

Evaluation of the Clinical, Laboratory, and Radiological Findings and Treatment of 19 Cases of Pancreatic Echinococcosis

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Background. Hydatid disease is a severe health problem in endemic areas. In recent years, the incidence of this disease in China has been increasing. As the imaging characteristics of pancreatic echinococcosis (PE) are similar to those of cystic diseases, such as cysts, tuberculosis, and tumors, PE is often misdiagnosed and mistreated.

Methods. The clinical manifestations, laboratory tests, radiological findings, and treatment of 19 patients with PE between January 2006 and December 2018 in 2 hospitals were retrospectively analyzed.

Results. The mean age of the patients was 38 years, and the ratio of women to men was 2. All patients came from rural areas. Clinical manifestations included cholestatic jaundice, mass, nausea, pain, and swelling. Hemagglutination inhibition test results were positive for all patients. Enzyme-linked immunosorbent assay test results were positive in 17 cases (89.5%). Foci in the pancreas were head in 52.6%, body in 26.3%, body and tail in 15.8%, tail in 5.3%. The size of lesions' diameter ranged from 1 to 12 cm (mean, 6.5 cm). The imaging features of PE included the presence of (a) daughter cysts on abdominal computed tomography (CT) and/or magnetic resonance imaging (MRI); (b) internal cyst wall dissection and ribbon sign on abdominal CT and/or MRI; (c) typical egg-shell cyst wall calcification on abdominal CT.

Conclusions. For patients with cystic lesions on CT and/or MRI combined with epidemiological history and positive echinococcosis serology, doctors can correctly diagnose PE earlier. Surgical treatment combined with drugs can reduce the mortality of PE, leading to a better prognosis.

Keywords. echinococcosis; hydatid; human; pancreas.

Zoonosis hydatid disease (cystic echinococcosis or alveolar echinococcosis) is most often caused by infection of the taeniid tapeworm with *Echinococcus granulosus* larvae. This zoonosis disease is distributed worldwide and represents a significant public health problem in China [1].

Previously, human hydatid disease was considered a rare disease; however, in recent years it has increased in incidence. The liver and the lungs are the most frequently involved organs in primary hydatidosis (70% and 20%, respectively). In contrast, pancreatic echinococcosis (PE) is a rare entity with an incidence ranging from 0.14% to 2% of patients with hydatidosis [2]. As the imaging characteristics of PE are similar to those of cystic diseases, such as cysts, tuberculosis, and tumor, PE is

often misdiagnosed and mistreated. Rupture of a hydatid cyst may lead to anaphylaxis. Preoperative diagnosis is critical, as the treatment modalities of other causes of cystic lesions are entirely different. From January 2006 to December 2018, a total of 19 patients with PE were treated in our department. Here we report the clinical, laboratory, and radiological findings and treatment.

METHODS

Study Design

We retrospectively analyzed the collected data of all patients diagnosed with PE in Xinjiang Uyghur People's Hospital and Xinjiang University Affiliated First Hospital between January 2006 and December 2018. The data collected included clinical features, laboratory tests, radiological findings, and treatment.

Study Methodology

All patients admitted to the hospital provided a medical history and underwent a clinical examination and laboratory tests, including measurements of serum agglutination and blood culture for echinococcosis, as well as purified protein derivative test, T-SPOT, and other tests to exclude other diseases.

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All patients also underwent computed tomography (CT), and 7 patients underwent magnetic resonance imaging (MRI). Sixteen cases were investigated with contrast-enhanced CT scans. Seven patients were examined with a MR short-TI-IR (STIR) scan, and 2 patients underwent magnetic resonance cholangiopancreatography (MRCP). CT and MRI examinations were performed with 1 week of hospitalization. Close contact with a dog or sheep was defined as having a dog or sheep at home.

Diagnosis of PE was made according to one of the following criteria: (1) The diagnosis was confirmed on histopathological examination. (2) The hemagglutination inhibition and enzyme-linked immunosorbent assay (ELISA) test results were positive.

All patients were followed up after leaving the hospital by either telephone or readmission. All patients had been followed up for at least 1 year.

