



Spectrum of Presentations of Hydatid Cyst at a Tertiary Care Hospital in Non-Endemic Region – A Prospective Study

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Abstract

Aims & Objectives

1. To evaluate different presentations of Hydatid cyst
2. To evaluate unusual locations of Hydatid cyst.
3. To evaluate Hydatid cyst related complications

Conclusions: Hydatid Cyst should be kept in mind as a possible differential diagnosis in all cystic lesions occurring in any organ in the body. Complications due to Hydatid Cyst are many & varied in nature Hence a systematic approach to Imaging & careful analysis of possible complications is mandatory to arrive at an accurate diagnosis .

Keywords: Hydatid cyst, Unusual locations, Complicated Hydatid cyst.

Introduction

Hydatid cyst is a zoonotic infection of the human caused by larval stage of Echinococcus Granulosus. In adults, the liver represents the most prevalent site (50 –70 %) and lungs are the second commonest site (10-30 %) In children the lungs are the most prevalent site⁽¹⁾. It can occur in any part of the body either as a primary infection or secondary to infection in a primary site.

Pathogenesis: Dogs or other carnivores are definitive hosts & sheep are intermediate hosts. Humans are secondarily infected by the ingestion of food or water that has been contaminated by dog feces containing the eggs of the parasite. After the outer capsule of the egg has been ingested, the freed embryo (oncosphere) enters a

branch of the portal vein by passing through the duodenal mucosa. Most of these embryos become lodged in the hepatic capillaries, where they either die or to grow into Hydatid cysts. Some pass through the capillary sieve and become lodged in the lungs and other organs ^(4,6,9) As the hydatid cyst grows in size, the organ reacts by forming a connective tissue layer, the pericyst, a layer embracing the parasite which is called the endocyst. The wall of the parasite itself consists of a thicker laminated chitinous outer layer, the ectocyst, and a delicate inner layer, the germinal membrane also called the endocyst. a layer embracing the parasite. The symptoms may be related to a toxic reaction due to the presence of the parasite The germinal membrane gives rise to

the hydatid fluid, brood capsules, scolices and daughter cysts⁽²⁾ and the local and mechanical effects depending on the location and nature of the cysts and the presence of complications (El-Tahir et al., 1992). The cyst fluid is crystal clear. It is a transudate of serum, contains proteins, and is antigenic. If it is released into the circulation of the host, it can cause eosinophilia or anaphylaxis, although cyst rupture may be clinically silent^(9,5).

Materials & Methods

This study was done between January 2014 and June 2015, a period of 18 months.

Inclusion criteria:

1. All patients in the age group of 10 -80 yrs were included in the study.
2. All patients with previously detected Hydatid cyst either on sonography or on a radiograph.

Exclusion criteria: Out of 150 patients in the study group, 38 patients were discarded after CECT showed the diagnosis of cystic lesions like pseudo cyst of pancreas or cystadenoma of ovary. A total number of 112 patients were included in the group. CT was performed on SOMATOM EMOTION 16 SLICE (SIEMENS Ltd).

Observation & Results

In our study, the age varied from 13 yrs to 80 yrs with majority of cases in 20 -50 age group. In our study group, there was a slight female predominance with a 52 males (46. 42%) & 60 females (53.57 %).

Site of involvement

In our study, there was a wide variety of presentation with respect to site of involvement.

Table 1: Distribution of cases according to site of involvement

Site	Number of cases	Percentage
Liver	78	69.64%
Retroperitoneal	7	6.25%
Brain	6	5.35%
Intraperitoneal	3	2.67%
Lung	3	2.67%
Liver & spleen	2	1.78%
Spleen	2	1.78%
Mesentery	2	1.78%
Heart	2	1.78%
Liver & intraperitoneal	1	0.89%

Paracolic gutter	1	0.89%
Pelvis	1	0.89%
Breast	1	0.89%
Disseminated	1	0.89%
Vertebral	1	0.89%
Kidney	1	0.89%
Orbit & brain	2	1.78 %

Complications

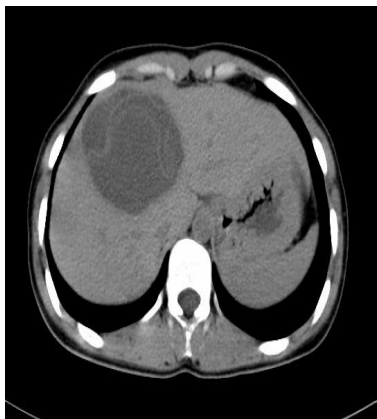
The following complications were encountered in our group of patients. A total of 18 patients (16.07 %) showed below listed complication while in 83.92 % of cases, Hydatid cyst presented as an uncomplicated cystic lesions.

