Echinococcosis still a serious cause of morbidity among children. Spectrum of imaging findings

Poster No.: C-2546
Congress: ECR 2017
Type: Educational Exhibit
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Keywords: Parasites, Infection, Cysts, Surgery, Ultrasound, CT, Lung, Liver, Abdomen
DOI: 10.1594/ecr2017/C-2546

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Learning objectives

Hydatid disease (HD) is a unique parasitic disease, which is endemic in different regions of the world, including the Mediterranean region. There is just few publications about radiologic findings of HD in children.

Our aim in this article is:

1-To introduce the different types of hydatid disease presented either as simple or as complicated disease, as well, as unusual cases.

2- To evaluate the imaging findings which help in the diagnosis of HD using US and/or CT as imaging modalities.
Background

HD continuous to be a serious health problem in our country with approximately 22 pts/year affected posing immediate need for prevention program implementation. It is caused by the larval stage of Echinococcus tapeworm. There are known four types of Echinococcus but only two types are of medical importance in humans; E.granulosus causing Cystic Echinococcosis or HD and E. multilocularis causing alveolar Echinococcosis.

Mediterranean region, which our country is part of, is endemic for HD mainly due to spread sheep and cattle raising areas. The primary hosts are dogs while sheep or cows become intermediate hosts. Humans are infected by consuming contaminated food with dogs’ stool.

Theoretically it may appear in every part of the body with liver being the most affected organ in adults followed by lung and less commonly heart, brain, bone, kidneys, adrenal glands etc.

We do not possess specific data for the incidence of HD in children, however it is believed that they are less commonly affected even though there are studies which report same incidence of HD in both children and adult patients. A recent study in our country concludes that HD may affect every active - age group.

Our hospital is a referral center for paediatric patients with HD.

Diagnosis is based on the correlation of clinical, radiologic and serologic findings. Patients are diagnosed either incidentally or after presenting with symptoms related to associated complications. Radiologic findings are variable depending on the stage of growth, associated complications and the affected organ. Serologic tests based on the detection of serum antibodies are necessary to establish the final diagnosis with a sensitivity range that varies between 85-98% for liver HD, 50-65% for lung and 90-100% for multiple organ cysts even though they remain of only a confirmatory use because of the variability in specificity. This is especially true with type I and type V cysts.  

Surgery with complete excision of the cyst and maximum preservation of the healthy tissue is considered as the mainstay approach for treatment. Many articles have been reported about HD and imaging findings that help in its diagnosis but few of them have been focused on the HD paediatric patients or introduced complicated and unusual cases in this age group.
Findings and procedure details

It was a single centred study which included 11 patients (pts) aged between 2-14 years. (table 1). It was previously believed that paediatric patients are less commonly affected than adults may be due to less studies including this age group. However recent articles report high incidence of HC even in children. In children HC appears more frequently in the lungs tending also to be bigger in size due to the low resistance of lung tissue which provides more rapid cystic growth.

Clinically HD may be asymptomatic and diagnosed incidentally during an examination for other reasons, or present with symptoms related to compression of the organ affected as well as related complications such as infection, membrane rupture, fistulisation etc.

<table>
<thead>
<tr>
<th>Age group (years old)</th>
<th>Organ affected</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Liver</td>
</tr>
<tr>
<td>0-2</td>
<td>-</td>
</tr>
<tr>
<td>3-6</td>
<td>-</td>
</tr>
<tr>
<td>7-10</td>
<td>2</td>
</tr>
<tr>
<td>11-14</td>
<td>-</td>
</tr>
</tbody>
</table>

Table 1.

Radiologic findings

Radiologic modalities play a crucial role not only in the diagnosis, staging and follow up but also in planning the appropriate treatment approach of patients.

Abdominal and Chest X-ray:

Usually give nonspecific information about HD. It may reveal a round/oval opacity I the lung with or without fluid air level depending on the cyst growth stage. Sometimes in calcified cysts it can visualize the ring-like calcifications in the periphery of the cyst raising doubt for a HC.
Ultrasound:

Is excellent for the evaluation of abdominal HC giving useful details about the clarity of fluid content with higher sensitivity for presence of debris (sand). US can also visualize septa when they are present and the presence of daughter cysts and membrane ruptures. Depending on these findings HC are classified according to a standardised WHO classification which is based in Gharbi et al classification of 1981. (table 2).

US poses little or no value in HD in the lungs, unless a peripheral localization of the HD or ruptured in the pleural space.

CT and MRI give better information for cases not accessible by ultrasound. Except for giving detailed cyst location they can also better evaluate patients with complicated cysts, broncho/biliary fistulas and calcifications which appear as round ring like hyperattenuating areas in CT and hypointense at MRI. CT is the preferred modality before planning for surgery.

Considering lack of radiation, MRI might be preferred to CT-scan when evaluating HC in children. Nevertheless, not only is not MRI cost effective but it also adds little information to CT-scan making the latter the best choice for examination.

In selected cases MRCP is used to better evaluate the possible communication of the cyst with biliary tree. It can demonstrate in more details (compared to CT) the level and degree of communication.

