Cystic echinococcosis

Summary

- Infection from eggs in dog feces.
- Larvae form large cysts with internal daughter cysts.
- Cysts in the liver and lungs, rarely in other organs.
- Often asymptomatic, sometimes symptoms due to pressure upon surrounding organs.
- Risk of rupture with anaphylaxis or dissemination.

General

There are several tape worms in the genus *Echinococcus*: *E. granulosus, E. multilocularis, E. vogeli, E. oligarthus, E. shiquicus.* The most important and frequent one is *E. granulosus,* causing cystic echinococcosis or hydatid disease*. Echinococcus granulosus* is a small tapeworm (a few mm long) which infects dogs and other canines. Its distribution is world-wide. In some regions the problem is very important such as North Kenya around Lake Turkana and Kyrgyzstan and the surrounding central Asiatic republics. Various animals (sheep, goats, cattle, pigs) may become infected with the eggs in dog faeces. In the animal's intestine the larva (called "oncosphere") emerges from the egg. It penetrates the intestinal wall and is carried by the venous blood towards the portal vein. After development of the parasite, hydatid cysts are formed in internal organs. The cycle is completed when a dog has the opportunity to eat offal containing hydatid cysts. In the dog's intestine adult *E. granulosus* then develop, after which egg laying can begin. Each hydatid cyst leads to multiple adult worms.

Humans are accidental hosts. If humans take water or food contaminated by dog faeces, they will develop one or more hydatid cysts. The cyst contains fluid and daughter cysts and is known as a hydatid cyst. On the inside of each cyst is a germinal membrane. From this membrane countless protoscolices (small heads) develop. There is thus multiplication at the larval stage. A capsule of connective tissue is formed around the cyst. This capsule consists of the cyst wall together with the germinal membrane. The majority of cysts are found in the liver and lungs, but other locations are also possible (brain, bones, spleen, kidneys). These

are often continuously growing cysts, which may produce pressure on surrounding organs, may rupture or die off and calcify. When the parasite has died and disintegrated the hooks which were situated at the former heads remain in the sandy fluid of the dead cyst, and these can be seen under a microscope. This is useful if there is doubt concerning the nature of a cystic lesion.

Clinical aspects

Humans are generally infected faecal-orally during childhood. The cysts grow very slowly, about 1 to 2 cm per year. The carrier may remain asymptomatic for a long time and symptoms are unusual before the cyst has reached 10 cm in diameter, at least in the liver, its preferred localization. There may be mechanical consequences. Pressure on surrounding organs leads to various symptoms and complaints. Hepatic cysts may lead to an enlarged liver with local discomfort, obstructive icterus with or without cholangitis. If localization is in the central nervous system this produces symptoms of a brain tumour, epilepsy, compression of the spinal cord or brain stem and even eosinophilic meningitis if there is spillage. If situated in the skeleton there is often bone pain, sometimes with fractures. This has to be differentiated from ordinary bone cysts or tumours. Lung cysts are usually asymptomatic, but sometimes there is a cough and thoracic discomfort. Renal cysts are sometimes found by chance and may cause unilateral kidney destruction. Allergic reactions may also occur, such as urticarial rash, bronchospasm, anaphylactic shock after rupture of a cyst (which may be spontaneous, after trauma or during surgery). After rupture there may be dissemination of the protoscolices in the peritoneum or pleura. Mechanical aspiration of a cyst may sometimes lead to rupture with allergic shock and dissemination.

Diagnosis

Plain X-ray of the abdomen (crescentic calcifications), X-ray of the lungs or CT scan. Ultrasound of the liver shows a round or oval hypodense zone with retro-acoustic intensification. The cyst can contain septa or daughter cysts. The wall may appear split (the endocyst separated from the pericyst) or it may be partially or completely calcified. Sometimes the cyst appears heterogeneous and produces a pseudo-tumorous image. Sometimes the diagnosis is made during surgery. In case of doubt as to the nature of a cystic mass, the content of the lesions may be examined for the presence of hydatid sand or the

presence of the typical small hooks which remain after the protoscolices degenerate. Serology may be negative in the case of well encapsulated liver cysts and lung cysts. Sometimes the serology is positive or the titer increases during treatment due to leakage of the cyst content and release of antigen which cause the immune response to increase.