Table 2: Distribution of cases according to type of complication

Complications	Number of Cases	Percentage
Contained internal rupture	3	2.6 %
External rupture	2	1.7%
Pleural effusion	5	4.46%
Ascites	2	1.7 %
Psoas muscle involvement	4	3.57%
Disseminated disease	1	0.8 %
Vascular complications	1	0.8%



Picture 1: Two cases showing presence of Hydatid cysts in lung



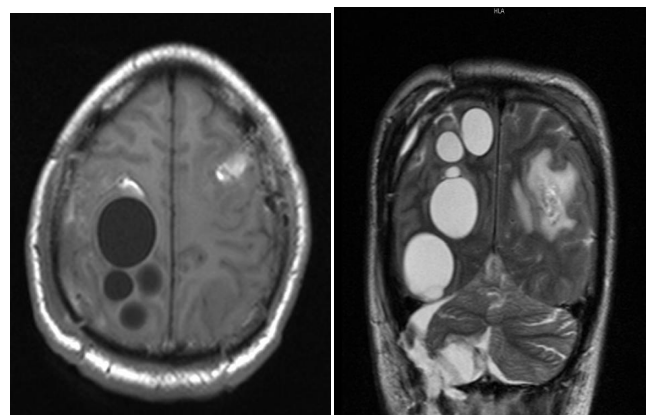
Pictures 2: case of hepatic Hydatid showing intracystic floating membrane



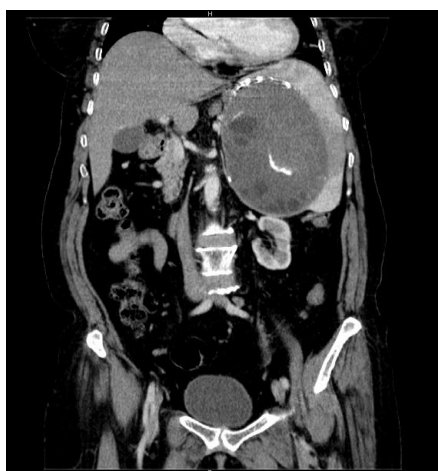
Pictures 5: case of renal Hydatid showing the presence of daughter cysts in right kidney, note the presence of enhanced renal tissue at the periphery of the cyst



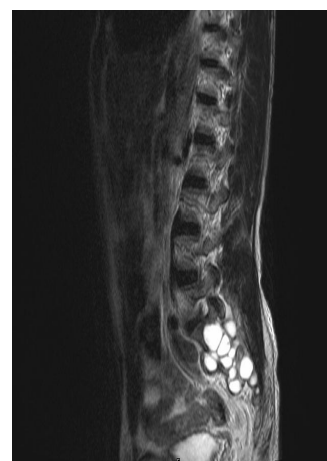
Picture 3: Presence of Hydatid cyst in the peritoneal cavity with thick rim of calcification



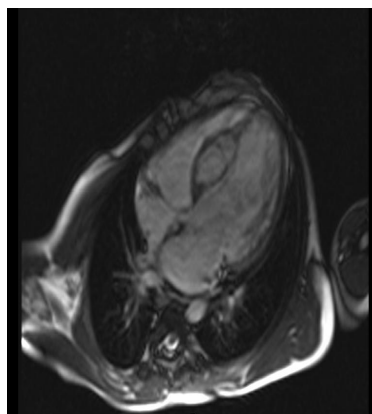
Picture 6: Fluid attenuating multi cystic lesion in posterior parietal lobe with no perilesional edema



Picture 4: Case of splenic Hydatid showing presence of daughter cyst, scattered calcifications



Picture 7: A case of multiloculated cystic lesion in lower lumbar spine proven case of Hydatid cyst with dissemination into multiple organs .



