affect any organ in the body. A 45 year old lady who presented with abdominal pain was subjected to ultrasonography and MRI of the abdomen which showed the characteristic “water lily sign” of hydatid cyst. Patient underwent pericystectomy, and on followup screening ultrasound has remained free of disease recurrence.

Keywords: Echinococcosis, Echinococcus granulosus, water lily sign, pericystectomy

1. Introduction

Echinococcosis (Hytadid cyst disease) is a zoonotic disease which is endemic in many of the countries of Europe, South East Asia including India, Africa, and South America1. Humans are accidental intermediate hosts who acquire the disease by ingesting Taenia eggs. Although the liver and lungs are the most common sites for the cysts to develop, hydatidosis can also infrequently involve kidneys, brain, peritoneum, heart.

2. Case Report

A female patient aged 45 years presented with history of right upper quadrant pain, on and off, since three months with no relieving or aggravating factors. Pain was described as dull aching, nonradiating, and not related to food intake. There was no history of fever, nausea, vomiting, jaundice, or passing high colored urine.

On examination, there was no jaundice. Per abdomen exam showed a soft and nontender abdomen with no palpable organomegaly. Other system examination was unrevealing. Laboratory testing was significant for a complete hemogram with a hemoglobin of 9.2 and eosinophils of 8% on the differential. Liver function tests were normal. Ultrasound of abdomen showed a large cystic lesion measuring 11 x 8.5 x 6.3 cms with multiple septations and membranes within it, suspicious for hydatid cyst. Patient underwent an MRI (Magnetic Resonance Imaging) of the abdomen which confirmed the diagnosis of hydatid cyst occupying segments V and VI of liver. There was also an incidental note of a complex right ovarian cyst.
3. Discussion

Echinococcosis or the hydatid cyst disease is endemic in Indian and other countries which raise livestock. It is caused by the larval forms of E. granulosus and E. alveolaris. Liver (75%) and lungs (22%) are the commonest locations for the cysts but can rarely affect other organs such as the kidneys, bones, spleen, and brain\textsuperscript{2}. Dogs are the definitive hosts, while sheep, cattle, and man act as intermediate hosts. Humans are dead end hosts.

The adult Echinococcus granulosus resides in the small intestine of the definitive hosts, dogs or other canids. Gravid proglottids release eggs that are passed in the feces. After ingestion by a suitable intermediate host (sheep, goat, cattle), the egg hatches in the small intestine and release oncospheres which penetrate the intestinal wall and migrate through the circulatory system into various organs, especially the liver and lungs where they develop into a cyst and produce protoscolices and daughter cysts that fill the interior of the cyst. The definitive host becomes infected by ingesting these cysts from the infected intermediate host. After ingestion, the protoscolices attach to the intestinal mucosa, and develop into the adult stage.

Structurally, the hydatid cyst consists of a fibrous inflammatory outer layer (pericyst) which is of host origin, a laminated cuticular layer (ectocyst), and an inner germinal membrane (endocyst) from which the protoscolices develop and secrete a clear fluid. The germinal layer forms invaginations from which daughter cysts arise.

3.1 Diagnosis

The diagnosis relies on clinical manifestations, imaging, and serological tests\textsuperscript{3}. Ultrasound is the most preferred and cost-effective imaging technique. Gharbi et al. classified the appearance of the hepatic cyst into five different types based on the viability of the cyst\textsuperscript{4}.

<table>
<thead>
<tr>
<th>Type</th>
<th>Appearance</th>
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<tbody>
<tr>
<td>I</td>
<td>Univesicular, hypodense cyst containing hydatid sand</td>
</tr>
<tr>
<td>II</td>
<td>Univesicular hypodense cyst with undulating membrane</td>
</tr>
<tr>
<td>III</td>
<td>“Mother and daughter” cyst which is highly specific for hydatidosis</td>
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<tr>
<td>IV</td>
<td>Detached germinal layer producing the classical “water lily” sign.</td>
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<tr>
<td>V</td>
<td>Cyst with reflecting, calcified, thick wall</td>
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The presence of a detached laminated membrane from the pericyst, the presence of daughter cysts, and the presence of calcifications in the cyst wall are the pathognomonic characteristics of hydatid liver disease. CT (Computed Tomography) and MRI are the most sensitive and accurate methods of diagnosis.

For several decades, the intradermal test described by Casoni was used for diagnosis but it has a low sensitivity and specificity and not used in the present era of immuno-diagnosis. Amongst the immunodiagnostic methods, IFA (Indirect immunofluorescence assay) is highly sensitive and specific test. Other tests include the enzyme-linked immunosorbent assay (ELISA), immunoelectrophoresis test (IEP), and Counterimmunoelectrophoresis or co-electroosyneresis (CoES)

**Figure 1:** MRI abdomen showing detachment of the endocyst membrane with floating membranes within the pericyst which mimics the appearance of a water lily
3.2 Treatment

The current treatment options available are divided into either medical, percutaneous aspiration, or surgery. Medical treatment consists of chemotherapy with benzoimidazole compounds such as albendazole or mebendazole and is indicated for patients who have inoperable disease or whose general condition does not permit them to undergo surgical procedures. A large cyst which is superficially located and an infected cyst are contraindications for chemotherapy. The recurrence rates are as high as 25% to 30%.

Percutaneous aspiration consists of the PAIR technique (puncture of the cyst wall, aspiration of cyst contents, injection of a scolecidal agent, and reaspiration of the scolecidal agent). PAIR should only be performed in highly specialized centers with appropriately trained and experienced staff.

Surgical treatment is the gold standard for hydatid cyst and can be done either by open surgical techniques or by laparoscopic methods where expertise is available. Pericystectomy is the most common conservative surgery performed for the disease, while radical methods involve the complete removal of the pericystic membranes and the parasitic contents.

4. Conclusion

Hydatid disease continues to be a public health problem in many of the endemic countries including India. The diagnosis requires a high index of suspicion by the practicing physicians in an endemic area. Imaging techniques provide an accurate method for diagnosis of the disease. Although not the standard of care, medical treatment with albendazole and mebendazole is still effective in resource-limited areas.

References