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Simple cyst with pure fluid collection</td>
</tr>
<tr>
<td></td>
<td>Normal 0 false false false HR X-NONE X-NONE</td>
</tr>
<tr>
<td>II</td>
<td>A fluid collection with a detached membrane</td>
</tr>
<tr>
<td></td>
<td>Normal 0 false false false HR X-NONE X-NONE</td>
</tr>
<tr>
<td>III</td>
<td>Fluid collection with multiple septa and/or daughter cysts</td>
</tr>
<tr>
<td></td>
<td>Normal 0 false false false HR X-NONE X-NONE</td>
</tr>
<tr>
<td>IV</td>
<td>Hyperechoic with high internal echoes</td>
</tr>
<tr>
<td></td>
<td>Normal 0 false false false HR X-NONE X-NONE</td>
</tr>
</tbody>
</table>
Table 2.

Type I hydatid cysts (type I HC) (fig.1, 2 & 3) are simple cysts with clear fluid inside and thin walls, which size may vary from a few millimeters to more than 10 centimeters. In certain cases these type of cysts might have a septate texture and/or sand. These type of cysts are thought to constitute the first stage of the disease.

In liver the cysts usually occupy the right lobe. US constitutes in most cases the initial imaging method of examination, with HC being found incidentally or in cases of examination due to clinical findings related to HC's complications. US findings show an anechoic or hypoechoic well-defined cystic lesion, with perceptible walls in most cases, which help in the differential diagnosis with other benign cystic structure in the liver, like the simple biliary liver cysts. In addition, US imaging offers a chance for dynamic examination of the cysts with echogenic sandy content. So, asking the patient to turn from supine to the side, can show live the echogenic corpuscular foci dissipate within the cyst, which constitutes another helpful imaging finding for HD.

These type of cysts may appear in the lungs, in any lobe, uni or bilaterally. As, X-ray in pediatric patients constitutes the first step of imaging modalities used in lung disease exploration, the imaging findings in type 1 cysts are rather non-specific, showing a quite well-defined opacity within the lung parenchyma, and it requires further exploration.

CT constitutes the next imaging modality of choice in most cases, with the main disadvantage being radiation in these age-group. Main findings include a well-defined hypodense-cystic mass, with near water HU density, with perceptible walls and in some cases septated structure. When in the liver, wall and septae enhancement are helpful clues in differentiating type I HC from simple liver cysts, with MRI constituting the next imaging modality when a more thorough differential diagnosis is needful, adding some more specific imaging findings, like "hypointense rim-sign" in T2W images.(8)

Type II hydatid cysts (type II HC), (fig.4) basically constitutes cystic structure with one/more multiple daughter cysts and matrix within it, thus showing quite variable macroscopic presentations. In some cases a "spoke-wheel" cystic structure might be seen in US, in cases of multiple daughter cysts within the same capsule of the mother cyst, containing/not echogenic areas. In some cases, type II HC might present with solid areas in US, which might make the differential diagnosis more challenging. These solid areas are thought to be due to consolidation of any of the daughter cysts.
The imaging findings in US, X-ray on one side and CT and MRI are quite correlative. In addition, CT findings are helpful in estimating the possible stage of the HC.\(^{(1, 8)}\)

**Stage IIA HC** findings include a well-defined cystic structure, with daughter cysts-regular round/oval cysts-arranged at the periphery of the mother cysts. In both stages IIA and IIB, one common finding is that the mother cyst content has a higher HU density comparing to the daughter cysts. Thus in Stage IIB HC, even though the daughter cysts-rather irregular shaped cystic structures in these stage of HC-are scattered throughout the whole volume of the mother cysts, due to the high attenuation of the mother cyst content, it gives stage IIB HC a rather septate cystic structure-much different from stage IIA HC-and many times compared to a "rosette."

While, in **stage IIC HC** the imaging findings in CT correlate to the stage of the cyst, which is that of an old degenerated cyst with an amorphous content. Thus, findings include a well-defined oval/round lesion, with a higher-attenuation content with dispersed calcifications. In some cases daughter cysts might be found even in this stage of HC.

**Type III hydatid cysts (type III HC)** findings are less variable, showing the remnant of a calcified dead cyst. Thus type III HC a strong posterior shadow in US and high-calcium-attenuation in CT, and hypointense in both T2/T1W images in MRI.\(^{(1, 8)}\)

**Type IV complicated hydatid cysts (type IV HC)** (fig. IV) findings are quite variable, depending on the complication. HC-both type I and II-might rupture and then may or may not become superinfected. Aging degeneration of the parasitic membranes, different chemical reactions and individual immunity reactions are thought to be some of the causes of HC rupture. The rupture in turn might be contained, communicating or direct, showing different imaging findings in each case correlating with the rupture mechanism and/or to where the cysts ruptures.

In contained ruptures, the rather specific sign is the wavelike undulating membrane, due to a disruption of the endocyst from the pericyst. US might show, as well, what is called a "snowstorm" image and it has the advantage of demonstrating the dynamic movement of the membrane with patient rolling.