Picture 8 : A case of suspected Hydatid cyst in the interventricular septum , post operative biopsy findings consistent with the Hydatid cyst

Discussion

Various Imaging modalities are available in diagnosis of Hydatid cyst like plain radiographs of chest or abdomen, ultrasonography, CT scan & MRI. Most often, they may be asymptomatic & are diagnosed on routine screening or coexistent with some other pathology. In our study group, there was a wide variety of presentation with respect to site of involvement & nature of complications.

Liver

In adults, the liver represents the most prevalent site (50–70%) for hydatid cysts. They may cause abdominal pain, discomfort, localized swelling. Hepatic Hydatid cysts show varied appearances according to the stage of the disease. On unenhanced CT, they appear as fluid density lesions with the well defined wall with wall appearing as high attenuation structure. Wall may show focal or concentric thick calcification. Appearance of daughter cysts within the maternal cyst vary according to the stage of disease either seen in the periphery or in the entire cyst resembling a rosette. CT density of mother cyst is higher than the maternal cyst. Enhancement of the wall is a characteristic feature of infection along with air fluid levels. Patchy contrast enhancement of neighbouring liver parenchyma represents inflammatory changes. CT is superior in detecting gas or air-fluid levels within the cyst.⁽⁵⁾ Magnetic

resonance imaging (MRI) reveals a homogeneous hypointense lesion on T1-weighted images and a homogeneous hyperintense lesion on T2-weighted images. The presence of a hypointense rim at the cyst periphery has been described as a characteristic of hydatid cysts (as opposed to non-parasitic cysts), but it is non-specific. This hypointense rim may be seen in long-standing Type I cysts due to a fibrotic response of neighbouring host tissue or to slight calcifications within the cyst wall⁽⁵⁾. Sonography, CT or MRI are helpful in detecting hydatid sand which floats & settles with change in patients position. The cyst may show detached irregular laminated membrane which floats inside the cyst due to detachment of endocyst. This stage is called a transitional stage between active & inactive stage of the disease. The detachment of the membrane may due to increased intracystic pressure, host response, degeneration, trauma or following therapy .In our study ,Out of a study group of 112 patients, Liver was the most prevalent site with 69.64 % of cases .In a study done by Pinar Polat et al ⁽⁶⁾, liver was the most common site .Our percentages on site of occurrence differ slightly from their study as the sample size of our study is smaller.

Table: 3 :Classification of Hepatic Hydatid Cysts on CT

Types	Phase of disease	Features	Complications
Type 1	Initial active disease	3 layers are intact	Rupture & Dissemination
Type 2	Active phase	Hour glass appearance	Dissemination without rupture
Type 2 A	Active phase	Daughter cysts in the periphery	Rupture & Dissemination
Type 2 B	Active phase	Daughter cysts in the entire cyst like rosette	Rupture & Dissemination
Type 2 C	Active phase	2A+2B+calcification in wall or in daughter cysts	Rupture & Dissemination
Type 3	Inactive or dead	Either wall calcification , calcification of membranes or entire cyst calcification	None
Type 4	Complicated Hydatid cysts	Enhancement of wall,rupture, mass effect,	Infection, rupture, dissemination

Lung

It is most common site to be affected in children & second most common site to be affected in adults after liver .The mode of spread is usually hematogenous however can occur due to

transdiaphragmatic spread from other organs or due to rupture from other organs commonly from liver. Pulmonary Hydatid cysts show a predilection to posterior segments & lower lobes. They are often multiple & bilateral and usually reach a very large size in lungs. Commonly, they are type 1 or type 2 cysts. CT shows fluid density hypo attenuating, well defined, oval to round cystic lesions. Various presentations of pulmonary hydatid cyst⁽⁶⁾ are 1. Thin walled translucent cavitating lesions containing air .2.Meniscus sign: large cysts produces erosions into the bronchioles included in the pericyst resulting in presence of air between pericyst & laminated membrane 3. Onion peel sign: when air is seen between pericyst & endocyst.4 .Water lily sign: detachment of the endocyst resulting in a freely floating membrane. The most common complications encountered in pulmonary hydatid cyst are rupture into parenchyma or pleura, spread to adjacent organs, consolidation, empyema, bronchial fistula & rarely pulmonary embolism. In our study group 2. 67 % of cases showed involvement of lung.

