Case Report

Chronic Hydatid Cyst in Malaysia: A Rare Occurence

Md Pauzi Suria Hayati[1], Chan Boon Teck Eugene[2], Bong Jan Jin[3], Isa Mohd Rose[4]

Submitted: 27 Feb 2014
Accepted: 17 Apr 2014

[1] *Department of Pathology, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latiff, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia*

[2] *Department of Parasitology and Medical Entomology, Faculty of Medicine, Jalan Raja Muda Abdul Aziz,50300 Kuala Lumpur, Malaysia. Universiti Kebangsaan Malaysia, Kuala Lumpur, Malaysia*

[3] *Department of Surgery, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latiff, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia*

[4] *Department of Pathology, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latiff, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia*

Abstract

Hydatid cysts are not endemic in Malaysia and are rarely seen. We hereby report a case of hydatid cyst of the liver in a 55-year-old Chinese-Australian lady who presented with a calcified liver cyst and negative hydatid serology. A liver segmentectomy was performed and revealed a well-circumscribed, calcified liver cyst containing only creamy whitish material without the typical daughter cyst. A histological examination revealed different layers of the cyst wall and the presence of loose, calcified scolices without a daughter cyst. The case highlights the importance of considering hydatid cyst in the differential diagnosis of liver cyst even in non-endemic areas, as the ease of travelling and migration allows the condition to be seen outside the endemic region.

Keywords: *hydatid cyst, Malaysia, histology, hepatic echinococcosis*

Introduction

A cystic lesion of the liver may present a diagnostic and therapeutic challenge to the attending physician. Many are found incidentally during imaging studies without any symptoms. A large liver cyst may sometimes be associated with serious consequences and morbidity. Apart from the usual symptoms of abdominal pain and possibly jaundice, a large liver cyst may also raise the suspicion of malignancy.

There are many type of liver cysts, including simple cyst, hepatobiliary cystadenoma, hepatobiliary cystadenocarcinoma, pyogenic or amoebic liver abcesses, and more rarely hydatid cysts.

Hydatid cyst is an infection cause by the *Echinococcus granulosus* tapeworm that is endemic in Mediterranean, the Middle East, Australia, and the South America (1-3). However, it is extremely rare in Malaysia. We hereby report a rare case of chronic hepatic hydatidosis presenting as a heavily calcified liver cyst with the absence of a daughter cyst. However, the histological features are suggestive of echinococcosis of the liver.

Case Report

A 55-year-old Chinese-Australian lady presented at our centre complaining of upper abdominal pain lasting one year. Further history revealed that she had emigrated from Malaysia to Australia many years ago and worked as an assistant nurse in Australia. There was no significant history of fever or jaundice. There was no history of direct contact with animals, particularly cattle or dog, to suggest a possible source of parasitic infestation. She has no known medical illness.

A blood tests revealed an increased alanine

transaminase (ALT) level (400 U/L), but other liver enzymes and blood parameters were within normal limits. The serology screening for Hepatitis B and C was also negative.

A chest radiograph showed a raised right hemidiaphragm, but no focal lung lesion was detected. An ultrasonography and computed tomography (CT) scan of the abdomen done in Australia revealed the presence of a peripherally calcified cystic lesion in segment seven of the liver measuring 50 mm in diameter. There was no presence of any internal vascularity or intraductal dilatation.

The finding of a calcified liver cyst raised the suspicion of a hydatid cyst. A hydatid serology test was then performed at Royal Perth Hospital in Australia, but the result was negative. Thus, no medication was started for hydatidosis. Due to financial reasons, the patient opted to obtain further treatment in Malaysia. She was seen by our hepatobiliary surgeon at the surgical clinic. The patient was later scheduled for excision of the liver cyst.

A non-anatomical, wedge resection of segment seven of the liver was performed. Intra-operative findings revealed the presence of a well-delineated liver cyst 50 mm in diameter at segment seven. Dense adhesion was also noted between the diaphragm and the liver. No additional focal hepatic lesion was observed. The patient's post-operative recovery was uneventful and she was discharged well one week later, with further clinical follow up to be done in Australia.