RESULTS

Demographic, Epidemiological, and Clinical Symptoms

Demographic characteristics, clinical presentation, and location of PE are summarized in Table 1. Of the 19 patients, 6 patients were male and 13 were female. The mean age of the patients (range) was 38.32 ± 17.65 (13–67) years. All patients lived in rural areas of Xinjiang and included 9 farmers, 9 herdsman, and 1 student. Fourteen patients had a history of close contact with dogs and sheep. There were 7 cases with a surgical history of hepatic hydatid.

Of 19 PE patients, cholestatic jaundice occurred in 10 cases (52.6%), abdominal mass in 7 cases (36.8%), nausea in 6 cases (31.6%), abdominal pain in 5 cases (26.3%), swelling in 3 cases (15.8%), and no clinical symptoms in 3 cases (15.8%).

Laboratory Tests

All patients were also evaluated with serological tests (hemagglutination inhibition and ELISA techniques) to determine the presence of PE. The hemagglutination inhibition test results were positive for all patients. The ELISA test results were positive in 17 cases (89.5%).

Imaging Findings

An abdominal CT was performed for all patients during the initial examination. The location, size, extrapancreatic location, and complications of PE are summarized in Table 1. Foci in the pancreas were head in 52.6%, body in 26.3%, body-tail in 15.8%, and tail in 5.3%. CT scans showed all cases of the cysts as hypodense space-occupying lesions, and the size of lesions' diameter ranged from 1 to 12 cm (mean, 6.5 cm). Sixteen cases were a single cystic with thin walls, and 3 cases appeared as a multiseptated cyst (Figure 1). Two hydatid cysts had an internal cyst wall dissection and presented ribbon sign (Figure 2). The cyst wall of 2 hydatid cysts had curved calcification (Figure 3A).

Eight patients also had intrahepatic or intralung manifestations of echinococcosis, including liver involvement ($n = 5$; 3 hepatic cystic echinococcosis, 2 hepatic alveolar echinococcosis) and lung involvement ($n = 3$). There were 2 cases of dilated biliary ducts. In 16 cases that were investigated with contrast-enhanced CT scans, hydatid cysts showed no enhancement in 3-phase. Angiography of 3 cases of PE showed significant displacement of vascular compression in the arterial stage (Figure 3B).

Seven cases had a complete record of MRI examination. Three cases were located in the pancreatic head (Figure 4A), 3 in the body and 1 in the body-tail. Six single cystic lesions showed a hypointense signal on T1-weighted images, a hyperintense signal on T2-weighted images, and an obviously hyperintense signal on MR STIR (Figure 4A). One case of multiple cystic lesions manifested grape cluster sign and showed up better on MR STIR. MRCP imaging for defining the lesion and its relationship with the pancreatic duct was performed in 2 patients. In 2 cases with head lesions, the bile ducts were dilated, including the common bile duct, the common hepatic duct, the pancreatic duct, and the gallbladder (Figure 4B).

Treatment and Follow-up

All patients who underwent surgical treatment received 400 mg of albendazole daily 1 month before surgery. The surgical procedure, outcome, and follow-up time of PE are summarized in Table 1. The complications of the surgical treatment were abscess formation in the residual cavity (2), pancreatic fistula (1), and postsurgical wound infection (1). We treated them accordingly. All cases underwent pathological detection, and the results of pathology were 17 cystic echinococcosis and 2 alveolar echinococcosis. After surgical treatment, all patients were treated with albendazole, praziquantel, and nitazoxanide (10 mg/kg body weight/d) for 6 months. All patients were followed up for at least 1 year and were evaluated by routine examination every year. The mean follow-up time (range) was 27.6 (12–56) months. A cystic echinococcosis patient (Case 7) relapsed and underwent surgical treatment.

DISCUSSION

Hydatid disease is a severe health problem in endemic areas. Human PE disease is rare, even in regions where hydatidosis is endemic [3–6]. In recent years, with the development of China's animal husbandry and tourism industries, the incidence of this disease in China is increasing. In this study, there was the most significant number of cases with PE in China.

