Hydatid Cyst of the Liver: A Case Report and Literature Review

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Taiwan is nearly free from hydatid disease. We report a case of hydatid cyst of the liver in a 37-year-old man who originally lived in India and had migrated to Taiwan 2 years earlier. He presented with right upper quadrant pain and intermittent low-grade fever. Both sonography and computed tomography (CT) demonstrated a cystic lesion with vesicles at its periphery in segments 6 and 7 of the liver. A hydatid cyst was diagnosed. The patient underwent radical excision of the cyst with total removal without opening the wall. He also received pre- and postoperative oral mebendazole. Pathology showed a hydatid cyst consisting of three layers: the inner single nucleated geminal layer, the middle acellular laminated layer, and the outer pericyst originating from inflammatory and hepatic cells. This case highlights that accurate preoperative diagnosis of hydatid disease can be made from personal history, typical sonography and CT study in non-endemic areas.

Key Words: hydatid cyst, *Echinococcus granulosus*, computed tomography, sonography, color Doppler sonography
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Echinococcus infection is commonly seen in humans and may cause hydatid cysts in the liver. This infection is an important public health problem in livestock grazing regions of the world, particularly in Central Europe, Africa, South America, New Zealand, Australia, Central Asia and China, but not in Taiwan. Life-threatening complications may develop if the infection is not treated properly. To date, only three cases of hydatid disease have been reported in Taiwan [1–3]; preoperative diagnosis was not made in any of these patients. We report on an immigrant male patient who had a hydatid cyst of the liver without specific symptoms and signs, but with typical sonography and computed tomography (CT) findings, including daughter cysts and internal septa. Accurate diagnosis and radical surgical resection with pre- and postoperative mebendazole led to a successful cure.

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Case Presentation

A 37-year-old Tibetan man was healthy until he experienced intermittent pain in the right upper abdomen, with low-grade fever and headache for about 2 years prior to admission. He lived in Tibet and then toured temples in India for 4 years before immigrating to Taiwan. When he visited our outpatient department, his body temperature was 36.5°C and he had mild right upper abdominal tenderness without evidence of liver enlargement on vertical span. His cardiovascular system and respiratory parameters were unremarkable. No other symptoms, such as nausea, vomiting, diarrhea, or skin rash, were noted. His white blood cell count was 4,500/mm^3, with 65% neutrophils and 2.6% eosinophils. Urine and stool analyses were

unremarkable. Biochemical tests for alkaline phosphatase, gamma-glutamyltransferase, aspartate aminotransferase, and bilirubin were within normal limits, but alanine aminotransferase was increased to 50 U/L. Tests for anti-hepatitis C virus antibody, hepatitis B surface antigen (HBsAg), and anti-HBsAg antibody were negative. Plain chest X-ray examination showed no abnormal findings.

Due to the abnormal liver function test and right upper abdominal pain, abdominal sonography was performed. A cystic mass in the right lobe of the liver was revealed. The mass measured 6.5 × 5.5 cm, with multiple cystic components presenting a honeycombed appearance (Figure 1). Some echogenic material was seen in the center of the mass. Color Doppler sonography revealed hypovascularity of the lesion. Pre- and post-intravenous contrast-enhanced dynamic enhanced CT scan of the upper abdomen demonstrated a well-defined non-enhanced mass measuring 7 cm in diameter in segments 6 and 7 of the liver (Figure 2). Both sonography and CT showed daughter vesicles at the periphery of the lesion. On the basis of the imaging findings and the patient's history, a diagnosis of hydatid cyst of the liver was made. Due to his headache, the patient underwent brain CT examination, but no significant abnormality was found. Sonoguided biopsy was not performed because of a fear of anaphylactic shock induced by leakage of the cystic content.

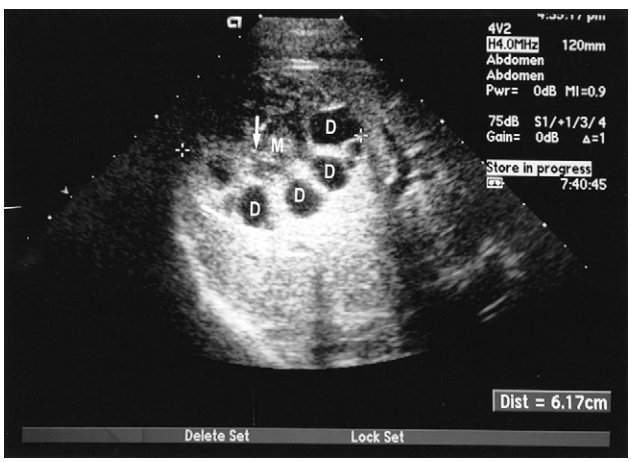

Figure 1. *Sonographic images show four distinct daughter cysts (D) with typical peripheral location within the mother cyst and a hydatid matrix (M) with a mixed echogenic pattern. Note the echogenic linear structures within the matrix representing collapsed membranes (arrow).*