In communicating ruptured HC, US and CT findings show well-defined lesion, with respectively echogenic and higher-attenuation content compared to contained ruptured HC, with CT having a higher sensitivity to showing the possible communicative fissure. The HC might rupture in the biliary tree, showing the sandy content spread throughout the affected biliary tree. In other cases a direct rupture to pleural or peritoneal cavity might be found.

In all cases, a ruptured HC might (25% of cases) or might not (75%) be superinfected, with imaging findings including heterogenous echogenicity, fluid-fluid levels, poor borders, etc.
While, air-fluid levels might be a finding of both superinfection and rupture—both direct and communicating. (1,8)

In children HC appears most commonly in lung where it may present in all different stages, uni or bilaterally, in small or huge dimensions. A rare imaging finding in this age-group is the cavitary-like appearance of the cyst which might be a certain stage of the natural growth of the cyst or in other cases, due to antiparasitic therapy (fig.6). In such cases differential diagnosis is challenging especially when the patient presents in this stage growth of the cyst and when it is a solitary lesion.

On the other hand heart HC is an unusual appearance with an incidence of 0.5-2% (13). When present they may predispose the patient to serious complications that can be lethal. Thus periodic check-up in endemic areas and in patients with medical history for HC, may prevent such complications. (fig.5)

Treatment of HC is case specific and depends on the cyst dimensions and characteristics, available medications and surgical expertise in referral areas. However different studies report complete excision of the cyst as the appropriate approach especially in endemic areas. In our country the surgical approach is the mainstay of treatment followed by three months of antiparasitic therapy and follow up with Chest X-ray and abdominal US. In bilateral lung cyst a staged surgical approach is applied.

Normal 0 false false false HR X-NONE X-NONE
Fig. 1: 6 year old patient (yo pt) presenting with abdominal pain on the right hypochondrium. CT-scan without contrast media shows a huge thin wall cyst with a diameter > 13 cm that extends in the right lobe of the liver. CECT (not shown) did not reveal any septations or solid nodules within the cyst. Serology was positive for echinococcosis and histopathology confirmed the diagnosis.

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Fig. 2: HC type I. 8 yo, male pt with abdominal pain. US (not shown) showed two cystic lesions in the right lobe of the liver. CECT-scan revealed two cystic lesion in the VII-th and V-VI segments respectively with thin walls and no pathologic inside enhancements. Serologic tests and histopathology were positive for echinococcosis.

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**Fig. 4:** Type II HC. 6 yo, female pt presented with cough and shortness of breath. CECT-scan shows a lesion in the lower left lobe of the lung measuring 3.2 x 2.5 cm, with thin walls and linear enhancements inside the lesion consistent with floating membranes within the pericyst. NOTE small parenchimal consolidations adjacent to the lesion in

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**Fig. 5:** 10 yo, male patient presenting with shortness of breath. NECT-scan and MRI (not shown) shows a cystic lesion on the right middle lobe with thin walls and no internal pathologic enhancements, consistent with a simple cyst. Another cystic lesion is seen within the left ventricle of the heart with thin walls but with thin ring-like calcifications consistent with a HC type III. Successful surgery resection was made for both cysts in staged procedures removing firstly the heart cyst.

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Fig. 6: Type II and type IV HC. 3 yo, male pt presenting with cough and temperature. CT-scan shows bilateral cystic lesions, one on the upper right lobe with floating membranes within the pericyst (water-lily sign)- fig. d) and the other on the upper left lobe. a) first day of examination: the cyst on the left lung shows air-fluid level and small parenchimal consolidations around it suggesting superinfection of the cyst. c) CECT obtained two days after the first CT scan: NOTE the cyst on the left upper lobe appears as a cavitary lesion with air content which is a rare finding of lung HC. During these days the pt was only under antibiotic therapy. The diagnosis of HC was confirmed after surgery.

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Fig. 7: Concomitant liver and bilateral lung HC. 8 yo pt with cough and temperature a) and b) Type IV HC. CT scan shows hypodense lesion in the right upper lobe with fluid HU values, floating membranes inside it and small air bulae inside consisting with superinfection of the cyst. NOTE the parenchymal consolidations adjacent to it. c) Type I HC. CT scan shows round hypodense lesion in the left lower lobe with fluid HU values inside and thin walls d) Small cystic lesion in the right lobe of the liver which showed reduction in dimensions after antiparasitic therapy confirming its hydatid nature.

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Fig. 3: Type I HC. 8 yo, male pt presenting with cough, fever and shortness of breath. CECT-scan shows a hypoattenuating lesion on the right upper lobe of the lung with thin walls and fluid HU density. Considering the central position of the lesion differential diagnosis with a bronchogenic cyst was reported on the medical report. Serologic tests were positive for HC.

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Conclusion

HD may present with a variable radiologic spectrum depending on the cyst stage, the organ affected and associated complications. Detailed knowledge of these findings is crucial for radiologists to primarily suspect the disease/contribute in the diagnosis of the disease and to provide the relevant information to the surgeon, especially in complicated cases. Paediatric patients are a specific age group which is affected by HD as well as adults. More studies are necessary to determine if there are age-related radiologic findings and treatment in order to be included in the approach guidelines.
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