Macrosopically, the liver cyst was surrounded by a capsule 2 mm thick. The cyst contained whitish to yellowish creamy material with no presence of a smaller cyst or of solid areas within the cyst (Figure 1). The surrounding liver tissue was grossly normal.

The histology of the cyst showed a thick capsule composed of different layers of fibrous tissue, a laminated anuclear layer and a degraded and calcified germinative layer (Figure 2). The cystic cavity mainly contained necrotic debris. Admixed within the debris were multiple small rounded empty structures, some of which were calcified. These round calcified structures were loose minute scolices (collectively known as hydatid sand) (Figure 3). Chronic inflammatory cell infiltration was seen in the capsule and the adjacent liver parenchymal tissue. The liver parencymal tissue was otherwise unremarkable. There was no evidence of malignancy.

The gross features of the liver cyst and the histology strongly suggest chronic hydatid cyst of the liver.

Figure 1: The resected segment of liver shows the presence of a well-circumscribed cyst with a calcified wall filled with whitish creamy materials.

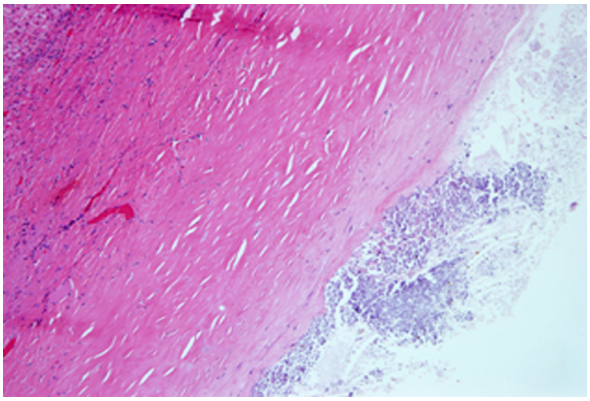

Figure 2: The different layer of the capsule of the cyst with calcified minute scolices within the cavity of the cyst (haematoxylin and eosin staining; 100× magnification).

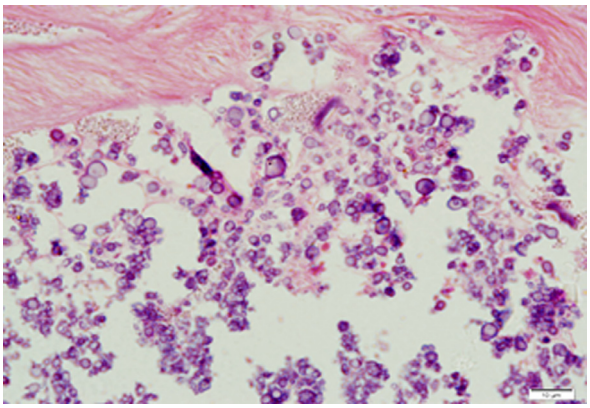

Figure 3: The scolices (hydatid sand) are better seen under high power view showing clusters of calcified empty cystic structures located just underneath the capsule (haematoxylin and eosin staining; 400× magnification).

Discussion

Echinococcosis occurs in humans as the result of ingesting the eggs of the tapeworm Echinococcus granulosus either by eating contaminated food or vegetables or after contact with dogs (1, 2).The definitive hosts are dogs and the intermediate hosts are sheep and humans. Cattle are infected by eating a contaminated grass. The most common sites of cyst development are the liver (60–70%) and the lungs (20–30%), although it may occur in any other organs (3).

Hydatid cysts are rarely observed in Malaysia. Our patient actually resides in Australia, where this disease is considered endemic. Jalleh et al. (1), reported a case of hepatic hydatidosis in Malaysia where the patient presented with acute symptoms of fever and right hypochondrial pain, with typical numerous daughter cysts found upon surgery. In contrast, in our case, the liver cyst showed a calcified wall with only necrotic debris without the presence of a daughter cyst. However, after considering the patient's history and supported by the gross and histological findings, we concluded that the patient had chronic hepatic hydatidosis.

