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Radiographic Characteristic of Hepatic Hydatid Disease

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Abstract:

Hydatid disease primarily affects the liver and typically demonstrates characteristic imaging findings. However, there are many potential local complications (eg, intrahepatic complications, exophytic growth, transdiaphragmatic thoracic involvement, perforation into hollow viscera, peritoneal seeding, biliary communication, portal vein involvement, abdominal wall invasion). Furthermore, secondary involvement due to hematogenous dissemination may be seen in almost any anatomic location (eg, lung, kidney, spleen, bone, brain). Ultrasonography (US) is particularly useful for the detection of cystic membranes, septa, and hydatid sand. Computed tomography (CT) best demonstrates cyst wall calcification and cyst infection. CT and magnetic resonance (MR) imaging may demonstrate cyst wall defects as well as the passage of contents through a defect. Chest radiography, US, CT, and MR imaging are all useful in depicting transdiaphragmatic migration of hydatid disease. CT is the modality of choice in peritoneal seeding. US and CT demonstrate rupture in most cases that involve wide communication. Indirect signs of biliary communication include increased echogenicity at US and fluid levels and signal intensity changes at MR imaging. CT allows precise assessment of osseous lesions, whereas MR imaging is superior in demonstrating neural involvement. Familiarity with atypical manifestations of hydatid disease may be helpful in making a prompt, accurate diagnosis.

Key words: Liver, Hydatid disease, US, CT, and MR.

Introduction:

Hydatid disease is a worldwide zoonosis caused by the larval stage of the echinococcus tapeworm, that is endemic in many parts of the world (in European, Middle Eastern, Mediterranean, South American and African countries)[1-4].

There are two types of Echinococcus infections: Echinococcus granulosis, the more common type, and Echinococcus multilocularis, the less common but more invasive.

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Hydatid disease is a relevant health problem in underdeveloped areas where veterinary control does not exist. The most frequent location of hydatid cystic lesions is in the liver (up to 80% of cases), followed by the lung (about 20% of cases), and with a lower reported incidence in any other organ or tissue in the body[2-4].

Dogs or other carnivores are definitive hosts, whereas sheep or other ruminants are intermediate hosts. Humans are secondarily infected by the ingestion of food or water contaminated by dog feces containing the eggs of the parasite. After the ingestion of the eggs, the freed embryo 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 grow into hydatid cysts. Some embryos pass through the hepatic capillaries and become lodged in the lungs and other organs.

The definitive diagnosis of liver echinococcosis requires a combination of imaging, serologic, and immunologic studies[4].

At biochemical analysis, there is usually eosinophilia, and a serologic test is positive in 25% of patients[5]. At histopathologic analysis, a hydatid cyst is composed of three layers: the outer pericyst, which corresponds to compressed liver tissue; the endocyst, an inner germinal layer; and the ectocyst, a translucent thin interleaved membrane[5].