Because there was only a single lesion in the liver, a decision was made to perform radical surgical resection of the mass 6 days after admission. Preoperative oral mebendazole 200 mg four times daily was administered for 4 days. Right posterior segmentectomy (S6 and S7) of the liver revealed a cystic lesion measuring about 7 cm. Incisions in the lesion allowed visualization of an encapsulated unilocular cystic space containing daughter cysts filled with gel-like fluid (Figure 3). Histopathologic study of the lesion showed four components in the wall: the outermost layer of the liver parenchyma infiltrated with non-specific chronic inflammatory cells; a middle layer of acellular laminated membrane; an inner layer consisting of a thin nucleated geminal membrane; and the innermost part containing ghost cells in the cysts (Figure 3). No scolices were identified in the lesion. The histopathologic features were compatible with hydatid cyst.

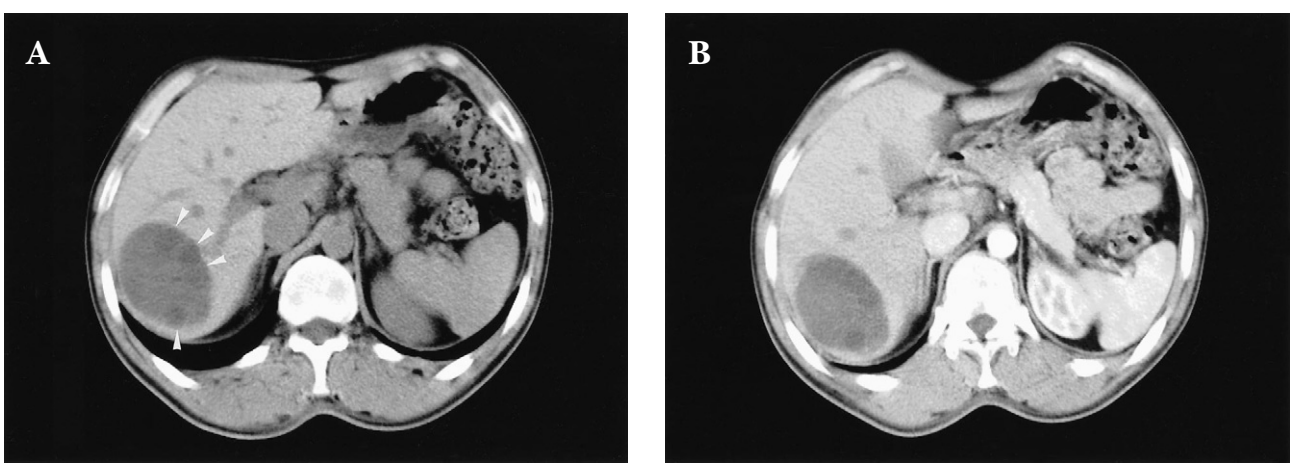

Figure 2. *(A) Unenhanced computed tomography (CT) scan shows an ovoid low-density mass in the right lobe of the liver. The mass has the typical peripheral location of daughter cysts (arrowheads) within the mother cyst. There is no definite calcification of the pericyst. (B) Contrast CT scan reveals no obvious enhancement in the mass.*