Human PE can happen only when a person consumes vegetables contaminated by the excreta of infected stray dogs. Most cysts are acquired in childhood and are not diagnosed until adulthood; only about 10%–25% of cases present in childhood [7]. In the current study, the mean age of the patients was 38 years, and those aged <18 years accounted for 0.36% of all

Table 1. Summary of Demographic and Clinic Characteristics and Follow-up in Patients (n = 19) With PE

Case No.	Sex	Age	Ethnicity	ELISA	Radiology	Location	Size, cm	Complication	Surgical Procedure	Recurrence	Follow-up, mo
1	F	27	Han	+	CT	Body ^a	2 × 1	NS	RS	No	16
2	F	62	Kazak	-	CT	Body ^a	1 × 1	NS	RS	No	21
3	F	42	Uighur	-	CT+MRI	Head	8 × 5	NS	PD	No	32
4	F	35	Han	+	CT	Tail	12 × 9	NS	PD	No	18
5	F	49	Uighur	+	CT+MRI	Body	4 × 4	NS	RS	No	26
6	M	28	Tibetan	+	CT+MRI	Body ^b	3 × 3	NS	RS	No	38
7	F	34	Han	+	CT	Head	4 × 2	NS	PD	Yes	43
8	F	45	Kirgiz	+	CT+MRI/CP	Body-tail	6 × 4	NS	PD	No	23
9	M	13	Kirgiz	+	CT	Body-tail	8 × 6	NS	PD	No	12
10	M	48	Uighur	+	CT+MRI	Head ^a	1 × 1	NS	PD	No	15
11	M	31	Han	+	CT	Head	5 × 4	NS	PD	No	41
12	F	25	Han	+	CT	Head ^a	2 × 2	NS	RS	No	21
13	F	59	Han	+	CT+MRI	Body	5 × 4	NS	PD	No	29
14	F	37	Uighur	+	CT	Body-tail ^b	7 × 5	NS	PD	No	33
15	F	20	Kazak	+	CT	Head	2 × 2	NS	RS	No	16
16	M	67	Han	+	CT+MRI/CP	Head	4 × 4	NS	PD	No	56
17	F	25	Han	+	CT	Head ^b	4 × 3	Dilated biliary ducts	PD	No	34
18	M	31	Han	+	CT	Head ^a	4 × 4	NS	PD	No	22
19	F	50	Han	+	CT	Head	3 × 3	Dilated biliary ducts	PD	No	29

Abbreviations: CP, magnetic resonance cholangiopancreatography; CT, computed tomography; ELISA, enzyme-linked immunosorbent assay; MRI, magnetic resonance imaging; NS, Not-saw; PD, percutaneous drainage; PE, pancreatic echinococcosis; RS, radical surgery.

^aAccompanied by hepatic echinococcosis.

^bAccompanied by pulmonary echinococcosis.

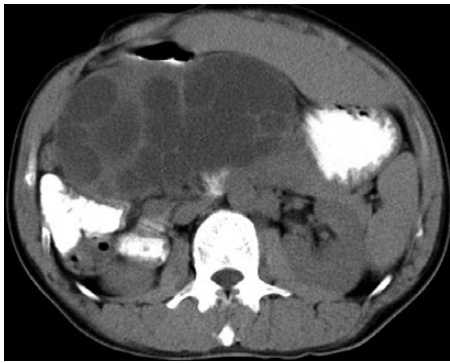


Figure 1. Case 3: Computed tomography axial showed that a bigger hydatid cyst had signs of daughter cysts in the pancreatic head.

cases. This study showed that the ratio of women to men was 2:1, and all patients came from rural areas.