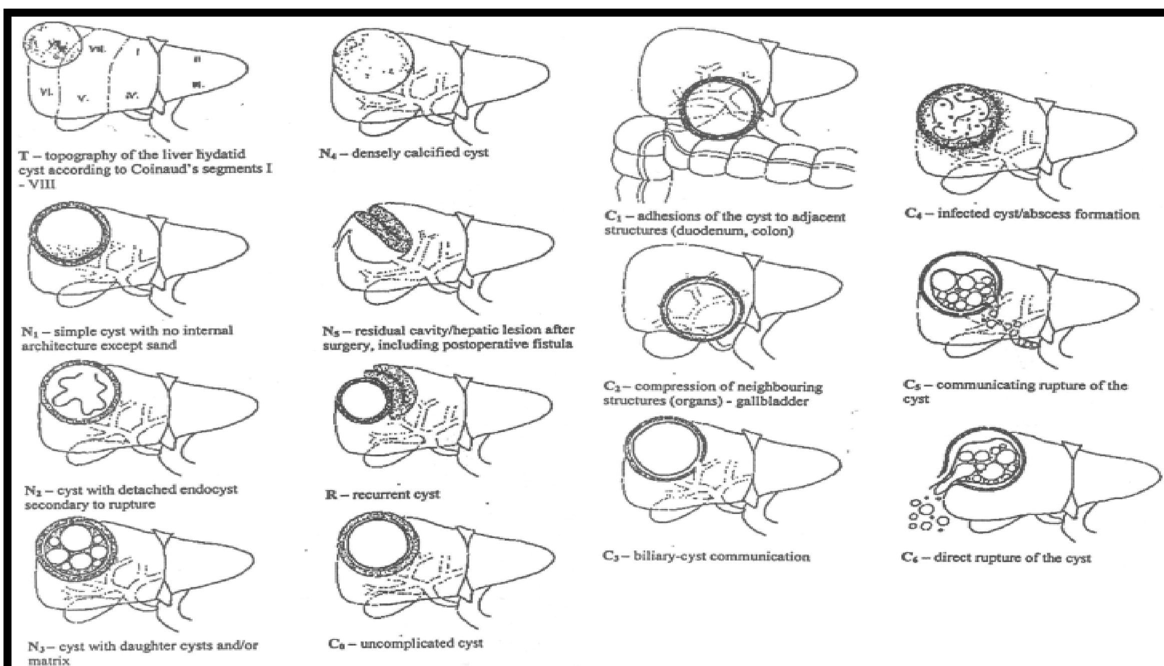
Imaging procedures are essential in diagnosis and evaluation of the extent of liver hydatidosis; ultrasound (US), computed tomography (CT), and magnetic resonance (MR) can depict hydathid disease[2-4,6].

The imaging method used depends on the involved organ, and the radiologic findings range from purely cystic lesions to a completely solid appearance[3]. US is the screening method of choice and is also used to monitor efficacy of medical therapy[2-4,6]. CT is always performed because it has a high sensitivity (94%)[7]. It is an important preoperative diagnostic tool to determine vascular, biliary or extrahepatic extension, to recognize complications, such as rupture and infections, and therefore to assess respectability[8-10]. MR is the best imaging procedure to demonstrate a cystic component. It helps to determine vascular or biliary tree involvement, as well as extrahepatic extension[10,11, 12].

There are many potential complications such as exophytic growth, transdiaphragmatic thoracic involvement, peritoneal seeding, biliary communication, portal vein involvement, abdominal wall invasion and hematogenous dissemination in any anatomic location (lung, kidney, spleen, bone, brain)[6,10,11, 12].

Life Cycle of E. granulosus

Humans become infected by ingesting eggs from the tapeworm E. granulosus either by eating contaminated food or through contact with dogs. The ingested embryos invade the intestinal mucosal wall, enter the portal circulation, and develop into a cyst in the liver. Humans are intermediate hosts. In the liver, cysts grow to 1 cm during the first six months and approximately 2–3 cm annually thereafter, depending on the host tissue resistance. [13-15].



the inner germinal layer is thin. Scolices, the infectious embryogenic tapeworms, develop from an outpouching of the germinal layer.

The middle laminated membrane and the germinal layer form the true cyst wall, usually referred to as endocyst; the acellular laminated membrane is occasionally referred to as the ectocyst. The thicknesses of these layers depend on the tissue in which the cyst is

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located. The layers tend to be thick in the liver, less well developed in muscle, absent in bone, and sometimes visible in the brain [16,17].

Imaging of hydatid disease:

The imaging methods used for diagnosis and evaluation of the extent of HD are ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI), and less commonly radiography and urography. USG is screening modality of choice and is also used to monitor the efficacy of treatment. It clearly demonstrates the hydatid sand, floating membranes, daughter cysts, and vesicles inside the cyst. CT has high sensitivity and specificity for HD. CT is an important diagnostic modality in detecting cyst wall or septal calcification, demonstrating internal cystic structure posterior to calcification, assessing complications, depicting osseous lesions and in cases where USG has limitations (obesity, excessive bowel gases, abdominal wall deformities, and previous surgery). MRI is superior for demonstrating cyst wall defect, biliary communication, and neural involvement.[17,18] Recently, MRI has been shown to be important in differentiating liver hydatid cysts from other simple cysts using diffusion-weighted sequence.[19].

