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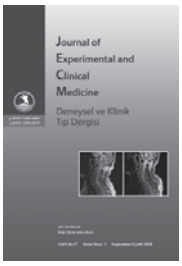

Review

Is calcified hydatid cyst absolutely dead? A case report and review of the literature

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ABSTRACT

How and when does a hydatid cyst die in humans? There is an ongoing debate in medicine about this subject. Calcification of the hydatid cyst wall is believed to indicate the death of the contents of the cyst . However, rarely, the cyst may be alive even though the wall is calcified. With this report, we presented a patient with a hydatid cyst with calcified wall. Our patient presented here was followed up for 8 years after the diagnosis and sufficient and effective treatment was not given thinking that the hydatid cyst was dead because of the calcified cyst wal, but intrabiliary rupture occurred after the follow-up. Criteria for the viability of hydatid cyst are discussed and the literature is reviewed.
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1. Introduction

In Cystic Echinococcosis, spontaneous death of the cyst is considered as natural healing. The exact mechanisms of spontaneous death of a hydatid cyst are not known. The two factors implicated in the spontaneous death are the immune resistance of the host and the infection of the cyst. These two factors play roles in similar frequencies in the death of the cyst (Czermak et al., 2008).

Antibodies produced in human body against hydatid antigens cause degeneration of the cyst contents and calcification of the cyst wall in 10% of the cases. Usually these patients are clinically silent. They are frequently identified coincidentally during examinations for other causes (Biava et al., 2001).

Another causative factor of death of the cyst is the infection. Through an ill understood mechanism, the infection of the cyst contents results in the death of the germinal membrane and the scolices in the cystic fluid. Infectious agents reach the cyst via the biliary system (Turgut et al., 2007).

Regardless of the cause of death, calcification of the wall occurs and, as a general rule, calcified cysts are considered as dead. However, there is no serological test to show whether the cyst is alive or dead (Hosch et al., 2007).

In general, a "coin-lesion" on plain radiographs is the manifestation of the calcification of all the cyst walls. A second, but more common appearance is a concentric calcification of the wall. Hosch (2007) and colleagues classified the calcification of the wall based on their appearances as "sprinkled", "eggshell-like", and "circular".

Here, we presented a patient who was diagnosed with calcified-dead hepatic hydatid cyst and followed up for 8 years without any treatment but subsequently developed intrabiliary rupture. Literature was reviewed.

2. Case

A 56-year-old male patient was hospitalized with abdominal pain in September 1999. Biochemical, hematological and serological examinations were unremarkable except a positive ecchinococcosis latex agglutination test at 1:6400. Computed tomography (CT) scan revealed a lobulated, heterogeneous, cystic mass, with partially calcified wall

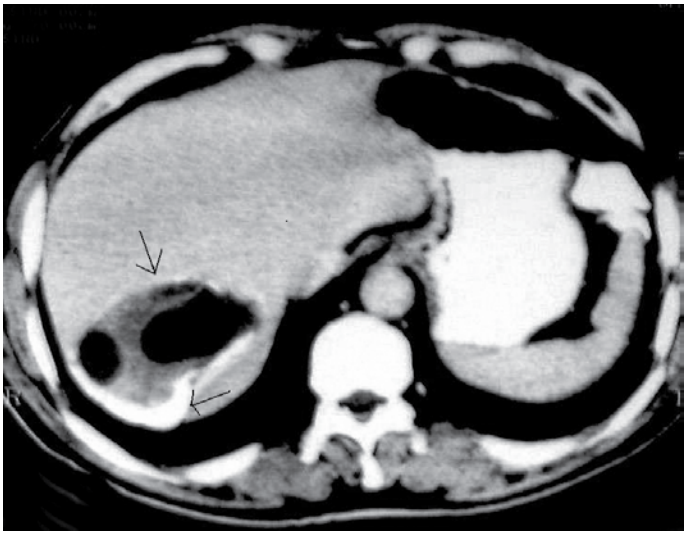

Fig 1: Computed tomography scan revealed a lobulated, heterogeneous, cystic mass, with partially calcified wall and measuring 4x5 cm in the posterior aspect of the right hepatic lobe (segment VII) in 1999. The arrows show the hydatid cyst and its calcified wall

and measuring 4x5 cm in the posterior aspect of the right hepatic lobe (segment VII) (Fig. 1). Cyst was considered "dead" due to its calcified wall. Patient was not given any treatment and discharged to attend a follow-up visit at 6 months.

During the follow-up visit on April 5th 2000, the same findings were observed on control CT scans and patient was scheduled for follow-up controls.

CT scan performed on May 21st 2001 showed that the cyst measuring 7x5x4.5 cm was hypodense, septated and had a calcified wall. Indirect hemagglutination assay (IHA) and enzyme-linked immunosorbent assay (ELISA) Ecchinococcus Ig G were positive.

In 2003, the patient was admitted to a private health center with abdominal pain where he underwent CT scan. He was told there was no change in the size and the content of the cyst and given symptomatic treatment.

