Acute uremia and intestinal obstruction due to a retroperitoneal hydatid cyst

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Hydatid disease is relatively common in endemic areas. The symptoms due to bulk effect, usually occurring over a certain period of time, depend on the site and size of the cyst. We report an unusual presentation of retroperitoneal hydatid cyst with rapidly developing uremia and acute intestinal obstruction.

Key words: Hydatid cyst, intestinal obstruction

INTRODUCTION

Retroperitoneal and retrovesical localizations of hydatid cyst (HC) disease are rare even in endemic areas. The pouch of Douglas is the preferred site of development for a secondary cyst in the pelvis, initially intraperitoneal and later subperitoneal (1).

The growth of HC is usually slow and asymptomatic, and clinical manifestations are caused by compression of the involved organ (2). We present a case of retroperitoneal pelvic HC that caused bilateral ureterohydronephrosis leading to acute uremia and acute rectal obstruction. To our knowledge, this is the first report of such a clinical manifestation.

CASE REPORT

A 21-year-old male was admitted with oliguria, frequency, mild abdominal pain, constipation and rectal bleeding. A medical examination in another hospital for nausea and vomiting lasting for the last two weeks revealed acute uremia, and he was referred to our emergency ward. His medical history revealed that he was given laxatives for constipation and local therapy for hemorrhoids for one year and that he had undergone abdominal surgery for liver HC disease with cholecystectomy four years ago.

Physical examination revealed mild abdominal tenderness and a mass in the suprapubic area on deep palpation. Digital rectal examination revealed the compression of the rectum. His white blood cell (WBC) was 11,500/ml, 8% of which were eosinophils; his BUN and creatinine were 72 mg/dl and 4.6 mg/dl, respectively. On ultrasound, he had a lower abdominal cystic mass of 15x12 cm, with septations inside. He also had bilateral hydroureronephrosis. Noncontrast computed tomography (CT) revealed that the pelvic mass was compressing the bladder and rectum (Figure 1). Initial diagnosis was uremia due to ureteral compression, and an emergency ureteral stenting was planned.

On cystoscopy, the surgeon was unable to reveal ureteric orifices because of bladder compression. The surgeon decided to place bilateral nephrostomies, after which the clinical condition changed significantly. At the end of the second day, a total of 4000 cc/day of urine was draining via nephros-
Retroperitoneal hydatid cysts are relatively rare and usually occur as a result of spontaneous, traumatic, or surgical rupture of the other organ or hepatic cysts or spillage during surgery. Primary retroperitoneal HC is extremely rare and can be considered only when no other cysts of the same etiology are present. El Ouakdi et al. reported 31 retroperitoneal tumors, six of which were retroperitoneal HC. Angulo et al. reported 10 cases during a 15-year period in an endemic zone, corresponding to 1% of newly diagnosed hydatidosis in that area. In another study, Prousalidis et al. reported 49 uncommon sites of HC, and two of them were in the retroperitoneal space. The case presented here is considered to be a secondary retroperitoneal HC.

Hydatid cysts in humans produce symptoms by two mechanisms: a generalized toxic reaction due to the presence of the parasite itself and local or mechanical symptoms depending on the location of the cyst. The mechanical symptoms depend on the size and the number of the cysts. In more than 40% of the cases, the complications, among which rupture, secondary infection, compressive syndromes and suppuration are the most common, precede the diagnosis of the disease. In retroperitoneal HC, upper urinary tract obstruction may be present at the time of diagnosis, usually causing flank pain and less often renal insufficiency. Digestive symptoms such as constipation or development of hemorrhoids may be confounding. All of these complications are due to the mass effect of the cyst and develop over a period of time, usually without causing acute uremia or acute intestinal obstruction. The case presented here had bilateral ureteral obstruction resulting in bilateral ureterohydronephrosis and acute postrenal azotemia. In addition, the mass resulted in obstruction of the rectum, resulting in constipation and hemorrhoids initially, and acute obstructive symptoms later. Acute onset of all these symptoms gives rise to the question of how rapidly a HC can enlarge. It is known that the embryo develops into HC, reaching a diameter of 1 cm in five or 6 months and thereafter grows according to the stiffness of the surrounding tissue. Knowledge on the growth rate of hepatic (75%) and pulmonary (15%), and only 10% occur in the rest of the body (3). According to Deve, fissuring or rupture of a primary hepatic, splenic or mesenteric cyst would seed its contents in the abdominal cavity. Retroperitoneal HC is usually the result of spontaneous, traumatic or surgical rupture of the other organ or hepatic cysts or spillage during surgery. Primary retroperitoneal HC is extremely rare and can be considered only when no other cysts of the same etiology are present (1). El Ouakdi et al. reported 31 retroperitoneal tumors, six of which were retroperitoneal HC. Angulo et al. reported 10 cases during a 15-year period in an endemic zone, corresponding to 1% of newly diagnosed hydatidosis in that area. In another study, Prousalidis et al. reported 49 uncommon sites of HC, and two of them were in the retroperitoneal space. The case presented here is considered to be a secondary retroperitoneal HC.