Clinical symptoms varied depending on the anatomical location of the cysts. Cysts in the body or tail were virtually asymptomatic and presented only as swelling with potential for subsequent effects. In the current study, we encountered 3 cases of clinically swollen pancreatic body cysts and 3 cases with no symptoms with cysts in the pancreatic body-tail. Symptoms arise because of pressure on neighboring tissues as the size of the cyst enlarges. Complaints such as abdominal pain, mass, and nausea may arise, although patients may be asymptomatic for a long time [8]. In the present study, as 36.8% had the large size of the cyst, the cysts appeared as an abdominal mass; 31.6% experienced nausea, 26.3% experienced abdominal pain. A similar clinical presentation has also been reported by others [9–11]. The cyst in the head could present with obstructive jaundice due to extrinsic compression of the common bile duct; only a few cases of this nature have been reported in the literature [2]. In the current study, we encountered 10 patients with cysts in the head, presenting with obstructive jaundice. Due to compression of the pancreatic duct from the head cyst, acute pancreatitis might occur [12]. In the current study, we did not encounter a case.



Figure 2. Case 9: Computed tomography axial showed that the cyst wall of a bigger hydatid cyst had internal cyst wall dissection and presented a ribbon sign in the pancreatic body and tail.



Figure 3. A, Case 4: Computed tomography (CT) coronal showed that there was a bigger hydatid cyst of the arc-shape calcification cyst wall in the pancreatic body-tail. B, Case 4: CT angiography coronal revealed that the vessel was significantly compressed and displaced by the hydatid cyst in the arterial stage.

In the current study, the hemagglutination inhibition test results were positive for all patients. The ELISA test results were 89.5% positive. Casoni's intradermal test has been abandoned owing to its relatively poor diagnostic accuracy.

Possible sources of infestation include hematogenous dissemination, local spread via pancreaticobiliary ducts, and peripancreatic lymphatic invasion [13]. The embryo of a hydatid cyst ends up in the pancreas mainly via the arterial route after passage through the liver and pulmonary filters. The relatively higher frequency in the pancreatic head can be explained by the fact that the head region is the most vascularized. In the current study, the head was the most frequent localization of the disease (52.6%), followed by body (26.3%), body-tail (15.8%), tail (5.3%). Similar findings had also been reported by others [2, 8, 14]. PE is usually isolated in 91% of cases [8].

Imaging plays an essential role in identifying pancreatic lesions. Localization within the pancreatic cyst may be based on CT or MRI [15]. CT images are characteristic. CT scans showed all cases of the cysts as hypodense space-occupying lesions, and the mean diameter of lesions (range) was 6.5 (1–12) cm. CT images revealed multiseptated cysts in 3 cases, ribbon sign in 2