The imaging findings depend on the organ involved, host reaction, stage of evolution, and maturity of disease. The findings can range from purely cystic lesions to solid-appearing masses. The cysts may be solitary or multiple, unilocular or multivesicular, and with or without calcification. Presence of daughter vesicles and membranes within the cyst, peripheral cyst wall, or internal matrix calcification are important findings for differential diagnosis of HD. Various classifications are being used to describe hydatid cysts. More commonly, depending on the imaging appearance the hydatid cysts are classified into four types. [20,21].

Type I: Simple cyst with no architecture: This type constitutes the initial and active phase of hydatid disease in which the three layers are intact [9]. During the initial stage of development of the hydatid cyst, it appears as a well-circumscribed, unilocular cystic lesion with no internal architecture, with or without internal septations or hydatid sand, with frequent enhancement of the cyst wall and septa on post-contrast CT [22]. On magnetic resonance imaging (MRI), hydatid cysts appear as a low signal intensity on T1-weighted images (T1WIs), and a high signal intensity on T2-weighted images (T2WIs) with a low signal intensity rim “rim sign”. The DWI sequence can differentiate Type I hydatid cysts from simple liver cysts [13].

Fig.2. Show the CT images of a 40-year-old male showing a well-circumscribed, round, unilocular hypodense cystic lesion centered in segment IV of the right hepatic lobe

Type II: Cyst with daughter cysts and matrix: This type represents the active phase in the parasite life cycle and in the dissemination of hydatid disease [23]. Cysts with multiple septa representing the walls of the daughter cysts inside the mother cyst are usually arranged in the periphery. According to the maturity and arrangement of the daughter cyst, Type II can appear as follows: (1) Type IIA contains multiple daughter cysts arranged at the periphery of the mother cyst with a high density central matrix forming a “wheel spoke appearance”, (2) Type IIB contains multiple irregular daughter cysts occupying the cyst forming "rosette appearance", and (3) Type IIC contains a hyperdense matrix and occasionally calcification or daughter cysts [12]. On an MRI, daughter cysts appear on a low signal intensity or isointense relative to the matrix on both T1WIs and T2WIs.

