

X-Ray-Evaluation

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Cystic Echinococcus of Liver in a 13 Years Old-Girl

Brief clinical information: NT, 13 years old, female patient, presented with stomach pain complaint. The case with intermittent stomach ache lasting for a year was referred to our hospital for advanced examination and therapy following the detection of cyst hidatid in the liver in the ultrasonography performed in the epicenter. The case has the story of 3-day hospital stay due to rashes on the body one year ago (why?). The family is in the stockbreeding business and look after a do at home.

PE; the liver is easily handled 2 cm under the rib and other system findings have been considered normally.

Laboratory tests: Echinococcosis IHA: 1/1280 positive, other blood test results were within the normal limits.

Abdomen USG (epicenter): the 93x89x75 mm diameter clear shaped cystic lesion with daughter vesicles in it in the supero-anterior segment of the liver right lobe (segment 8) was considered as cyst hidatid. Moreover, there was an irregular shaped 29x17 mm diameter hyper echoic lesion (hemanjiom?) in the supero-anterior segment of the liver right lobe (segment 5).

A brief clinical and radiological evaluation of the patient: In the CT analysis of the patient; a cystic lesion with a small calcification on its wall and a water lily flower image which is atypical image of the germanium membrane with a fluctuating course detached from cyst wall is detected. The un-changing round-oval appearance of the cyst leads us to think that the lesion is within the transition zone (it may be compatible with CE3; CE3a or CE3b) (It should be remembered that the

WHO's classification is a US-based classification; the pre-admission symptoms of the patient supports the view that they are compatible with the phase CE3 of the WHO). It was considered appropriate to continue the albendazole treatment of the patient and to demand a consultation from Pediatric Surgery for a percutaneous drainage. Besides, in the follow-up of the patient, it was decided that CT scan would not be performed (due to radiation risk) unless there was an additional indication. The patient followed-up by serial ultrasonography, that the findings of inactivity or whether a new lesion developed to be recorded and that the patient to be serologically monitored.

Brief general clinical information about the cystic echinococcus in the liver and its agent *Echinococcus granulosus*:

Among the four types of echinococcus (*Echinococcus granulosus*, *E.multilocularis*, *E. vogeli*, *E. oligarthrus*, the first two are more frequent) causing infections in humans, *E. granulosus* is the most frequent one in the world and especially in our country and typically causes cystic echinococcosis. Many infections are acquired in childhood; this primary infection is always asymptomatic and stays asymptomatic for years and it generates clinical symptoms in adults (or in late childhood) after many years, or is accidentally detected in an asymptomatic patient. The size of the cysts typically increases 1-5 cm per year; however, this is very variable (1, 2). It is involved the most in the liver (2/3 of the patients, 60-85% in the right lobe), followed by lung (nearly 25% cases) and the other organs. There is 85-90% one-organ involvement in the *E. granulosus* infections and only one cyst is detected in >%70 cases (3). There is usually no significant symptom in cysts smaller than 10 cm; in the larger cysts, especially in the right

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Figure 1 a-c. In the contrasted abdominal CT of the patient; a cystic lesion with a small calcification on its wall and a germinative membrane with a fluctuating course detached from cyst wall within this cystic lesion are visible in the right lobe antero-superior segment of the liver (segment 8). This tomographic image is compatible with the phase CE3 in the USG-based classification of the WHO. (By the written permission from the archive of the Pediatric Thorax Council of the Department of Pediatric Infectious diseases of the Uludağ University Medical Faculty).

CT: computed tomography; WHO: World Health Organization; US: ultrasonography

CL	CE1	CE2	CE3	CE4	CE5
Cystic lesion	Active	Active	Transitional	Inactive	Inactive
Specification					
CL	CE1	CE2	CE3	CE4	CE5
CL: Nonspecific cystic lesion; uniform, anechoic image	Unilocular, simple cyst, uniform, anechoic, cyst wall is visible, lesion may be round or light, thin echogenic fluctuation via position changing (hidatid sand; snow flake sign)	Multivesicular, multiple septal cyst, cyst wall is visible (round or slightly oval), septations may be in the shape of a wheel, virgin vesiculars may form the shape of a badge or honey comb.	Wavy, moving germinative membrane; water lily flower image (C3a) unilocular cyst inclusive of virgin vesicular, fundamentally, predominantly solid structure rather than hypoechogenic start to dominate (C3b). It is important to detect the virgin vesicular in this phase in guiding the treatment.	Heterogenic hypoechoic or hyperechoic degenerative encysted structures, virgin vesicular may be invisible, degenerative membranes may seem like a ball of wool.	Thick, calcified cyst wall (may seem like a bow) calcification may be partial or complete. (NOTE: Differential diagnosis in CE4 and CE5 may not be made only through ultrasonography.)
Recommended treatment					
	<5 cm; albendazole >5 cm; albendazole +PAIR,	Albendazole + percutaneous catheter or surgery	C3a; <5 cm; albendazole >5 cm; albendazole +PAIR, C3b: albendazole + percutaneous catheter or surgery,	Follow-up	Follow-up