**DISCUSSION**

Hydatid disease is one of the oldest diseases known to mankind. The locations are mostly hepatic (75%) and pulmonary (15%), and only 10% occur in the rest of the body (3). According to Deve, fissuring or rupture of a primary hepatic, splenic or mesenteric cyst would seed its contents in the abdominal cavity. Retroperitoneal HC is usually the result of spontaneous, traumatic or surgical rupture of the other organ or hepatic cysts or spillage during surgery. Primary retroperitoneal HC is extremely rare and can be considered only when no other cysts of the same etiology are present (1). El Ouakdi et al. reported 31 retroperitoneal tumors, six of which were retroperitoneal HC. Angulo et al. reported 10 cases during a 15-year period in an endemic zone, corresponding to 1% of newly diagnosed hydatidosis in that area. In another study, Prousalidis et al. reported 49 uncommon sites of HC, and two of them were in the retroperitoneal space. The case presented here is considered to be a secondary retroperitoneal HC.

Hydatid cysts in humans produce symptoms by two mechanisms: a generalized toxic reaction due to the presence of the parasite itself and local or mechanical symptoms depending on the location of the cyst (8). The mechanical symptoms depend on the size and the number of the cysts. In more than 40% of the cases, the complications, among which rupture, secondary infection, compressive syndromes and suppuration are the most common, precede the diagnosis of the disease (5,9). In retroperitoneal HC, upper urinary tract obstruction may be present at the time of diagnosis, usually causing flank pain and less often renal insufficiency. Digestive symptoms such as constipation or development of hemorrhoids may be confounding (5,6). All of these complications are due to the mass effect of the cyst and develop over a period of time, usually without causing acute uremia or acute intestinal obstruction. The case presented here had bilateral ureteral obstruction resulting in bilateral ureterohydronephrosis and acute postrenal azotemia. In addition, the mass resulted in obstruction of the rectum, resulting in constipation and hemorrhoids initially, and acute obstructive symptoms later. Acute onset of all these symptoms gives rise to the question of how rapidly a HC can enlarge. It is known that the embryo develops into HC, reaching a diameter of 1 cm in five or 6 months and thereafter grows according to the stiffness of the surrounding tissue (10). In the liver, the growth rate is approximately 1-2 cm/year (11). Knowledge on the growth rate of
retroperitoneal HC is lacking, mainly because HC
disease is not to be followed up but rather to be
-treated initially. The growth rate can be surveyed
by the reports of under-evaluated or missed cases
of HC.

This case was evaluated for hydatid disease and
was operated for liver HC four years ago. No evi-
dence of retroperitoneal HC was present at that
time. Whether he was under-evaluated at that
time for a small primary retroperitoneal cyst or
had spillage of the disease due to surgery is spec-
ulative, but nevertheless, it is evident that a small
or nonexisting HC became a 15 cm mass in four
years. In the last year of this period there were
mild symptoms of the retroperitoneal mass, and
the acute onset of obstructive symptoms occurred
in less than one month. To our knowledge, this is
the only report of a HC case with acute onset of
uremia and intestinal obstruction in such a short
period of time.

Hydatid disease should be considered in the dif-
ferential diagnosis of all cystic masses in all
anatomical locations, especially in areas where
the disease is endemic (12). The combination of
clinical history, imaging findings and serologic
test results usually aid in the diagnosis
(2,6,7,12,13). Aspiration biopsy may not be as
risky as previously considered, and this procedure
can be risk-free if performed with systemic alben-
dazole therapy (14). In our patient, the diagnosis
was made with CT and fine needle aspiration biop-
sy, and no complications occurred during or after
the biopsy.

For treatment of retroperitoneal HD, total exci-
sion may not be possible because of dense adhe-
sions to major vessels such as iliac vessels. The
choice of surgery should be evacuation of the cyst
and excision of the redundant portion of the peri-
cyst leaving the rest of the cavity open, as in our
case (7).

In conclusion, a retroperitoneal HC may develop
to a diameter of 15 cm in four years. Although not
usual, HC may cause acute uremia and acute
intestinal obstruction with a rapid onset. HC
should be considered in the differential diagnosis
of retroperitoneal masses, even in acute onset of
obstructive symptoms.

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