Ultrasound

Various types of cysts can be identified by ultrasound. The following signs are regarded as pathognomonic for cystic echinococcosis (CE):

- Unilocular, anechogenic round or oval lesions with a pronounced laminated membrane or with snow-like inclusions.
- Multivesicular cysts or cysts with multiple septa with a wheel-like appearance.
- Unilocular cysts with daughter cysts which may exhibit a honeycomb appearance.
- Cysts with floating laminated membranes (“water-lily sign”) which may also contain daughter cysts.

Ultrasound is also of utmost importance to stage the liver cysts according to the 2010 WHO classification (see Fig 2 below), between active (or early: CE1 and CE2), transitional (C3a and C3b) and inactive (or late: CE4 and CE5) lesions. This has immediate implication for the prognosis and treatment of cystic echinococcosis

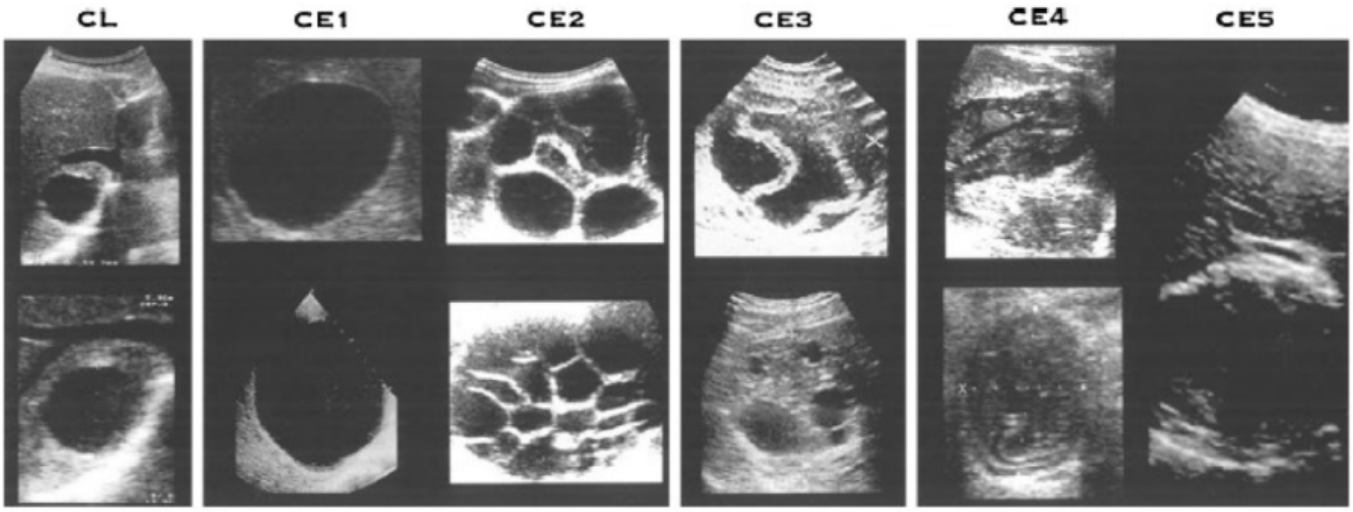

Fig. 2. WHO-IWGE standardized classification.

WHO Classification of hepatic hydatid cysts

CL	Unilocular anechoic cystic lesion without any internal echoes and septations
CE 1	Uniformly anechoic cyst with fine echoes settled in it representing hydatid sand
CE 2	Cyst with multiple septations giving it multivesicular appearance or rosette appearance or honey comb appearance with unilocular mother cyst; this stage is the active stage of the cyst
CE 3	Unilocular cyst with daughter cysts with detached laminated membranes appearing as water lily sign; this is the transitional stage of the cyst
CE 4	Mixed hypo and hyperechoic contents with absent daughter cysts, these contents give an appearance of ball of wool sign indicating the degenerative nature of the cyst
CE 5	Arch-like thick partially or completely calcified wall; this stage of cyst is inactive and infertile

Treatment

Waiting

Many cysts remain stable, calcify or even involute spontaneously. Small, calcified cysts in the elderly can usually be left untreated. As a whole, a wait and see attitude is recommended for CE4 and CE5.

