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Review

Primary renal hydatid cyst – A review

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Abstract

The aim of this study was to analyze the clinical presentation, diagnosis and treatment of primary renal hydatidosis. A thorough review of the literature was made using the PubMed database. Cystic echinococcosis is a disease that results from infection with the larval stage of *Echinococcus granulosus*, a tiny tapeworm found in dogs (definitive host), sheep, cattle, goats, foxes and pigs, amongst others (intermediate hosts).

Humans are accidentally intermediate hosts, who become infected by direct contact with infected dogs, by ingesting contaminated raw or undercooked vegetables or by drinking contaminated water. Renal involvement by hydatid disease is uncommon (less than 3-4%), most cystic lesions occurring in the lungs or liver. Patients may be asymptomatic or they can present with flank pain, hematuria or hypertension. Hydaturia, considered a pathognomonic sign, is seen in less than 20% of cases.

Imaging investigations are essential in making the correct diagnosis, the most commonly used means being ultrasonography and CT scans. Surgery is the mainstay of treatment, but other therapeutic options are also available.

Keywords

: renal, cyst, primary, hydatid disease, echinococcus, cystic, taenia

Highlights

- ✓ Renal hydatidosis is a very rare disease that can be associated with potentially severe complications, such as vascular compression, cyst infection, shock, sepsis and death.
- ✓ There are numerous options for the treatment, however the surgical approach (either open or laparoscopic) remains the most effective treatment of choice, which ensures the removal of the cyst.

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Introduction

Human hydatid disease (hydatidosis or echinococcosis) is caused by larval stages of cestodes (tapeworms) of the genus *Echinococcus*. There are multiple subtypes of *Echinococcus*, such as: *E. Granulosus* that causes cystic echinococcosis (the most common form encountered), *E. Multilocularis* responsible for alveolar echinococcosis, *E. Vogeli* that leads to polycystic echinococcosis and *E. Oligarthrus* which is an extremely rare cause of human echinococcosis.

Echinococcus Granulosus is generally found in Asia, Africa, Europe, Central and South America, the Middle East and, in rare cases, in North America. The disease is frequently encountered in people involved in sheep breeding, due to the sheep's role that acts as an intermediate host of the parasite, and also due the presence of dogs that are allowed to ingest the offal of the infected sheep.

The adult *Echinococcus Granulosus* worm, which measures 3 to 6 mm long, resides in the small intestine of the definitive hosts (dogs or other canids). Gravid proglottids release eggs that are then passed into the feces. These eggs are swallowed by suitable intermediate hosts, such as sheep, goat, swine, cattle, horses, camel, under natural conditions. After ingestion, the egg hatches in time in the small bowel where it releases an oncosphere which penetrates the small intestinal wall and migrates within the circulatory system towards several organs (the most commonly involved being the liver or the lungs). In these organs, the oncosphere develops into a cyst which expands gradually, leading to protoscolices and daughter cysts which fill the entire cyst interior. The definitive host becomes infected only after ingesting the cyst-containing organs of the infected intermediate host. The protoscolices often evaginate after ingestion, attaching the most appropriate intestinal mucosa and evolving towards adult stages within 32-80 days. Humans are accidentally intermediate "dead-end" hosts, who become infected by direct contact with infected dogs, ingesting contaminated raw or undercooked vegetables or by drinking contaminated water (1).

Because the liver is the first filter, it is the most commonly affected organ (75%). The oncospheres that are able to escape the hepatic filter will also involve the lungs in approximately 15% of the cases. The spleen, the kidneys, the pancreas, the bones and the brain are among the most rarely affected organs. The oncospheres that settle in the organs grow slowly and cause symptoms late in the evolution of the infection. The diagnosis can be made even 20–25 years after the initial infection (2, 3).

The renal hydatid cyst is an extremely rare entity (encountered in 2-3% of cases), usually involving the cortex of the kidney, most commonly being a solitary lesion (4, 5). It is not clear how the echinococcus oncosphere reaches the kidney in cases of primary hydatid disease, but it is postulated that it must pass through the portal system into the liver and retroperitoneal lymphatics (6).