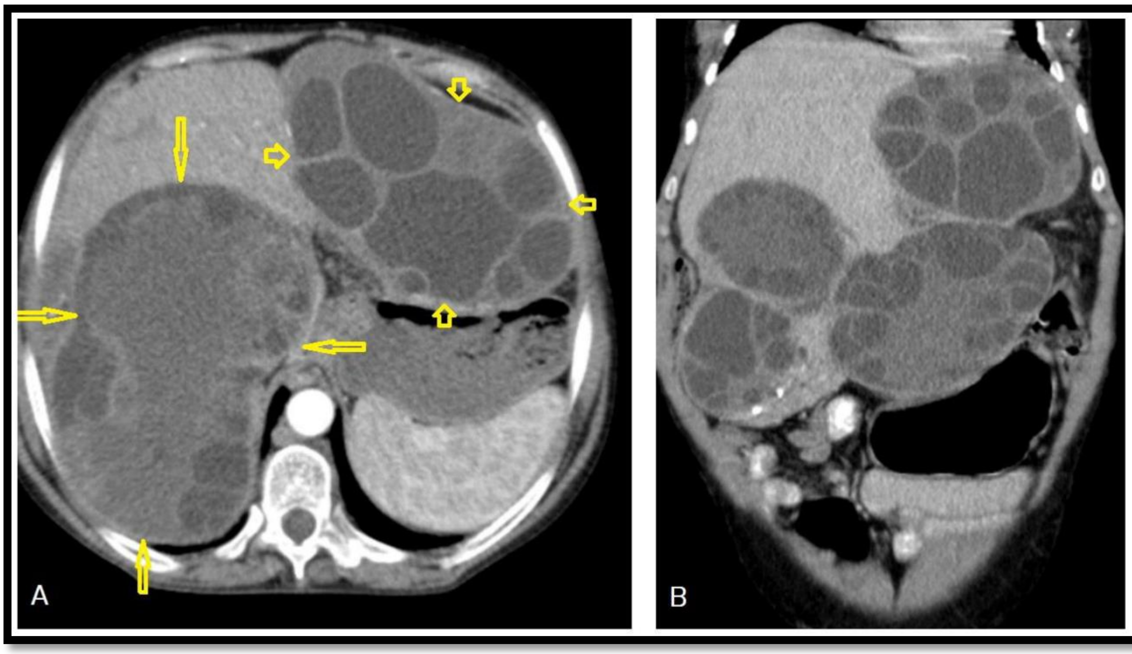

Fig.3. Show the axial (A) and coronal (B) CT images of a 55-year-old male showing multiple well-defined cystic lesions occupying the liver:

Type III: Calcified cysts: This phase constitutes the inactive dead phase of hydatid disease, which cannot spread, has no mass effect or complications, and does not require surgery [**9**]. It appears as a calcified lesion with posterior acoustic shadowing on ultrasound imaging, a round hyperdense lesion on CT, and a low signal intensity area on an MRI [**23**]. CT is the gold standard imaging modality used to diagnose calcified hydatid cysts [**24**].

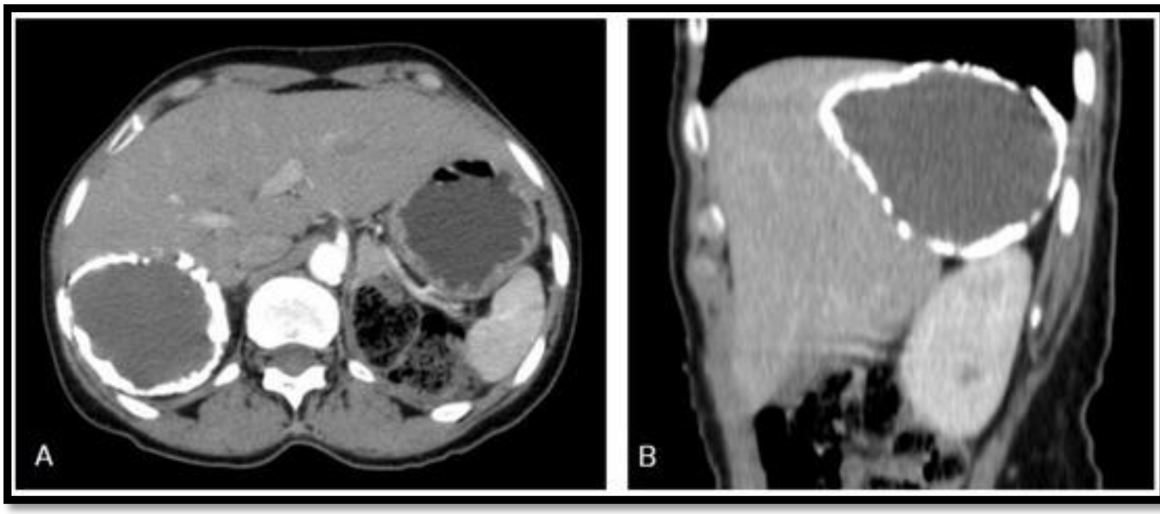

Fig.4. Show the axial (A) and sagittal (B) CT images of a 56-year-old female presenting with right upper quadrant pain, showing a well-circumscribed unilocular cystic.

Type IV: Complicated hydatid cyst: This is a hydatid cyst with a rupture or superinfection, which may be seen in both Types I and II [**13**]. CTs and MRIs play a major role in diagnosing complicated hydatid cysts [22-26]. The rupture of hydatid cysts occurs in 50% of cases, mainly due to age degeneration of the parasitic membrane or a defense mechanism [**13**]. Complications of hydatid cysts include the following: (1) Mass effect: This is when a cyst reaches a large size, which can cause biliary duct dilatation either by compression of the nearby duct or by perforation into the biliary duct. (2) Rupture of hydatid cyst: A rupture may be internal, communicating with the passage of the cystic contents into the biliary ducts, or direct, when cystic content spillage into the peritoneal cavity causes disseminated disease. (3) Hydatid disease infection is generally seen in ruptured hydatid cysts, which permits bacteria to pass easily into the cyst. Air within the cyst cavity is a clue, as it is thick and enhances walls after contrast administration on CTs and MRIs. (4) Exophytic growth occurs via the bare area of the liver with transdiaphragmatic migration to the lung or mediastinum, or via the gastrohepatic ligament into the peritoneal cavity. (5) Peritoneal seeding is almost always secondary to hepatic hydatid disease. It occurs due to previous hepatic hydatid surgery or