Surgery

Pericystectomy or partial liver resection. Sometimes what is known as the “frozen-seal” method is applied. Using liquid nitrogen, a funnel is frozen onto the liver capsule to prevent accidental spillage.

The liver is opened and the cyst content evacuated. During the operation, lavage is carried out with a scolicidal agent. Surgery is the treatment of first choice for large cysts (> 10 cm), for CE2-CE3b lesions, if there is superinfection or communication with the biliary tract. For extrahepatic cysts, surgery is always the treatment of first choice. Albendazole is administered ideally prior to surgery (but optimal timing is unknown, up to 4 weeks), and praziquantel is given at the time of the operation. This is done in order to diminish the risk of disseminated infection in case of accidental rupture or spillage during operation. Post-operative complications are not unusual.

Medication

Mebendazole is no longer used (only at high dose, in case of albendazole toxicity). Long-term therapy with albendazole (e.g. 800 mg daily for 6 to 9 months, blocks glucose uptake by the parasite) is usually used alone for CE1 and CE3a lesions < 5 cm and in combination with PAIR or surgery for bigger lesions or in CE2 and CE3b lesions. It is used in extended duration for inoperable and/or disseminated disease.

Previously this was given in cycles, but nowadays the medication is administered daily without interruption. The efficacy of medical therapy varies greatly (overall cure rate of 30%)

and clearly leaves much to be desired. Higher levels of albendazole sulphoxide (ricobendazole), the chief active metabolite, may be obtained by higher dosage, ingestion with a fatty meal, or by combination with praziquantel or cimetidine [cimetidine inhibits the breakdown of both albendazole and praziquantel].

Albendazole cannot be used during pregnancy. The combination albendazole (10-15 mg/kg daily divided in two doses) with praziquantel (40 mg/kg once a week) is probably more effective than either drug alone.

PAIR

Percutaneous treatment with the PAIR technique (puncture-aspiration-injection-reaspiration) can be used for CE1 and CE3a lesions. Daughter cyst should be ruled out, since their presence reduces the likelihood of successful treatment with PAIR. Experienced surgeons can perform a laparoscopic variant of this technique. In hospitals where the necessary equipment is available, after detection of a cyst an endoscopic retrograde cholangiography is carried out. This permits determination of whether there is any communication between the cyst and the biliary tract. Under ultrasound or CT guidance the cyst is punctured transhepatically with a fine needle. The cystic pressure can be measured. Vital cysts have a pressure of 8-75 cm water. Dead cysts have a low pressure (0-2 cm water). Subsequently 10-15 ml of cystic fluid is aspirated. Live protoscolices are actively motile upon microscopic examination.

Biochemical analysis of the fluid for the presence of bilirubin is carried out to exclude communication with the biliary three. If there is sufficient evidence of active echinococcosis, the remaining cystic fluid is aspirated. Afterwards a protoscolicidal agent is injected (generally 95% ethanol or15-20% hypertonic salt). As a guideline the amount injected should by 1/3 of the volume of the aspirated fluid.

After 10 to 30 minutes the cyst content is then aspirated again. The risk of rupture, dissemination or anaphylaxis is minimal if there is at least 1 cm (preferably 2 cm) between the liver capsule and the cyst wall.

If there is a cyst-to-biliary tract fistula, the PAIR technique cannot be used due to the risk of

sclerosing cholangitis. It is advisable to begin albendazole one week before and to continue administering this until 4 weeks after the procedure. PAIR cannot be used for extra-hepatic lesions. Those who have no experience with PAIR are advised to leave this to an expert as the complication rate is quite high.

Prevention

De-worm dogs and prevent them from eating offal.

Keep dogs out of slaughterhouses.

The first results of a recombinant vaccine (EG95) administered to sheep and goats, are encouraging, and show protection of 83-100% for these animals.

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