Brain

Hydatid cysts in brain are extremely rare, occur in upto 2 % of cases .They are more common in children & can occur anywhere in the brain but most of them are supratentorial & commonly seen in the territory of middle cerebral artery & in parietal lobe. The usual appearance of Hydatid cyst is either as unilocular or multilocular fluid attenuating lesion on unenhanced CT. Characteristic features of cerebral Hydatid cyst are lack of perilesional edema & presence of significant mass effect. Arachnoid cyst, epidermoid tumor, porencephalic cyst, brain abscess, cystic astrocytoma are the lesions which should be considered in the differential diagnosis. They can invade the meninges, can cause calvarial erosions or rupture into subarachnoid spaces .In our study group, 5. 35 % of cases showed involvement of brain.

Orbit

Orbital hydatid cysts are extremely unusual & represent 1- 2 % of cases. they usually present as unilateral proptosis, decreased visual acuity, exophthalmous, impaired ocular movements pain & swelling. They are usually unilateral & solitary. They can occur either as a primary disease or as secondary to hydatid disease elsewhere in the body .An orbital hydatid cyst tends to involve the retro bulbar tissues either within the muscle cone or outside in the superolateral or superomedial angle⁽⁸⁾. Inferiorly located cysts are extremely rare. Differential diagnosis includes other ocular and orbital diseases such as retinoblastoma, rhabdomyosarcoma, inflammatory pseudotumors, capillary hemangioma, chloroma, glioma, lymphangioma etc. In our study group 1. 78 % of cases showed involvement of orbits secondary to involvement of brain. There were no cases in isolated primary involvement of orbit.

Heart

Hydatid cysts are very unusual in heart representing 0.5 -2 % of cases ⁽¹⁵⁾ and maybe due to hematogenous spread or rupture of lung Hydatid cyst. The most commonly affected cardiac chambers (in decreasing order of frequency) are the left ventricle (50%–60% of cases), interventricular septum (10%–20%), right ventricle (5%–15%), pericardium (10%–15%), and right or left atrium (5%–8%) (6).Cardiac MRI is the investigation of choice as motion artefacts are commonly decrease the efficacy of CT scan. According to the literature, Common sites involved in heart are pericardium, interventricular septum, costophrenic angles, superior mediastinum & hila. Type 1 to Type 3 cysts are commonly seen. On chest radiographs, deformation of cardiac borders or lung contours may be seen. Chest radiograph, Transthoracic echocardiography, CT, MRI are the common imaging modalities used in the detection. In our study group, 1. 78% of cases showed involvement of orbits secondary to involvement of brain. There were no cases in isolated primary involvement of orbit.

Spleen

The commonest mode of Mode of transmission of disease is by spread from adjacent organs like liver, peritoneum etc or as a part of systemic dissemination. Primary splenic involvement is very rare less than 2%⁽⁵⁾. Splenic Hydatid cyst is almost always solitary & imaging features are similar to their hepatic counterparts of all stages. Other splenic cystic lesions such as epidermoid cyst, pseudocyst, splenic abscess, hematoma and cystic neoplasm of the spleen should be considered in the differential diagnosis⁽⁵⁾ In our study group, 1.78 % of cases showed involvement of spleen secondary to involvement of liver. There were 1.78 % of cases in isolated primary involvement of spleen.

Kidney

Renal Hydatid cysts are very rare, 3 % of cases^(6,5) They present with dysuria, flank pain, hematuria, persistent fever, renal colic, hypertension, renal calculi⁽¹⁰⁾ They are usually located in upper or lower poles. They are often solitary, generally seen in the renal cortex. 18 % of renal hydatid cysts can rupture into collecting system & produce hydatiduria^(11,6). Imaging features are similar to Hepatic Hydatid cysts of all stages. Mural calcification and daughter cysts often coexist^(5,6). Differential diagnosis include simple renal cyst, necrotic renal cell carcinoma, renal abscess and infected cysts & necrotic renal cell carcinoma. On excretory urogram, it may cause bulging of renal outline or may appear as round mass which elongates the infundibula and calyces^(11,6). On plain CT scans, commonest presentation is that they are seen as hypoattenuating masses with daughter cysts. In our study group, 0.89 % of cases (1 case) showed isolated involvement of kidney.

Peritoneal Cavity

13 % of cases present in peritoneal cavity. It is always secondary to traumatic or surgical rupture of hepatic, mesenteric or splenic hydatid cyst⁽⁵⁾ but sometimes can present as a primary disease.