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after a spontaneous or traumatic rupture. CTs and MRIs are valuable imaging modalities for diagnosing peritoneal hydatid cysts [**27**].

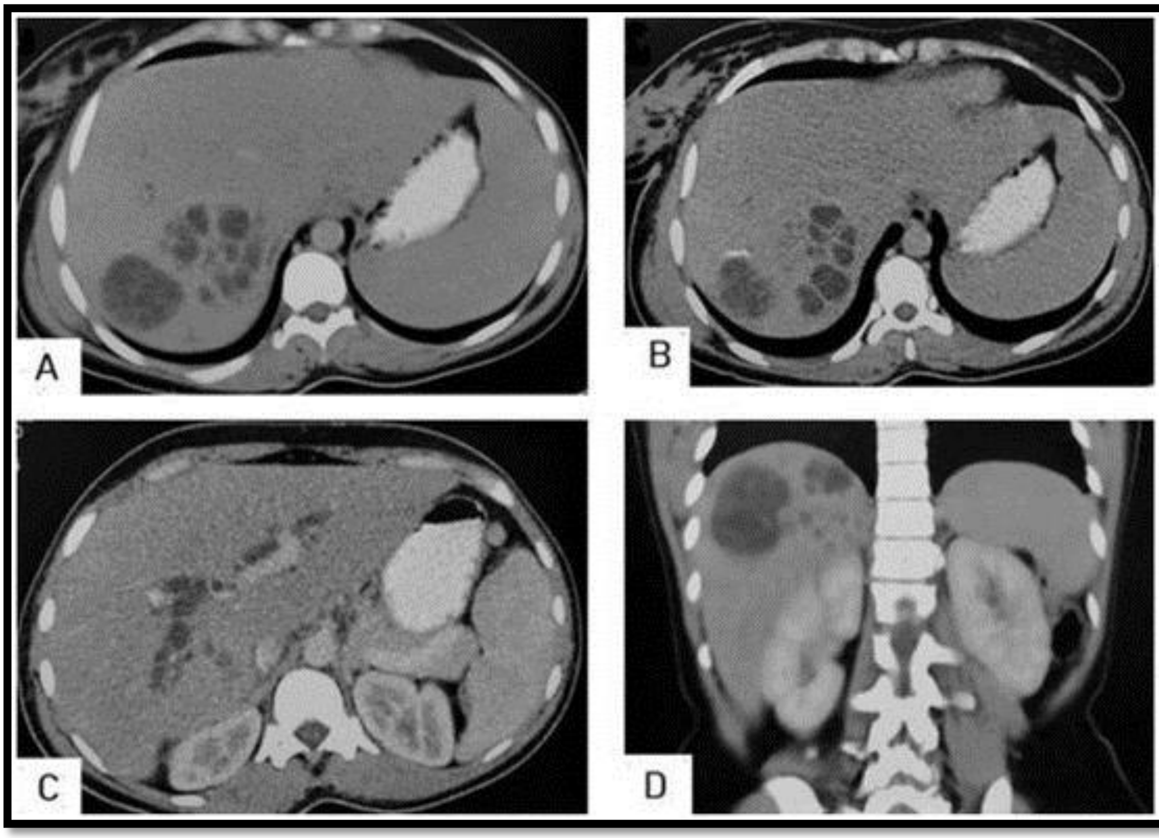

Fig.5.Show the CT images of a 30-year-old male showing multiple well-defined grouped cysts in the right lobe of the liver