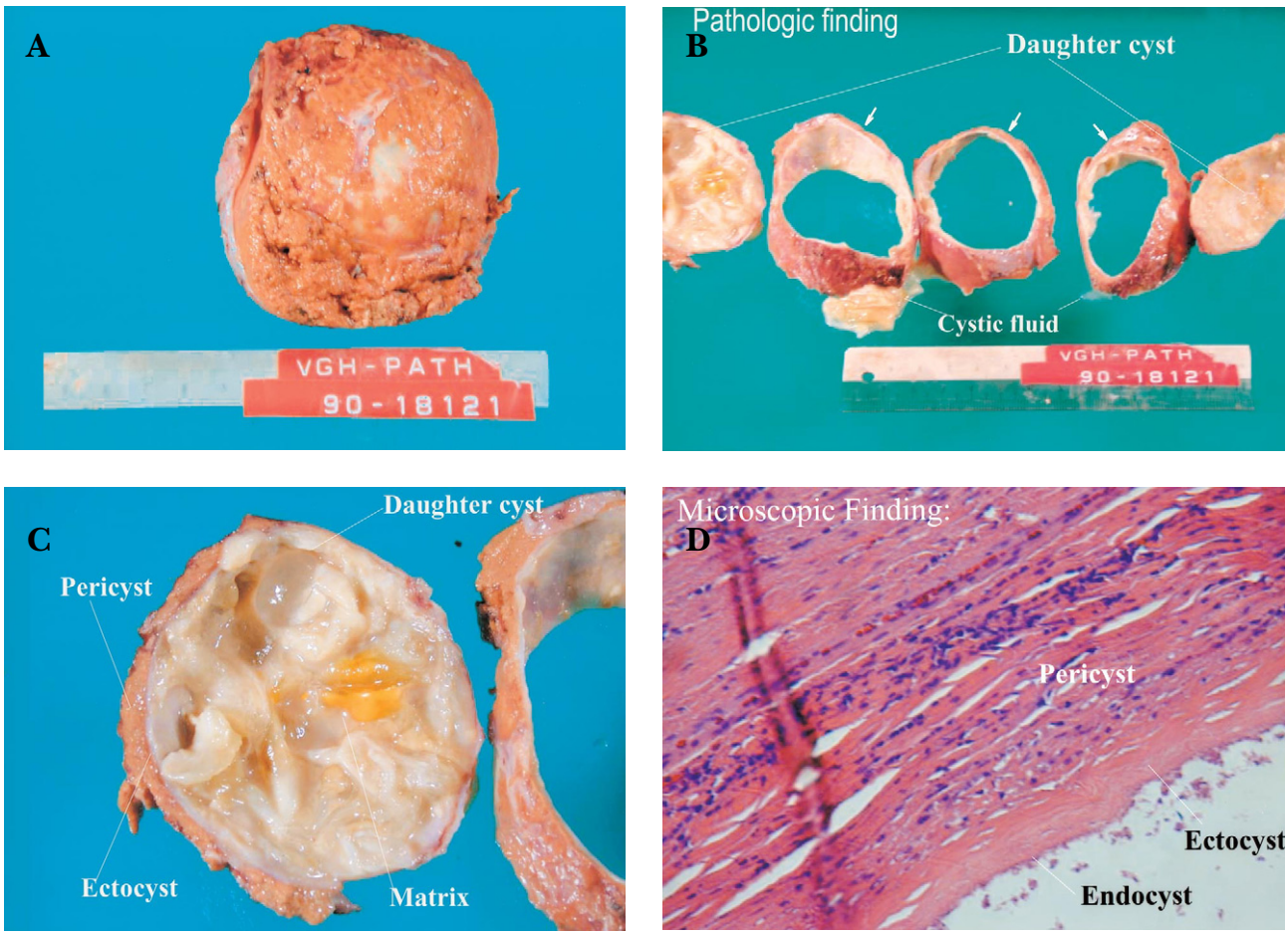

Figure 3. *(A) The specimen consists of brown liver tissue measuring 9 × 7 × 4.5 cm. (B) A large and encapsulated unilocular cystic mass is divided into five sections and contains daughter cysts filled with gel-like fluid. The outer pink layers of the central three sections (arrows) are pericysts. (C) Gross findings of one section reveal that the outer pink layer is the pericyst, the white layer is the ectocyst, and orange material is the matrix. There is a daughter cyst at the internal periphery of the mass. (D) Microscopic findings show multilocular cysts of finely and amphophilic laminated membrane (pericyst) and geminal layers (endocyst) surrounded by fibrinous walls (ectocyst). No scolices are identified in the cysts (hematoxylin & eosin, × 200).*

The patient recovered uneventfully, and there were no postoperative complications except for wound pain, which was treated conservatively. The patient was discharged in good health 2 weeks later. Long-term postoperative mebendazole 200 mg four times daily was prescribed. The patient was still well without imaging or clinical evidence of recurrent hydatid cysts 6 months after surgery.