Most patients with hepatic hydatidosis have a benign clinical course, and the majority present with an asymptomatic abdominal mass (4). In symptomatic patients, right hypochondrial pain is the most common symptom, whereas hepatomegaly and right upper quadrant abdominal masses are the most frequent clinical findings (3). A patient may experience symptoms of acute cholangitis when the hydatid cyst ruptures into the biliary system. Lung and thorasic involvement would result in hemoptysis, lower chest pain and a productive cough. Bilioptysis may also occur when the lesion is complicated by a biliobronchial fistula (5). Our patient presented with a long history of abdominal pain but no acute symptoms of fever or jaundice. The finding of liver cyst was only noted upon radiologic imaging.

Imaging studies have an important role in the investigation of liver cysts, helping to differentiate between the various possible liver lesions. However, in many cases, particularly in suspected liver infection, the radiologic findings are not sufficiently specific to help identify the type of infection. Thus, an aspiration or biopsy may be needed for final conclusion (2). A clinical history and epidemiologic information are also crucial in making an accurate presumptive diagnosis (2).

Various classifications are proposed to categorise hydatid cysts depending on the ultrasonographic appearance and pattern. Gharbi et al. proposed a morphologic classification of hydatid cyst ranging from type I (with pure fluid collection) to type V (reflecting a thick cyst wall). According to the five categories proposed by Gharbi, types II and III are characteristic of hydatid cysts, type I and V are suggestive of hydatid cysts in endemic areas and type IV simulates a pseudotumour (5,6). Lewall et al. later introduced a classification of hydatid cysts reflecting the pathology and natural history of the disease (7). Taking into account the fact that our patient came from Australia where echinococcosis is considered endemic, the presence of a thick wall and densely calcified liver cysts (which may be classified as Gharbi type V and Lewall's type III, respectively) is strongly suggestive of hydatid cyst of the liver.

Serologic studies are usually performed to supplement the radiological findings in the diagnosis of hydatid cyst. The test detects antibodies to hydatid cyst fluid-derived native or recombinant antigens that have high sensitivity for hepatic (85–95%) but lower sensitivity (50–60%) for lung echinococcosis (8). These serologic studies have largely replaced the Casoni intradermal test (1, 3). The major disadvantage of this serological techniques is lack of species specificity, with a cross-reaction occurring with other parasitic infections (fascilosis, schistosomiasis, amebiasis, cysticercosis, and filariasis) (8).

A general consensus points towards an association between a heavily calcified liver cyst and negative serology (9). This was likely the reason for a negative hydatid serology test in our case, as the cyst wall was found to be intact. Therefore, there was possibly no leakage of parasitic-derived materials that may stimulate her body to produce the antibody.

Histologically, a typical hydatid cyst is composed of three layers: the outer pericyst, which corresponds to fibrosed and compressed liver tissue; the middle endocyst, which is an inner germinative layer; and the ectocyst, a thin, translucent interleaved membrane. Maturation of the cyst is associated with endocyst invagination forming multiple daughter cysts at the periphery. Peripheral calcification can be observed in both viable and non-viable cysts (2).

The spontaneous death of a hydatid cyst can occur and is considered a natural healing process (10). The possible reasons for this are host's immune reaction and infection of the cyst. Antibodies produced by the infected person against the hydatid antigen may cause cyst degeneration and calcification of its wall (10). A patient with a degenerated cyst is usually

asymptomatic. The death of a hydatid cyst can also be caused by infection that can reach the cyst through the biliary system. This ascending infection can result in the death of the germinal membrane and the scolices in the cystic fuid (10).

A dead cyst will become calcified. As a general rule, a calcified cyst is considered dead, but unfortunately, there is no serological test to prove this (10). A serology test can be positive for years, even after the treatment or death of a cyst, with no definite time frame for the serology to revert back to negative (10). Erzurumlu et al. (10) reported a case of a calcified hydatid cyst that was followed up for eight years without any treatment. However, this patient later presented with intrabiliary rupture of the cyst contents. Therefore, it is possible that a cyst is still viable even though its wall is calcified.