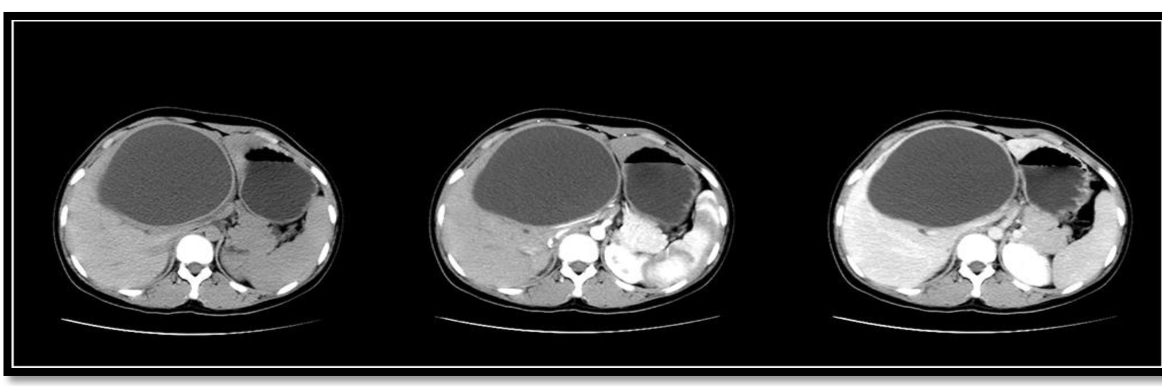

Fig.6. show the Cystic echinococcosis—the simple cystic type.

Fig.7. Show the CE of Cystic echinococcosis—the multi-daughter cystic type.

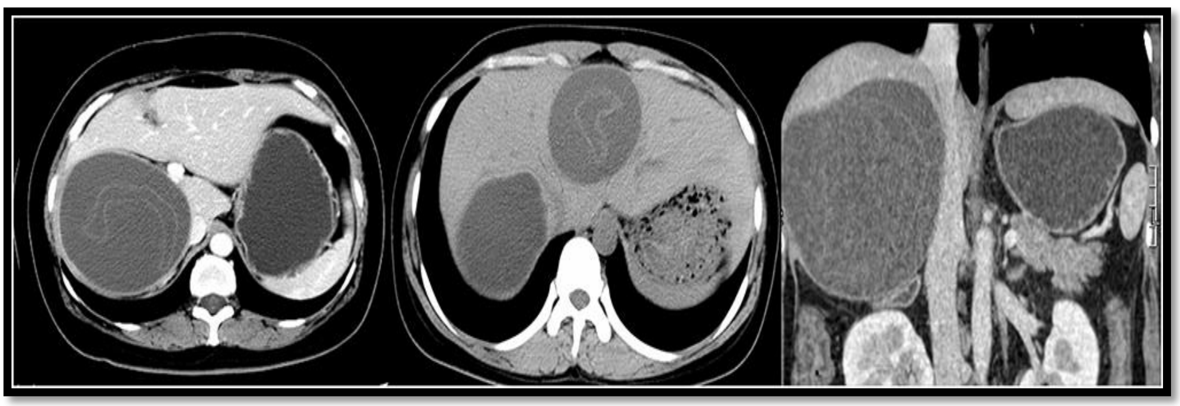

Fig.8. Show the CE of cystic echinococcosis—the inner capsule detached type.

Epidemiology

Hydatid disease is a global zoonotic infection caused by Echinococcus in its larval stage. Hydatid disease is caused by *E. granulosus* and *Echinococcus multilocularis* which are the two main types. *E. granulosus* is mainly seen in New Zealand, Australia, the Middle East, and Africa which makes it the most common type of hydatid disease in humans [29-31]. The classical findings are well known, however, findings related to unusual anatomic locations and extensions are less frequently described in the literature [6]. Here, we present an interesting case of hepatic hydatid cyst with exophytic growth and transdiaphragmatic thoracic involvement with likely impending rupture.