Discussion

Hydatid disease is a parasitic tapeworm disease caused by the larval stage of *Echinococcus granulosus* or *Echinococcus multilocularis* [4,5]. The liver is the most frequently involved organ in both forms, followed in frequency by the lung [6,7]. However, these two species of *Echinococcus* are quite different, with different endemic regions and clinical manifestations. *E. granulosus* tends to produce a typical large, single, round or ovoid, well-defined hydatid cyst and is the more prevalent species [8]. It is most common in sheep- and cattle-raising areas, such as Central Europe, Africa, South America, New Zealand, Australia, Central Asia, and China. In contrast to *E. granulosus*, *E. multilocularis* is restricted mainly to cold and high-altitude regions, with a higher prevalence in Alaska, Canada, Central Western Europe, Siberia, and Japan. It causes an irregular, small, fluid-filled cavity or an invasive spongy mass resembling an infiltrating malignant hepatic tumor, rather than a well-defined expansile cyst [8,9]. There have been three sporadic reported cases of hepatic hydatid cysts in Taiwan [1–3]. Two were caused by *E. granulosus* and one by *E. multilocularis*. One of the two *E. granulosus* cases showed

complicated hydatid hepatic cysts with superimposed infection, and the other showed uncomplicated multilocular cysts with focal calcification. The third case was of complicated *E. multilocularis* hydatid liver disease, which manifested with a calcified mass that occupied the right hepatic lobe with extension to the left hepatic lobe. It invaded the diaphragm, right adrenal gland, inferior vena cava, and right portal vein.

All of these cases were treated using radical resection without accurate diagnosis before surgery [1–3]. The present case is the first in Taiwan to be diagnosed as a hepatic hydatid cyst by imaging before hepatic resection. The liver lesion was a unilocular, ovoid, well-walled marginated cystic lesion, compatible with those developed due to *E. granulosus* infection of the liver. Hydatid cysts of the liver grow to 1 cm during the first 6 months and 2–3 cm annually thereafter, depending on host tissue resistance [4]. This suggests that this patient may have been infected in India about 2 – 3 years prior to presentation.

The clinical symptoms of hepatic hydatidosis are highly variable and diverse [10]. Uncomplicated hydatid cysts of the liver may remain asymptomatic for years or even decades, depending on the size and site of the developing cyst [5]. In cases of uncomplicated hydatid cyst, pain in the right upper quadrant is the most important diagnostic symptom for the disease [5]. In a review of 157 patients with hydatid disease of the liver, hepatomegaly was the major sign in hepatic hydatidosis [11]. Other symptoms of hydatid disease of the liver include nausea, dyspnea, and dysphagia. Jaundice may occur, depending on the local mass effect of a gradually enlarging cyst or the rupture of a hydatid cyst into the biliary tree. Fever and chills and severe anaphylactic shock are usually absent unless cysts become secondarily infected or rupture [4]. We do not consider that the fever in our case resulted from infected or ruptured hydatid cyst because sonography, CT, and pathologic study did not show dilatation of biliary trees, air bubbles, increased blood flow, or ring enhancement.

Eosinophilia (> 3%) may or may not be present and has been cited as a diagnostic criterion [11]. It is an inconsistent sign in cases with uncomplicated hydatid cysts, but may be helpful in cases with complicated cyst. In our case, the eosinophil cell count (2.6%) was within the normal range. Alkaline phosphatase is elevated in proportion to the volume of the cyst, and is constantly elevated in all complicated cases [10]. The serum alkaline phosphatase level in our patient was not elevated. We consider our patient an uncomplicated case.

There is no single serologic test that definitively establishes the diagnosis before imaging and surgery [9,12]. Even so, immunologic studies are helpful when imaging studies are inadequate in distinguishing hydatid cysts of the liver from pyogenic abscesses or cystic neoplasms [11–13]. Serologic studies were not performed in our case because the findings on sonography and CT images were indicative of a hydatid cyst.

Lewall suggested that the hydatid cyst always starts as a fluid-filled, cyst-like structure (stage I), which may proceed to a stage II lesion if daughter cysts and/or matrix develop [14]. In some instances, the stage II lesion becomes hypermature and, due to starvation, dies to become a mummified, inert, calcified, stage III lesion. When formed elements completely replace the nourishing hydatid fluid, the stage II lesion is starved, dies, and eventually becomes a calcified and biologically inert stage III lesion [14]. The pathology of our present case revealed a typical hydatid cyst containing daughter cysts filled with gel-like fluid (Figure 3). Microscopic findings on sections showed mutilocular cysts with an outer-layer ectocyst and a single-layer endocyst surrounded by an acellular laminated pericyst. No scolices or proscolices were identified in the cysts, except necrotic tissue and some ghost cells. There are three possible causes to explain the loss of proscolices and scolices. First, the patient received preoperative treatment with mebendazole; however, the 4 days of treatment was insufficient to successfully sterilize the cyst. Second, the hydatid cyst had been infected, but laboratory data, sonography, and CT did not support this possibility. Third, the lesion was a Lewall stage II hypermature hydatid cyst with a dead space due to starvation. We believe the third cause might explain the loss of proscolices and scolices.