They are often multiple, can present as any type of hydatid cysts & can arise anywhere in the peritoneal cavity. CT & MRI help in accurate diagnosis of peritoneal Hydatid cyst. They need to be differentiated from mesenteric cysts, duplication cysts or cystadenoma of ovaries. In our study group, 2.67 % of cases showed isolated involvement of peritoneum & 0.89 % of cases showed peritoneal involvement secondary to involvement of liver.

Retro Peritoneum

It is always secondary to traumatic or surgical rupture of hepatic, mesenteric or splenic hydatid cyst⁽⁵⁾ but sometimes can present as a primary disease. Primary retroperitoneal hydatid cyst is extremely rare and only occasional cases have appeared since Lockhart and Sapinza first described this entity in 1958. It is defined as a zone of hydatidosis occurring in the fatty tissue in the space lying behind the posterior parietal peritoneum without any parasitic foci in other organs⁽¹⁴⁾ It usually remains asymptomatic unless the cyst grows and produces symptoms due to pressure, rupture into the pleural or peritoneal cavity, secondary infection, or an allergic reaction^(14,12) An isolated retroperitoneal hydatid cyst could be caused by haematogenous dissemination of protoscoleces after bypassing the liver and the lungs or by the gastrointestinal tract into the lymphatic system⁽¹³⁾ The differential diagnosis of retroperitoneal cysts also includes soft tissue tumors, retroperitoneal abscess, cystic lymphangioma, embryonal cyst, ovarian neoplasms, teratoma, and other cystic and necrotic solid tumors⁽¹²⁾. It can also rupture into great vessels like the aorta forming false aneurysms or may present as retrocaval ureter⁽¹⁴⁾. In our study group, 6.25 % of cases showed involvement of retro peritoneum.

Spinal Cord

They account for less than 1 % of cases. They are typically multiple & often do not show calcifications. They are more common in thoracic spine

but can occur in any part of the spine. Spinal Hydatid cysts are classified into five groups: intramedullary, intradural extramedullary, extradural intraspinal, vertebral, and paravertebral⁽⁶⁾. CT & MRI help in accurate diagnosis of Hydatid cyst in these locations. Tuberculosis spondylitis or chronic osteomyelitis should be considered in differential diagnosis. Lack of osteoporosis and sclerosis in involved bone, absence of damage to intervertebral disk spaces and vertebral bodies, paraspinal extension, and (in the thoracic spine) involvement of contiguous rib are the most common features of spinal Hydatid cyst⁽⁶⁾. In our study group, we have one case of vertebral involvement secondary to peritoneal involvement & another one case as a part of disseminated disease involving multiple organs including spine.

Vascular Complications

Include rupture of aorta & encasement of adjacent vessels to the lesion. There pericyst gets adherent to the adjacent vessels especially in large peritoneal lesions. It is mandatory to look for clear zone of separation from adjacent vessels on all imaging modalities. CT is superior to others. Failure to recognize this complication may lead to peritonitis, rupture, dissemination of disease or it can lead to vascular injury during surgery which may prove fatal. In our study, we report a case of secondary retroperitoneal hydatid cyst presenting as iliopsoas abscess adherent to the iliac & femoral vessels. During surgical excision there was an iatrogenic traumatic rupture of femoral vessel which was later repaired.

Bone

Seen in less than 2 % of cases. The most commonly involved bone structures are the spine (35% of cases), pelvis (21%), femur (16%), tibia (10%), ribs (6%), skull (4%), scapula (4%), humerus (2%), and fibula (2%). CT & MRI help in accurate diagnosis of Hydatid cyst in these locations. These cysts show much thinner walls, rarely calcify & are generally irregular in shape.

Extensive involvement can lead to destruction of bone or may lead to pathological fractures & may spread to extraosseous soft tissues. Parasitized bone has heterogeneous medium to low signal intensity on T1-weighted MR images and high signal intensity on T2-weighted images. CT can demonstrate the calcification in extraosseous Hydatid cyst⁽⁶⁾.

Other unusual locations reported in literature include omentum, soft tissues, ovary, pancreas, thyroid, adrenal glands, scrotum, mediastinum & pleural cavity.

Conclusions

Hydatid Cyst should be kept in mind as a possible differential diagnosis in all cystic lesions occurring in any organ in the body. Complications due to Hydatid Cyst are many & varied in nature. Hence a systematic approach to Imaging & careful analysis of possible complications is mandatory to arrive at an accurate diagnosis.

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