Figure 2. WHO ultrasonographic classification of the Cyst Hidatid (3, 11)
CL: Cystic lesion, CE: Cystic echinococcus (stage)

upper quadrant, there may be abdominal pain, nausea, vomiting, sometimes pressure symptoms, cystic leakage or allergic symptoms including anaphylaxis in the presence of rupture, biliary colic in the presence of rupture in the biliary tree, hepatitis and cholangitis. Laboratory tests are usually non-specific; leukopenia, thrombocytopenia, nonspecific liver function tests, sometimes eosinophilia in the presence of antigenic material leakage may be observed. Calcification in cysts may usually develop after 5-10 years; it is frequently observed in hepatic cysts and the total calcification of the cysts wall leads us to think that the cyst is inactive.

Serology is useful in primary diagnosis and follow-up after the treatment. Antibody detection is more sensitive than E. granulosus antigen detection (4). Serologic antibodies may be detected by different methods (such as complement fixation, latex agglutination, haemagglutination, ELISA, IgG and IgE); however, ELISA is the most sensitive (80-92%) and the most specific one (98-100%) (3).

The treatment of cyst hydatid can be performed by medical (albendazole), surgical, percutaneous drainage methods (PAIR or evacuation via catheter) or the combination of all these. In medical treatment, albendazole 10-15 mg/kg/d divided in two doses is given for the recommended period (sometimes for months). Mebendazole therapy may alternatively be given. Surgical treatment is recommended in complicated cysts (cyst rupture, cysts forming biliary fistula, pressure on vital structures, secondary infection or hemorrhage) or the presence of multiple virgin vesicular cyst (such as phase CE2 CE3b of the WHO), surface cysts with a high rupture risk due to trauma, and very large (>10cm) plus out-of-liver other region cysts where percutaneous drainage treatment is not appropriate. Postop complications are rare (<1%), 2-25% recurrence is possible (4-7). Percutaneous treatment or the PAIR method (punction, aspiration, scolexidal agent injection, and repeated respiration and phase CE2 CE3b of the WHO excluding daughter vesicles) or the method of discharging all the cyst via a big catheter may be performed (it is recommended especially with phase CE2 CE3b of the WHO including virgin vesiculars or after the relapse difficult to drain via PAIR). Percutaneous drainage may be performed under the guidance of ultrasonography or CT. Before surgery or percutaneous drainage, albendazole should be started at least one week earlier and can continue at least for a month or longer after the operation (3, 8).

The imaging methods are important in the diagnosis of cystic echinococcosis. Plain radiography may reveal the cystic calcification, but cannot detect non-calcified cysts; therefore, it is not sufficient for the differential diagnosis. Its ultrasonographic sensitivity is 90-95%. Ultrasonography is used both in the diagnosis and follow-up. It is seen in the shape of anechoic and round (may be difficult to differentiate from the benign cyst) cyst in the ultrasonography. If there is daughter vesicles, typical encysted internal sep-

tation may be seen. If the patient moves around during the ultrasonography scan, it may seem like as if it is snowing or there are moving sand grains within the cyst. Sometimes, internal wall folds and germinative membrane detachment (water lily flower appearance) may be detected. Ultrasonography makes various hydatid cyst classification possible and the most accepted one is the WHO classification (Figure 1) (9-11). Based on the symptoms showing the biological activities of the cyst, ultrasonographic classification can be active cyst, transition period or inactive cyst. Inactive lesions are collapse, non-round, oblate ellipse-like cyst, germinative membrane detachment from the cyst walls (water lily flower appearance), rough, thick echos inside the cyst and cyst wall calcification. Calcified cyst may generate the image of an egg shell. According to the WHO classification, CE1 and CE2 is considered as active cyst and CE4 and CE5 inactive cyst; CE3 is the transition zone. The increasing echogenicity levels together with the calcification in the repeated ultrasonographies show that the cyst is always nonviable.

CT is suggested to be more sensitive (95-100%) than ultrasonography. CT is the best way in assessing the size of the cyst, its number and anatomical localization. It is also better than ultrasonography in detecting complications such as infection and extra hepatic cysts and may be used in establishing the result of the treatment and its recurrence. MR imaging is not more advantageous than CT and in most cases, there is no need to demand it in addition to CT.

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