E. granulosus infection of the liver is readily detected on sonography as a cystic cavity, mostly in the right lobe [6]. In approximately 50% of cases, there are daughter cysts [15]. Before the budding of daughter cysts begins, the lesion is indistinguishable from a simple cyst on sonography [15]. The sonographic features of hepatic cystic echinococcosis can be classified into five types [13,16]: I) univesicular cysts with pure fluid collection; II) fluid collection with a split wall and very marked enhancement of back-wall echoes; III) fluid collection with daughter cysts and a visible laminated layer; IV) cysts with heterogeneous echo patterns; and V) cysts with reflecting thick walls and varying degrees of calcification. The hydatid cyst in the present case can be classified as Type III (Figure 1).

CT is an accurate means of diagnosis for hepatic hydatid disease [10], and is especially helpful when sonography

fails, owing to patient-related difficulties such as obesity, excessive intestinal gas, abdominal-wall deformities, previous surgery, or disease complications [4]. However, CT imaging in this patient provided less information about daughter cysts, internal septa, and detached membranes in the matrix than sonography because the density of the cyst fluid was relatively high and the septa or detail was lost on CT images (Figure 2).

The differential diagnoses of hepatic hydatid cyst include liver abscess, sarcoma, and metastastic tumor. Hydatid cyst of the liver without marginal calcification should be differentiated from other hepatic cystic low-density lesions such as pyogenic liver abscess, hepatic cysts with infection, hemangioma, mesenchymal hamartoma, biliary cystadenoma/cystadenocarcinoma, liver metastasis, and hepatic sarcoma. The differential diagnosis is dependent on the margin and contrast enhancement of the hepatic mass on CT and sonography, and the gender of the patient.

Surgical removal of liver hydatid cysts remains the preferred method of treatment [13,17], using either radical or conservative surgery [2]. Conservative surgery, including capitonnage, partial pericystectomy, and cystojejunostomy, results in complete removal of the endocyst and cyst contents, but not of the pericyst [3]. Our patient underwent major liver resection over posterior segments 6 and 7 with pre- and postoperative mebendazole medical treatment. No surgical complications such as local recurrence or anaphylactic shock were detected.

According to the World Health Organization guidelines, chemotherapy with mebendazole or albendazole is the preferred treatment when the disease is inoperable, when surgery or percutaneous aspiration and injection of hypertonic saline (PAIR) is not available, or when the cysts are too numerous [18]. PAIR is used to kill all viable protoscolices or separate them from the pericyst within 5 to 10 minutes [13,15]. Indications for PAIR include Type I–IV hydatid liver cysts that contain a considerable amount of fluid, or an infected hydatid cyst without communication with biliary trees [18]. Chemotherapy combined with percutaneous drainage has been reported to achieve better results in the treatment of uncomplicated cyst [2]. However, PAIR was not indicated in the present case because the preoperative diagnosis of hydatid cyst was not confirmed and the cyst contained considerable non-drainable matrix.

In conclusion, hydatid disease of the liver can occur worldwide and rare cases have been diagnosed in non-endemic areas. Due to increased chances of travel, immigration, and contact with domestic pets and wildlife, the specific sonographic and CT appearances of hydatid cysts should be kept in mind whenever a hepatic cystic lesion is encountered. Aggressive surgical intervention concomitant with oral mebendazole or albendazole, before and after surgery for uncomplicated hydatid liver cysts, is still preferred in the treatment of this disease.

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肝胞蟲囊病：病例報告及文獻回顧

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台灣幾乎沒有本土感染之胞蟲囊病。我們報告一個原本住在印度並且移居到台灣兩年的肝胞蟲囊病個案。他表現出右上腹痛以及間歇性輕微發燒。超音波以及電腦斷層影像顯示一個位於肝臟第六及第七節周邊囊狀病兆，病兆周圍內側有數個水泡。基於上述特殊影像表現以及病人經歷，因而診斷為肝胞蟲囊病。病人之後接受全病兆切除且並未將病兆外壁破壞。病理結果顯示病兆有三層：最內層是單一有核細胞增殖層，中間是無細胞多層，第三是最外層由發炎細胞及肝細胞組成。病人經由結合術前及術後口服 mebendazole 加上全病兆切除手術後得到成功治療。這個案點出一項明顯的事實 — 就是經由個人旅行史，病徵及典型超音波以及電腦斷層影像研究可以在非流行地區對胞蟲囊病於術前做出正確診斷。

關鍵詞：胞蟲囊，粒層性胞蟲囊，電腦斷層影像，超音波影像，彩色都卜勒超音波

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