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Case Study

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HYDATID CYST OF LIVER, A CASE REPORT

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INTRODUCTION

The hydatid cyst is zoonosis caused by Echinococcus species. It is recognized as a global public health problem by WHO. Hydatid cysts have non-specific history and clinical manifestation. Ultrasound is a cheap and widely used modality for diagnosis, treatment and prognosis of this disease. We present a typical case of hydatid disease.

CLINICAL REPORT

A 40-year-old woman presented to the cardiologist with chief complaints of non-radiating, right sided chest pain and breathlessness for the past 1 – 2 weeks. Her chest work up included ECG, echocardiography and TMT. All these tests were within normal limits and RAT was negative. She was stable and had no significant past medical and surgical history of note; in particular, there was no history of cancer or predisposing factors for chronic lung or liver diseases. The patient's clinical examination findings; laboratory test results, including complete blood count; and liver function test results were within normal limits.

As part of her initial work-up, she was sent to our center to undergo chest radiography and routine abdominal ultrasonography (USG). In USG of the abdomen, a large well defined cystic lesion with multiple small daughter cysts and central solid matrix was noted. Cyst wall was thin and irregular with few foci of intermittent calcification. This cystic lesion was located in segment V-VI of liver (Image 1). No other cystic lesion or abnormalities were noted in liver or rest of abdomen.

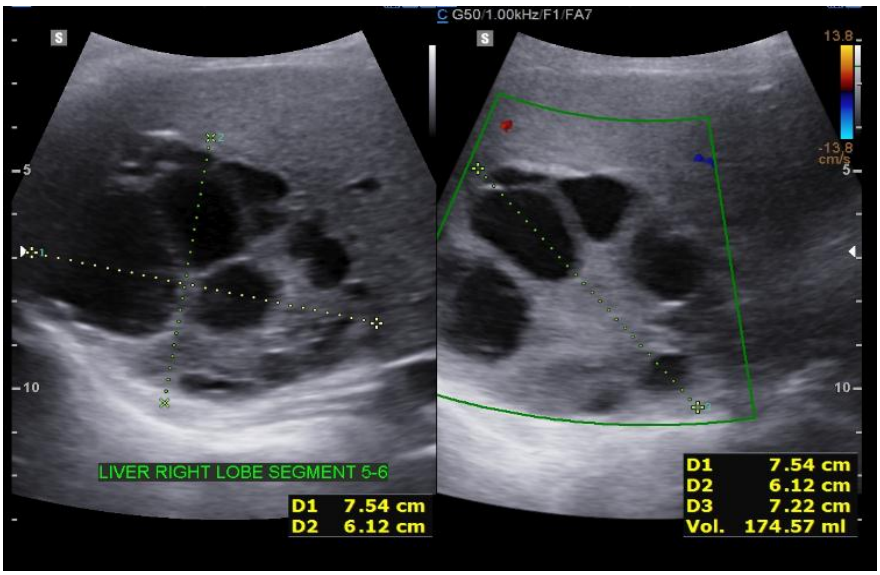


Image 1: USG of abdomen shows a large cystic lesion with mutiple daughter cysts and soild martix. Wall of the cyst is irregular with intermittent calcifications. No central vasularity was noted on doppler.

Based on these USG findings, the patient was advised for Computed tomography (CT) scan. Patients underwent plain plus contrast enhanced CT (CECT) of chest and abdomen; this confirmed the USG findings as non-contrast enhancing cystic lesions with multiple small daughter cysts and central solid matrix (Image 2). No other obvious cystic or solid lesion was identified in rest of the abdomen and in chest. Based on USG findings, the hepatic hydatid cyst was classified as WHO CE 3b (transition stage).

