Inflammation and infection

An unusual case of a huge hydatid cyst of ectopic pelvic kidney

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Introduction

Hydatid disease is endemic in parts of Africa, Australia, South America, Asia, Southern Europe, and the Middle East.1 The liver is the most commonly involved organ, followed by the lungs. Hydatid disease of the urinary tract is uncommon, accounting for only 2–3%, and may cause considerable diagnostic difficulty for the clinician. The pelvic kidney has a congenitally short ureter and an aberrant blood supply. We report here the case of a huge Hydatid cyst of an ectopic kidney. To our knowledge, this case is the first hydatid cyst of pelvic kidney reported to date.

Case

A 67-year-old woman, living in an urban area in west of Tunisia, with a history of hydatid cyst of liver presented to our hospital complaining of dull pain followed by occasional passage of small, pearly white grape like structure per urethra during urination for the last 4 months. Physical examination revealed guarding, tenderness and a mobile mass in the right quadrant of abdomen. ELISA test for echinococcus antibodies was positive. Ultrasonography revealed a multivesicular well defined cystic mass extending from the right retroperitoneum to the pelvis where it appears to be attached to the ectopic pelvic kidney. The liver, spleen and left kidney were normal. The computed tomography confirmed the large cyst measuring 15 × 12 cm arising from cortex of upper pole of right ectopic pelvic kidney replacing the renal tissue. The cyst was multivesicular and well-defined with a thin non enhanced wall containing microcalcifications (Fig. 1A). The cyst was close to pancreatic uncinate process, the third part of the duodenum and right colonic flexure. Inflammatory changes in the pericyst fat were noted (Fig. 1B and C). Excretory phase showed a delayed right excretion and calyceal distortion. The diagnosis of isolated hydatid cyst of ectopic pelvic kidney was made. The abdomen was opened by right Kocher's incision. There was a cystic mass measuring 15 cm which covered the entire kidney. Initially, the hypertonic saline (20% sodium chloride solution) was injected into the cyst and surrounding tissues were protected using gauze soaked in hypertonic saline. After cyst removal, the right kidney parenchyma was completely destroyed. To perform nephrectomy we did a ligature of triple blood supply (bifurcation, common iliac and hypogastric).

Gross pathologic examination showed a large medullar white cyst containing translucent fluid and daughter cysts of varying size. Histopathology study revealed an hydatid cyst with three layers: outermost pericyst is fibrous, middle ectocyst layer is laminated, hyaline and acellular (Fig. 2A) and the inner endocyst is the germinative layer which consists of daughter cysts and brood capsules with scolices. It's associated to granulomatous foreign body reaction (Fig. 2 B). The adjacent medullary renal parenchyma shows lesions of chronic pyelonephritis (Fig. 2 C). Patient was discharged on postoperative day 3. No signs of recurrence showed after 6 months of follow-up.

Discussion

Hydatid cyst disease, also known as hydatosis or Echinococcosis, is a zoonotic and parasitic disease caused by the larval stage of Echinococcus granulosus.1 This infection may involve liver, lung and other organs. Renal involvement is a rare condition. Isolated renal hydatid cyst disease may reside symptomless for a long time. Clinical
presentations include fever, malaise, flank pain, palpable mass, hematuria and hydatiduria. Although hydatiduria characterized by the presence in urine of gelatinous material is a pathognomonic sign, it has been seen only in 5%–25% of renal hydatid cyst disease. 

Serology is useful in the diagnosis of hydatid disease. However, the sensitivity of serological tests is affected by the site and condition of hydatid cysts in the body. 

Preoperative diagnosis of renal hydatid cyst is usually based on radiological investigations. Renal hydatid cysts usually present as a multivesicular large cyst containing daughter cysts. Some ultrasound findings are suggestive of hydatid cyst such as a thick bilayered wall, the presence of multiple daughter cysts with a heterogeneous echogenic fluid containing membranes and hydatid sand. CT scan show the accurate location of the renal cyst, confirms the type presumed by ultrasound and allows a complete preoperative investigation. Type 1 cysts are unilocular and can be confused with a simple renal cyst. Type 2 cysts contain multiseptated daughter cysts developing in an echogenic fluid. Type 3 cysts are considered as non evolving completely calcified masses. Ruptured or infected cysts are considered as type 4 cysts. In our case, the renal hydatid cyst was a type 2 with inflammation of the pericystic fat suspecting a cyst cracking or an infection with an ectopic kidney. The pelvic kidney has a congenitally short ureter and an aberrant blood supply. Blood supply to the ectopic renal tissue was demonstrated by autopsy, surgical and arteriographic studies in 33 patients. 17 kidneys had a solitary artery arising at or just distal to the bifurcation of the aorta, 12 kidneys had a double vascular supply (one artery from the bifurcation and one from the ipsilateral or contralateral common iliac artery), 3 kidneys had a triple arteries and 1 kidney had 4 vessels. 

Abstention is considered for non evolving completely calcified masses (type 3). In general, surgery is the treatment of choice for all other types of renal hydatid cyst and it should be based on the size of cyst, location, number, renal function and surgical methods. Both open and laparoscopic methods have been described in the treatment of renal hydatid cyst. 

![Fig. 1](image1.png) A: Contrast enhanced CT scan in the axial plane showing a multivesicular and well-defined cyst with a thin non enhanced wall containing microcalcifications. (B) and (C): Contrast enhanced CT scan in the sagittal and coronal plane showing the cyst close to pancreatic uncinate process, the third part of the duodenum and right colic flexure.

![Fig. 2](image2.png) A: Histological examination showing middle laminated hydatid membranes. (B): Histological examination showing granulomatous foreign body reaction and giant multinucleate giant cells surrounding laminary hydatid membrane. (C): Histological examination showing renal parenchyma with lesions of chronic pyelonephritis (HES X20).
literature. There is fear of cyst rupture and dissemination during laparoscopy. To avoid the spread of the disease by the infective daughter cysts, the cyst should be removed without rupture. Many scolicides are used: 10% formalin, 30% saline solution, 0.5% silver nitrate, 1% iodine and hydrogen peroxide. We use either 30% saline solution or hydrogen peroxide. Intraoperative use of hypertonic saline injected in the cyst can exterminate the infective daughter cysts. Extreme care should be taken to prevent leakage during the surgery. Kidney sparing hydatid cyst removal (cystectomy with pericystectomy), is possible in most cases (75%). Nephrectomy (25%) is considered only if the kidney is destroyed by the cyst.

Histopathologically a renal hydatid cyst shows a non nucleated laminated membrane composed of innumerable delicate laminations and a nucleated inner germinative membrane. High-power photomicrograph of the cyst depicts an *E. granulosus* scolex with shark tooth hooklets floating in proteinaceous cyst fluid.

**Conclusion**

In this case, we reported an unusual presentation of hydatid cyst disease in ectopic kidney. A limited number of isolated renal hydatid cysts are reported in the literature, while the disease is often misdiagnosed as a simple lumbar pain or a malignant renal mass. Careful diagnosis and complete pathophysiology of urinary tract hydatidosis are needed to be clarified.

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