Renal hydatidosis usually remains asymptomatic for many years, thus cysts can reach enormous size. Symptoms may vary according to the size and the extent of the cyst, but they are usually nonspecific and subtle. The renal hydatid cyst is considered closed if the pericyst, ectocyst and endocyst are intact. A hydatid cyst is considered exposed if it is no longer protected by the pericyst or by the lining of the collecting system. An open or communicating cyst has all three layers ruptured, resulting in free communication with the calyces and the pelvis. Cystic rupture into the collecting system, causing hydaturia, is only seen in 10-20% of all renal hydatidotic cases. Hydaturia is pathognomonic for this pathology and it is usually microscopic. Gross passage is rather uncommon, but it has a tremendous diagnostic utility. It is characterized by an episode of renal colic, followed by the passage of a small white grape-like structure in the urine (4, 7, 8). If the cyst is closed and does not communicate with the renal pelvis, patients may remain asymptomatic. Symptomatic patients can present with flank or abdominal pain, hematuria and hypertension. A palpable mass can be the only clinical sign. Acute urinary retention and anuria have also been reported (9, 10).

Discussions

Routine blood tests are generally normal, except for eosinophilia, which is found in only 50% of cases. Serological tests, such as immunoelectrophoresis, indirect hemagglutination, western blot, enzyme linked immunosorbent assay (ELISA) technique and complement fixation are useful in diagnosing primary renal hydatid cysts, but these investigations are not very specific and usually associate a high incidence of false negative results. El-Shazly AM et al have compared the use of indirect hemagglutination with the ELISA technique and showed that the sensitivity and specificity of ELISA were 96.7% and 97.5% respectively, while those of indirect hemagglutination were 86.7% and 95% respectively (11). Chemical tests, such as Casoni test, which can detect up to 90% of the cases, and Ghedine-Weinberg test, which can detect up to 80% of cases, are not very specific and outdated. Gharde et al have stated

that a combination of serological investigations yields a proper diagnosis in only 50 % of cases (12).

Imaging plays a key role in the diagnosis and staging of renal hydatid cysts. Ultrasonography (US) is an essential tool for diagnosing kidney hydatid cysts, indicating floating membranes and daughter cysts. It can also detect hydatid sand, which is demonstrated by changing the patient's position under real time ultrasound, resulting in the movement of the hydatid sand, thus giving rise to the “falling snowflake pattern.” The major disadvantage of US is its accuracy in diagnosing renal hydatid cysts which depends on the operator's experience. The WHO/IWG-E – 2010 classification system for the diagnosis and treatment of cystic echinococcosis is based on ultrasound imaging and classifies hydatid cysts as active, transitional and inactive (13, 14). Computed tomography (CT) has a better sensitivity and specificity in diagnosing renal hydatidosis. Hydatid cysts are seen as unilocular or multilocular cysts with defined walls, which are frequently calcified. The use of MRI offers no real advantage over CT scan. Plain films are usually unspecific and mostly unrevealing. They can show a soft-tissue mass in the renal area and if calcification is present, it is usually peripheral, curvilinear and eggshell-like (15).

Treatment options for renal hydatidosis include: medical treatment, PAIR (percutaneous aspiration, injection of chemicals and re-aspiration), open or laparoscopic surgery and, in selected cases, a conservative approach like “watch and wait” is the best. However, surgery remains the most effective treatment in order to remove the cyst, this approach leading to a complete cure. The cyst type, its location, size and complications are all taken into consideration when deciding what therapeutic method should be used. Patients with small cysts or multiple cysts in several organs can be treated successfully with benzimidazoles, such as Albendazole or Mebendazole. According to literature data, between 30% and 50% of patients can achieve a significant regression of the cyst size, as well as important symptoms' alleviation under chemotherapy. Both Albendazole 10 to 15 mg/kg body weight per day (max. 800 mg orally in two doses) and Mebendazole 40-50 mg/kg body weight per day continuously for several months (as a second treatment of choice) have been highly effective. Albendazole can be very effective when administered preoperatively, as well as in the postoperative period (for one month), for the intended purpose of facilitating the safe surgical manipulation of the cysts by inactivating it, altering the integrity of the cyst's membranes and decreasing the chance of anaphylaxis, as well as reducing

the turgidity of the cysts (1). Nephron-sparing surgery (partial nephrectomy or removal of the hydatid cyst with pericystectomy) is possible in most cases (75%), while nephrectomy must be reserved for damaged kidneys (which are encountered in 25% of cases). Extreme care must be taken during surgery to prevent cyst rupture that may lead to secondary disseminated hydatidosis. Throughout the nephron-sparing surgery, scolical agents such as hypertonic saline solution should be used before opening the cavities, to inactivate the daughter cysts and therefore to prevent further spread or anaphylactic reaction (16). Pedicled omentoplasty can be used to reconstruct large soft-tissue defects that arise in the kidney after cystectomy, this technique promising good results (17). Trans-peritoneal and retroperitoneal laparoscopic management can be used, both with good results (18, 19). PAIR (percutaneous aspiration, injection of chemicals and re-aspiration) under ultrasonography guidance has been shown to be effective and it is indicated for patients with relapse after surgery, failure of chemotherapy alone, or for patients who refuse to undergo surgery. The major drawback of this method is the high incidence of dissemination of daughter cysts and fatal anaphylaxis (1, 20).