Role of ultrasound and computed tomography (CT)

Ultrasound is the primary modality of diagnosis but CT scan can help in various cases like excessive abdominal gas, high build status, and history of previous surgeries or disease extensions (communication with the biliary tree). CT has high sensitivity and specificity for hepatic hydatid disease [32]. CT scan help in the diagnosis of hydatid cyst by showing the high attenuation wall of hydatid cyst which can be seen both when it is calcified or even without calcification, but certain setbacks like in cases of contrast enhancement, hemochromatosis, and drug therapy (like amiodarone, etc.) can increase hepatic attenuation which can obscure visualization of wall of the hepatic hydatid cyst [6]. So non-contrast CT (NCCT) is better than contrast-enhanced CT scan as contrast can obscure the high attenuation cyst wall. A free-floating membrane within the cyst seen as high attenuation membrane is the peri-cystic laminated membrane of hydatid. Multiple, round peripherally arranged vesicular structures within the main cyst are the daughter cysts that contain cystic fluid of low attenuation as compared to the main cyst [33].

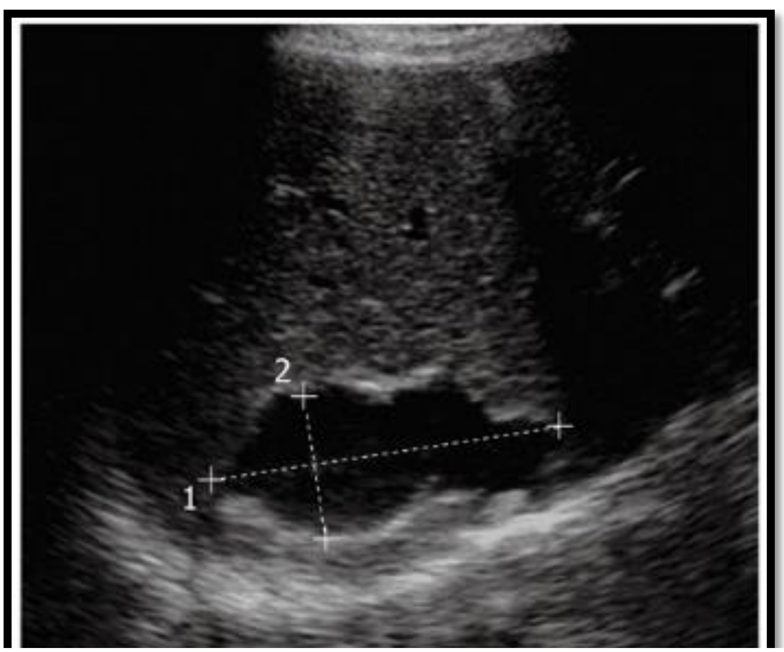

Fig.2. show the liver hydatid disease in a 55-year-old man appears as a well-defined anechoic mass without hydatid sand and septa

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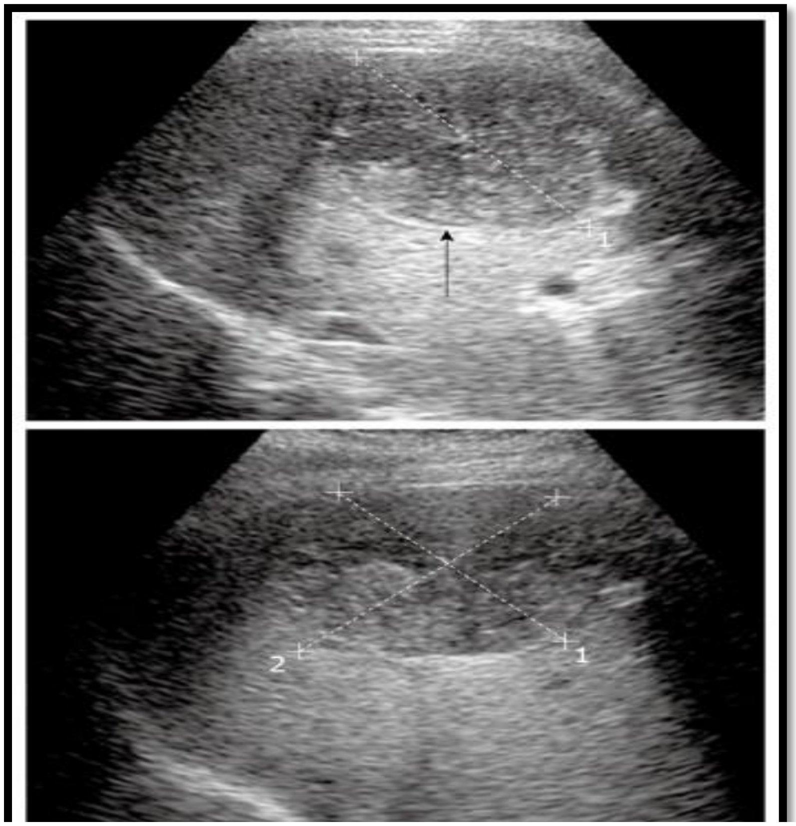