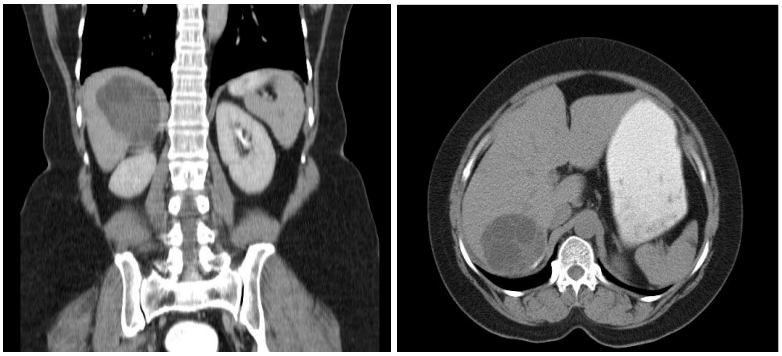


Image 2: CECT of abdomen (axial and coronal images) shows cystic lesion with multiple daughter cysts and solid matrix. Wall of the cyst is irregular with intermittent calcifications. No obvious contrast enhancement was noted.

DISCUSSION

The hydatid cyst or Cystic echinococcosis (CE) is accidental infection of the human with the eggs of Echinococcus granulosus, followed by the development of the larvae. Most common location of infection is the liver (50-70%), and other less common organs are the lungs, spleen, kidneys and brain.

Animal hosts of the parasites comprise wild carnivores, farm and domestic animals, and other small mammals including rodents. Infection in humans occurs by ingestion of Echinococcus eggs that are most commonly shed in faeces of dogs and other animals lik wolves, jackals, and coyotes. CE has been strongly linked with the sheep-raising industry and dogs which act as intermediate and definitive hosts.

CE has been recognized as a global public health problem. CE is globally distributed and found in every continent except Antarctica. Alveolar echinococcosis is confined to the northern hemisphere, in particular to regions of China, Russia and countries in continental Europe and North America as reported by WHO.

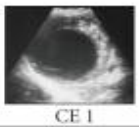
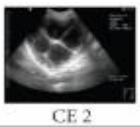
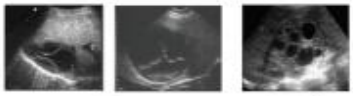
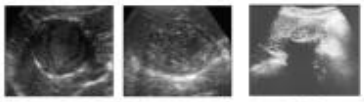
In India, the highest prevalence is reported mostly from the southern part Andhra Pradesh and Tamil Nadu 2-6. Highest incidence in rural areas, where exposure to domestic animals like cattle including sheep and stray dogs is common. History of consuming raw vegetables and water contaminated with excreta of an infected dog is common findings.

There are various ways to investigate and confirm a suspected case of CE but USG is a cheap and most widely used. Radiography has no role in abdominal CE. CT and MRI are used to confirm diagnosis and to rule out complications from CE.

Based on the USG findings of CE, Gharbi proposed classification in 1981[7]. In 2003, the World Health Organization Informal Working Group on Echinococcosis (WHO-IWGE) proposed a standardized USG classification based on the active-transitional-inactive status of the cyst (table 1).[8,14]

The WHO-IWGE classification sets both the staging of hepatic hydatid cysts and the therapeutic attitude depending on this staging. This standardized classification scheme is intended to promote uniform standards of diagnosis and treatment.

Table 1: Classification WHO and Gharbi from A. da saliva 'Human echinococcosis: a neglected disease'.

Gharbi 1981	WHO classification (cyst types)		
Type I	Univesicular anechoic cystic lesion with double line sign (CE1)	Active	 CE 1
Type III	Multiseptated, "rosette-like"/ "honeycomb" cyst (CE2)	Active	 CE 2
Type II	Cyst with detached membranes "water-lilly-sign" (CE3a) Cyst with daughter vesicles in solid matrix (CE3b)	Transition	 CE 3a CE 3b
Type IV	Cyst with heterogenous content (hypoechoic/hyperechoic). No daughter vesicles (CE4)	Inactive	 CE 4 CE 5
Type V	= CE4 plus calcified wall (CE5)	Inactive	

The WHO classification provides a rational basis for choosing an appropriate CE treatment and follow-up. Following are types of CE according to WHO classification

The CE1 stage is simple unilocular cyst with anechoic content and a visible double cystic wall. In early stages when the cysts are smaller than 4–5 cm and especially in children, the thick walls may not be seen. Therefore, differential diagnosis with simple liver or kidney cysts may sometimes be difficult.