The prevention and control of the disease are also very important. The CDC recommendations in terms of prevention of cystic echinococcosis are as follows: prevent dogs from feeding with the carcasses of infected sheep, control stray dog populations, restrict home slaughter of sheep and other livestock, do not consume any food or water that may have been contaminated by fecal matters from dogs, wash hands with soap and warm water after handling dogs and before handling food, teach children the importance of washing hands to prevent infection (1).

In our practice, we have encountered the case of a 74-year-old male, from a rural area, who presented for a left renal tumor, displaying cystic characteristics, diagnosed by CT scans. From the patient's medical history, we have found out that his symptoms had started two years earlier, when he began to have mild allergic-type manifestations (21-23). At that moment, the triggering factor was not identified. Considering the cystic characteristics of the tumor, the mild allergic-type symptoms and the fact that he was from a rural area, we have tested the patient serum for anti-hydatid cyst antibodies and the result was positive. The patient's eosinophils count was normal. After making the diagnosis, the patient began a one-month treatment with Albendazole, prior to surgery, for the inactivation of the cyst, as well as for decreasing the

chance of anaphylaxis and reducing the turgidity of the cysts. We have chosen an open retroperitoneal surgical approach, through a subcostal incision and the hydatid cyst was removed by pericystectomy. The retroperitoneal approach was preferable so that we could avoid possible contamination of the peritoneum, in the event of a cystic rupture. The histopathological report confirmed the diagnosis. After surgery, the patient received Albendazole for two more months.

Ameur et al have studied 34 cases of primary renal echinococcosis and found that the clinical features were dominated by flank or abdominal pain in 63% of cases, followed by hematuria (31%) and a palpable mass in 26% of patients. Hydaturia, although considered a pathognomonic sign, was present in less than 20% of patients (in a study, 11.4% of cases had this symptom) [24]. Ultrasonography was the preferable method of diagnosis and CT was performed whenever the diagnosis remained uncertain. In 23 cases, they have opted for the resection of the prominent dome, five patients were treated by pericystectomy, six by total nephrectomy and one by partial nephrectomy (24).

Benchekroun et al have reported a series of 45 cases of renal hydatid disease in which they have encountered similar results, flank pain being the dominant symptom, followed by a palpable mass and hematuria. The majority of the patients have undergone nephron-sparing surgery, such as cyst roof dissection, pericystectomy, partial nephrectomy, but due to the hydatid cyst complications, 18 patients needed to undergo total nephrectomy (25).

Even though the recommended treatment is the surgical excision, studies have demonstrated that the medical treatment may be an alternative, allowing the preservation of the organ. Soares et al have presented the case of a 14-year-old male, whose diagnosis was made by imaging studies and specific serological tests. His treatment consisted of 4-week cycles of Albendazole and resulted in a progressive reduction of the cyst as well as in a negative specific serology (26).

Hydaturia can be an unusual cause of renal colic. In their study, Unsal et al, have communicated the case of a 38-year-old female, with a 3-year history of episodes of renal colic and occasional removal of grape-like material. The diagnosis was made by ultrasonography and CT imaging. Due to the connection between the cyst and the renal collecting system, right nephrectomy was performed (7).

Conclusions

Renal hydatidosis is a very rare disease, that can be associated with potentially severe complications such as vascular compression, cyst infection, shock, sepsis and death. Patients are usually asymptomatic for long periods of time and if symptoms are present, they are frequently nonspecific and subtle (most commonly flank or abdominal pain or hematuria). Imaging investigations are essential in making the correct diagnosis, the most commonly used means being ultrasonography and CT scans. There are numerous options of treatment, however the surgical approach (either open or laparoscopic) remains the most effective treatment of choice, which ensures the removal of the cyst.

Conflict of interest disclosure

There are no known conflicts of interest in the publication of this article. The manuscript was read and approved by all authors.

Compliance with ethical standards

Any aspect of the work covered in this manuscript has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

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