Fig.3.Ultrasonography images of hydatid disease show multiple internal septa and floating membranes inside the cyst. Note the cyst wall is visible as double echogenic lines

concentric layers acidophilic, soft and acellular, milky characteristic appearance. And another is an internal germinal membrane, thin, grainy and nucleate. The cyst contains a liquid compound NaCl, carbohydrate, protein and lipid. This liquid has a spring water appearance if the cyst is healthy, opalescent or purulent if infected. Daughter vesicles fertile cyst containing 10 to 100 scolex, born of the germinal membrane and bathed in liquid hydatid. This explains the interest to evacuate the block cyst to avoid dissemination. Between brain tissue and cyst volume increase, forms a third membrane or adventitious, easy peel and helps to better remove the cyst fluid (33).

Diagnosis

Imaging procedures such as ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) play a great role in the diagnosis and evaluation of the extent of HD [34-36]. The choice of the most appropriate imaging technique for the proper diagnosis of HD depends on the organ involved and the stage of the disease. US is considered as the best imaging method of choice for the initial diagnosis of HD since it allows cysts categorization into different types according to its nature as; solitary univesicular, solitary multivesicular, solid echogenic mass, multiple univesicular, multiple multivesicular, or collapsed, flattened and calcified [18]. It can define the number, site, dimensions, and viability of the hydatid cysts [19]. The most widely used and standardized classification for hydatid cyst was proposed by the World Health Organization (WHO) in 2001 [37] and based on Gharbi's classification (1981) [21]. The WHO classification system relies on some CE-specific US imaging features which help in the diagnosis, staging and follow-up of CE cysts [20]. In addition, US is considered the preferred tool in monitoring the efficacy of anti-hydatid therapy before and after surgical removal of the hydatid cysts [38]. On the other hand, CT scan has high sensitivity and specificity over the US for HD diagnosis [21] and is required in cases where the US proved inadequate due to patient-related difficulties such as obesity, excessive intestinal gas, previous surgery [21-24] and dead or calcified cysts [39]. MRI is a useful imaging technique and superior to any other imaging modalities in demonstrating neural involvement [40]. Furthermore, MRI can demonstrate cyst wall defects, cyst contents passage through defected, floating membranes and any irregularities of the cyst rim representing incipient detachment of the membranes [25-27]. The MRI characteristic sign of HD is represented by the low-signal-intensity rim on T2-weighted images "rim sign" [28-30]. The "snake sign" is another typical MRI finding, which represents collapsed membranes due to damaged r degenerated hydatid cyst. Both CT and MRI are very important in the diagnosis and management of HD as they provide a broad knowledge of the lesion size, location, and relations to other structures. CT and MRIbased hydatid cyst classifications were described by Kalovidouris, et al. [31] and von Sinner, et al. [32], respectively. Radiological diagnosis can be further confirmed by several antibody-based immunological assays. The high rates of false negative results of serological tests restrict their confirmatory role [28] especially at early cyst stage (hydatid fluid-filled endocysts) and late stage of involution (solid and wall calcified cysts).

Conclusion

Imaging plays a primary role in liver hydatidosis. It is used for diagnosis, for assessment of extension, for identification of possible complications, for classification and for monitoring the response to therapy. US, MDCT and MR have different roles depending on accuracy in depicting the different goals.

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