The CE2 cyst is completely filled with daughter vesicles. It appears as "septa" are not true septa but the cyst walls of the daughter vesicles adjacent to one another

CE3 cysts include two stages, CE3a and CE3b, which differ in morphology and clinical characteristics. CE3a is classical "water-lily" sign, floating membranes, i.e. the endocyst detached from the cyst outer wall (pericyst). CE3b is a predominantly solid lesion with daughter vesicles. CE3a may go on to become "solid" (inactive) or may give rise to daughter vesicles, in which case it becomes a CE2 cyst.

CE4 iss coarse variable (hyper, hypo) echogenic echotexture cyst without daughter vesicles. The "ball of wool" sign, is detached endocyst in hypoechoic folded structure embedded in a hyperechoic matrix, in USG. However, often a definitive diagnosis of CE in this stage cannot be made by USG findings alone. If the CE4 stage is reached spontaneously, these cysts tend to remain inactive over time and, if asymptomatic, need only USG monitoring.

CE5 cysts are partially (with an egg-shell calcified wall) or completely calcified with shadowing. These cysts are not viable in the vast majority of cases. Definitive diagnosis cannot be made by ultrasound findings alone.

The "CL" category when findings are of undifferentiated 'cystic lesion' that requires further investigations before a definitive diagnosis. As such, strictly speaking, CL is not a "stage" but rather a temporary label assigned to a cyst whose parasitic nature is still undefined. This is very helpful in ultrasound surveys in endemic areas.

CE cysts have to be differentiated from other conditions, such as non-parasitic cysts, single or multiple hemangiomas, pyogenic or amoebic liver abscesses, hematoma, and neoplasia with hemorrhage and necrosis (e. g., large adenoma, hepatocellular carcinoma, metastases, lymphoma), biloma and post-surgical sequelae and textiloma.[9] Most frequently, simple cysts are encountered but atypical cysts sometimes pose a diagnostic challenge. These include biliary cysts, polycystic liver disease, mucinous cystic neoplasms (cystic (biliary) adenoma, cystadenoma) and cystic metastases. Additionally other infectious agents must be considered: fungal, bacterial and amoebic abscesses.[10] In most uncertain cases, diagnosis can be achieved using aspiration. Only under particular circumstances, small and very large (>50 mm), asymptomatic and uncomplicated simple cysts, may be monitored. This can be done by serial ultrasound at six-month intervals for the first two years following diagnosis. Significant growth, the development of progressive symptoms, or any suspicion of neoplastic change requires a definite diagnosis and surgical intervention.

Determining whether a cystic lesion is echinococcal depends on the presence of a double wall and is obvious when membrane detachment is present. Simple or minimally complex cysts, as well as biliary cystadenocarcinomas or abscesses, lack these features.

TREATMENT

USG has a crucial role in percutaneous treatments and evaluation of treatment response, particularly in assessing for inactivity of type CE4 and CE5 asymptomatic liver cysts when managed expectantly, i. e. the so-called "watch and wait" approach.[11]

The PAIR technique (puncture, aspiration, injection of 95% ethanol solution or hypertonic saline solution, re-aspiration) is applicable to the hepatic hydatid cyst in stages CE1, CE2, CE3 liver cysts.[9]

CE2 and CE3b cysts are not responsive to PAIR.[12] Although daughter vesicles can be punctured individually, these stages show growth of new daughter vesicles in the weeks following a procedure.[9,12] Successful drainage of the entire cyst content via large bore catheters has been reported but studies with larger cohorts of patients are needed to compare these methods with surgery.[13] No interventions is generally required for inactive cysts (CE4 and CE5), in which the "watch-and-wait" attitude can be adopted.

CONCLUSION

It can be said that patients with hepatic CE form a heterogeneous group with unspecific history and clinical manifestation. USG allows diagnosis, differential diagnosis, treatment guidance and follow-up. The use of WHO classification in reporting of CE is aimed to provide uniform reporting and clinical treatment to patients. This will lead to minimally invasive methods, high applicability, less frequent complications and shorter hospitalization.

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