Due to recurrent abdominal pain, the patient underwent another CT scan in the private health center in 2006 that showed a subdiaphragmatic lesion with heterogenous density in the right hepatic lobe, measuring 8.5x6.0x6.0 cm in size. Its wall was partially calcified and there were cystic areas inside the cyst, the largest one measuring 2 cm .

Patient was admitted to the Department of Gastroenterology with pain and jaundice in May 2008. Ultrasound and MR-cholangiography were carried out. They showed marked dilation of the extrahepatic bile ducts and determined a hydatid cyst possibly inactive and draining into the biliary ducts in the right hepatic lobe. Within the lumen of the choledochus, dense bile content or a membrane of the hydatid cyst, isointense with soft tissue, located in the distal end of the lumen and obstructing the bile flow was also reported.

During ERCP+sphincterotomy, debris composed of vesicles and cyst material was noted in the choledochus and irrigated.

Upon persistant jaundice and occasional abdominal pain, patient was admitted to the Department

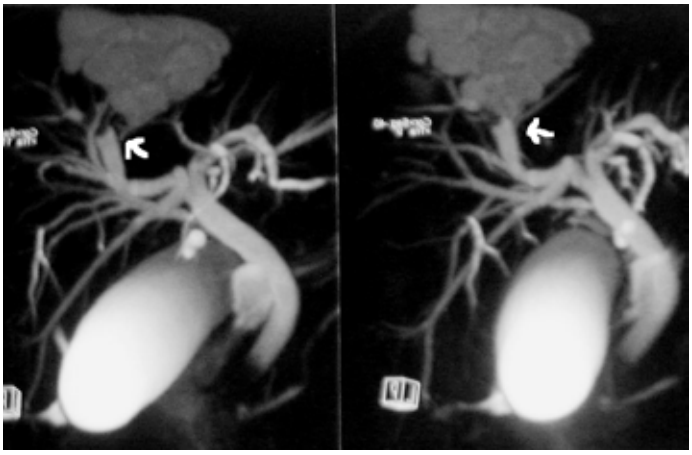

Fig 2: ERCP demonstrated cysto-biliary fistula in 2008 (the arrows show the fistula between the cyst and the intrahepatic biliary duct).

of Gastroenterology once again and ultrasound was repeated. US depicted an 8x5 cm cyst, cholelithiasis and cholesterolosis of the gall bladder. MRI was obtained and patient underwent ERCP that clearly demonstrated cysto-biliary fistula (Fig. 2). Patient was referred to our clinic for treatment.

Routine hematologic tests did not show a pathological finding other than mild anemia. Biochemical tests were normal. IHA and ELISA Echinococcus Ig G were positive.

During a joint meeting with the Department of Radiology, all radiological examinations performed in the last 8 years were re-evaluated in chronological order by a radiologist who concluded that:

In CT scan of 1999: A mass lesion measuring 8x5 cm with calcified boundary, heterogeneous solid and cystic content was identified in segment VII of the liver. It was argued that the vesicles could have been alive despite the calcified wall.

In CT scans of May 2000 and April 2006: The size of the cyst, peripheral calcification and internal structure were comparable. Cystic components had thin walls and were alive. Further, frank cystobiliary communication could be visualized on the images of 2006.

In MR examination of May 2008: Biliary ducts were dilated. Especially those adjacent to the cyst were more prominently dilated and connected to the cyst. Solid components were comparable with previous examinations but cystic components were enlarged. MR-cholangiography revealed similar findings in addition to hypo-intense areas within the lumen of distal choledochus where it opened into the duodenum. Pancreatic canal was normal.

When all findings were considered, the case was thought to be a hydatid cyst that has stayed silent for the past seven years, contained daughter cysts and opened into the biliary ducts, causing dilation of the ducts and obstruction of the distal end of the choledochus, possibly by the cyst contents.

Patient was scheduled for operation. He was given 10 mg/kg/day albendazol for 3 days preoperatively. An 8-cm hydatid cyst was identified in Segment VII during laparotomy on September 2, 2008. Moreover, a gallstone

of 0.5 cm was palpated in the gall bladder. Samples were taken from the live vesicles for parasitological examination. Cyst contents were evacuated. Cyst cavity was sterilized by albendazole solution, as described in an earlier study (Erzurumlu et al., 1995). Contents comprised of degenerated and dead daughter cysts as well as live daughter cysts. Cyst cavity and suprahepatic area were irrigated by albendazole solution. Cysto-biliary communication couldn't be seen in the cyst cavity. A thorough inspection could not be accomplished due to the localization. Biliary stain was not observed in cyst contents. Cholecystectomy and choledochotomy were performed. There was no cyst content within the lumen of the choledochus. Proximal and distal segments were irrigated by saline. Choledochoduodenostomy was performed. Omentum was prepared for omentoplasty. Capitonage+ omentoplasty was carried out for the cyst.

Patient did not have any complaints in early postoperative days and discharged on day 10. On day 20, patient presented with signs of infection. Infection of the cyst cavity was diagnosed and patient was treated medically.