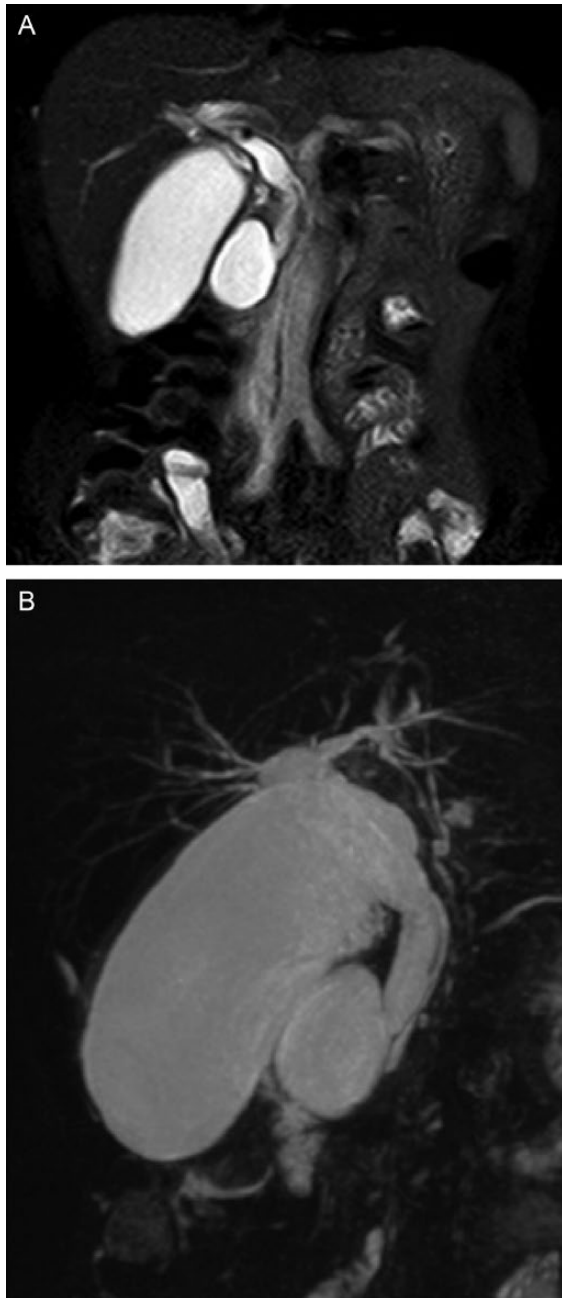


Figure 4. A, Case 16: Magnetic resonance imaging short-T1-IR coronal showed that the bile ducts were dilated, including the common bile duct, the common hepatic duct, and the gallbladder in the pancreatic head. B, Case 16: This is magnetic resonance cholangiopancreatography imaging of the same case.

instances, and curved calcification in 2 cases; these signs led to a correct diagnosis of PE [16, 17]. In the current study, there were 4 cases with concurrent liver echinococcosis and 4 cases with lung echinococcosis; 6 cases had a surgical history of hepatic cystic echinococcosis. The results of CT may suggest diagnosis of PE before surgical treatment. There were no qualitative results of CT in 2 cases before surgical treatment. Moreover, CT reconstruction may provide information about the relationship between the cyst and bile ducts and wall calcification, and

CT angiography can show significant displacement of vascular compression in the arterial stage.

MRI, due to its higher soft-tissue contrast and capability of multiplanar imaging, gives a better depiction of the fluid content of cystic lesions and communication with the pancreatic duct. MRI documented a cystic mass of the head of the pancreas. All hydatid cysts showed a hypointense signal on T1-weighted images and a hyperintense signal on T2-weighted images and an obviously hyperintense signal on MR STIR. One case manifested better grape cluster sign on MR STIR. MRCP imaging for defining the lesion and its relationship with the pancreatic duct was performed in 2 patients. MRCP findings revealed gross dilatation of the common bile duct, the common hepatic duct, the pancreatic duct, and the gallbladder as cysts located in the pancreatic head. MRCP helps delineate the biliary tree and pancreatic duct when the pancreatic cyst is located in the head of the pancreas and causing ductal compression [18].

PE often needed to be differentiated from cystic lesions of the pancreas, including cystadenomas and cystadenocarcinomas, cystic islet cells, papillary cystic tumors, cystic metastasis, pseudocysts, and tuberculosis [19]. The diagnostic features of PE include the presence of (a) the characteristic epidemiological setting; (b) history of a liver and lung echinococcosis; (c) daughter cysts and typical eggshell cyst wall calcification on abdominal CT; (d) internal cyst wall dissection and imaging presenting ribbon sign; or (e) positive echinococcosis serology.

Surgery remains the treatment of choice for hydatid disease [14]. After surgical procedures, all patients were treated with the antihelminthic drug for 6 months. In the current study, the mean follow-up time was 27.6 months, and 1 patient relapsed and underwent surgical treatment again.

This study has 2 limitations. First, this was a retrospective study with a small number of patients. Second, prospective studies with more sample patients are needed to confirm our findings.

CONCLUSIONS

This study is the largest to date studying patients with PE in China. For patients with cystic lesion CT and/or MRI diagnosis combined with epidemiological history and positive echinococcosis serology, doctors can correctly diagnose PE earlier. Surgical treatment combined with drugs can reduce the mortality of PE, leading to a better prognosis.

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Availability of data and materials. The data sets cannot be made publicly available, and restrictions apply to the availability of these data. Data can be requested from the authors and will require permission from Xinjiang Medical University affiliated First Hospital and Xinjiang Uyghur People's Hospital.

Ethics approval and consent to participate. This study was conducted in accordance with the principles of the Helsinki Declaration and was reviewed and approved by the institutional review board for exemptions and exemptions from written informed consent requirements.

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