The options for treatment of hydatid cysts include non-surgical and surgical approach. The non-surgical option is chemotherapy, and the surgical options include percutaneus aspiration of the liver cyst and liver resection. A combination of pre- and post-operative albendazole or mebendazole therapy reduces the risk of recurrence (3,4).

Conclusion

In conclusion, although the disease is not endemic in Malaysia, the ease of travelling and the migration of people from other countries allows hydatid cysts to be encountered in non-endemic area. Thus, a differential diagnosis of echinococcosis must always be considered in any case of cyst of the liver. An accurate diagnosis can be made by correlating the clinical history, radiological appearance, serology test and its histological features. This is important in deciding patient management and clinical follow–up, with a reduction in rate of recurrence, morbidity, and mortality.

Acknowledgement

The authors would like to acknowledge Professor Datin Dr Norhayati Mokhtar from Department of Parasitology & Medical Entomology, Universiti Kebangsaan Malaysia for her expert opinion in this case.

Conflicts of Interest

None.

Funds

None.

Authors' Contributions

Conception and design: SHMP
Drafting of the article: SHMP, IMR
Critical revision of the article for the important intellectual content, final approval of the article: SHMP, IMR, CBTE
Provision of study materials or patient: BJJ

Correspondence

Dr Suria Hayati Md Pauzi
MD, Doctor of Pathology (Anatomic Pathology) (UKM)
Department of Pathology
Universiti Kebangsaan Malaysia Medical Centre
Jalan Yaacob Latiff, Bandar Tun Razak
56000 Cheras, Kuala Lumpur, Malaysia
Tel: 603-91455356/5357
Fax: 603-91737340
Email: su_hayati@yahoo.com

References

1. Jalleh R, Nuruddin R, Krishnan M. Hepatic hydatidosis: diagnostic and therapeutic aspects. *Sing Med J*. 1989;**30**:210-212.

2. Mortelé KJ, Segatto E, Ros PR. The Infected Liver: Radiologic-Pathologic Correlation. *Radiographics*. 2004;**24(4)**:937–955. doi: 10.1148/rg.244035719.

3. Krokos N, Mihailidou E, Margioulas A, Karakatsanis A, Drizis T. A giant hydatid cyst of the liver. *Hellenic J Surg*. 2011;**83(2)**:102–106. doi: 10.1007/s13126-011-0019-0.

4. Arif SH, Wani NA, Zargar SA, Wani MA, Tabassum R, Hussain Z, et al. Albendazole as an adjuvant to the standard surgical management of hydatid cyst liver. *Intern J Surg*. 2008;**6(6)**:448–451. doi: 10.1016/j.ijsu.2008.08.003.

5. Dziri C, Haouet K, Fingerhut A. Treatment of hydatid cyst of the liver: where is the evidence? *World J Surg*. 2004;**28(8)**:731–736. doi: 10.1007/s00268-004-7516-z.

6. Gharbi H, Hassine W, Brauner M, Dupuch K. Ultrasound examination of the hydatic liver. *Radiology*. 1981;**139(2)**:459–463. doi: 10.1148/radiology.139.2.7220891.

7. Lewall DB, McCorkell S. Hepatic echinococcal cysts: sonographic appearance and classification. *Radiology*. 1985;**155(3)**:773-775. doi: 10.1148/radiolog .155.3.3890008.

8. Schweiger A, Grimm F, Tanner I, Müllhaupt B, Bertogg K, Müller N, et al. Serological diagnosis of echinococcosis: the diagnostic potential of native antigens. *Infection*. 2011:**40(2)**;1–14. doi: 10.1007/s15010-011-0205-6.

9. Scherer K, Gupta N, Caine WP, Panda M. Differential diagnosis and management of a recurrent hepatic cyst: a case report and review of literature. *J Gen Intern Med*. 2009;**24(10)**:1161–1165. doi: 10.1007/s11606-009-1062-1.

10. Erzurumlu K, Polat C, Dicle K, Kiliç E, Güngör B, Hökelek M. Is Calcified Hydatid Cyst Absolutely Dead? J *Experim Clin Med*. 2011;**27(3)**.