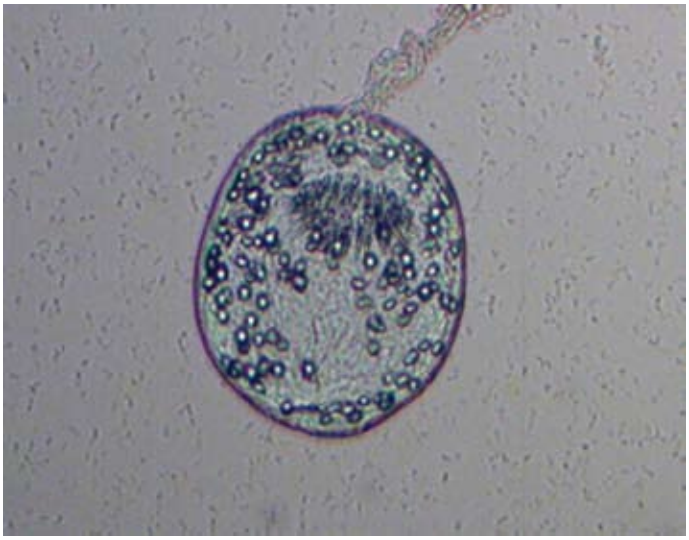

Fig 3: Live protoscolices were observed in the parasitological examination. Histopathological examination reported "hydatid cyst".

Live protoscolices were observed in the parasitological examination (Fig. 3). Histopathological examination reported "hydatid cyst".

3. Discussion

In humans, especially when located in the liver, calcification of the cyst wall is generally accepted as the indicator of the death of cyst (Hosch et al., 2007).

There are three findings that suggest the death of a cyst. These are calcification of the cyst wall, positive serological tests turning negative, and decrease in the intracystic pressure (Czermak et al., 2008).

Calcification of the cyst wall is the most frequently observed and easily identified sign. Whether it is a coin-lesion or partial calcification, they are interpreted in favor of cyst death. Although Sayek (2004), argued that calcification of the wall does not indicate cyst death, there is no data in the literature with regard to the presence of protoscolices or vesicles in calcified cysts. Therefore, calcified wall is accepted as a sign of cyst death.

Serological tests are widely used for the diagnosis of hydatid cyst. However, none of the tests has perfect sensitivity and specificity. IHA test, among the other tests, has the highest sensitivity and specificity. Biava et al (2001) reported that the most commonly used tests (indirect immunofluorescence assay, indirect hemagglutination assay, immunoelectrophoresis and co-electrophoresis with antigen 5 identification) had specificity of 80-94% and 65% for hepatic and pulmonary hydatid diseases, respectively. If the cyst is calcified or located outside the liver or lung, they recommended ELISA, Western-blot, polymerase chain reaction. They emphasized the importance of serological follow-up in medically or surgically-treated patients. However, there is no data in the literature as to when the tests become negative. A positive serology persists for a long time when the cyst is treated or died spontaneously. Unfortunately it is not possible to give a definite time. Hence, serological tests becoming negative are not very useful to determine cyst death (Filippou et al., 2007).

The cavity pressure in the live cysts is in the range of 50-80 cm water. In Gil-Grande's study (1993), it was noteworthy that a 3-week albendazole treatment was 94% effective on protoscolices and decreased cyst cavity pressure.

Based on these, it is difficult to set forth a conclusive finding indicating cyst death. In the light of the facts, calcification of the cyst wall is the most commonly observed and widely accepted sign of cyst death. On the other hand, it would not be totally correct to claim that there is no life in calcified cysts. As in our case, it is possible that dead vesicles co-exist with live ones in cysts with calcified wall. Viability can easily be diagnosed by experienced radiologists if the daughter cysts are big enough to be visualized. Difficulty arises when imaging modalities are insufficient to depict small-sized viable cysts. Therefore, we recommend benzoimidazol treatment for 2-3 months in cases with calcified cyst wall except those with eggshell-like or coin lesion-like calcifications. Such cases should be followed-up every six months by imaging methods. Morphological changes besides an increase in calcification may be an indicator of cyst's viability.

Having reviewed the CT images of 1999 and 2000 retrospectively, it was possible to see the live daughter cysts. Simple and frank cystobiliary fistula could have been seen developing gradually in CT, US and MR images taken since 2003. In 2008, live protoscolices have been found in the contents of hydatid cyst with calcified wall for more than 9 years.

Various treatment modalities are being utilized in cases which cystobiliary fistula developed. Choledochus exploration, biliary-digestive anastomosis,T-tube drainage, transduodenal sphincteroplasty are among conventional methods preferred by many surgeons. Some authors plan

surgery after ERCP+sphincterotomy. We evaluate and treat all cases of intrabiliary rupture by an algorithm previously reported (Erzurumlu et al., 2005). Primary mode of treatment of cyst is surgery, whichever method is used for choledochal evacuation and drainage. In the present case, ERCP+sphincterotomy+choledochal drainage were performed initially but was not successful. The surgery was later added.

In conclusion, physicians should carefully examine whether or not live vesicles are present within the cyst cavity in hydatid cysts with calcified wall. In the event of a slightest doubt, medical treatment should be started and patients should be scheduled for follow-up. Besides wall calcification, morphological changes in the form of enlargement should bring surgery into mind.

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