

Case reports

Isolated cystic echinococcosis of kidney burlesquing as renal cell carcinoma: a diagnostic pitfall

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ABSTRACT. *Echinococcus granulosus* infection affects worldwide population, including countries of South and Central America, Middle East, sub-Saharan region, Russia and China. In cystic echinococcosis diagnosis is related to the presence of single or multiple well-delineated spherical primary cyst, commonly seen involving liver followed by lungs. Renal hydatid cyst is rare, comprising of about 2–3% of all locations, isolated renal involvement is even rarer. We present a case of hydatid disease in a 45 year male patient who presented with the history of vague flank pain, mild fever and hematuria. IVU showed a filling defect while USG and CT scan displayed multiloculated cyst in the upper pole of right kidney, correlating the clinical and radiological findings a differential diagnosis of cystic renal cell carcinoma and cystic nephroma was derived. Total nephrectomy was done but the histopathological examination of the lesion revealed hydatid cysts and final diagnosis of renal cystic echinococcosis was made.

Key words: hydatid disease, echinococcosis, renal cell carcinoma, cystic nephroma

Introduction

Renal cystic echinococcosis (hydatid disease) is caused by the larval stage of *Echinococcus granulosus* [1]. Isolated hydatid cyst involving the urinary tract is very infrequently seen. The cyst grows slowly in the kidney and can remain asymptomatic for several years, that can be as long as even up to 10 years [2]. Occasionally, these cyst may simulate an intra-abdominal tumour or even may be complicated with gross ascites [2].

Herein, we are reporting a case of 45 year male presenting with multiple small cysts in his right kidney, which was misdiagnosed as a renal malignancy on radiology. We therefore present this case with the aim of recapitulating the consideration of this differential diagnosis for clinicians and radiologists those dealing with the renal space occupying lesions.

Case presentation

A 45-year-old Indian male presented to the surgery outpatient department with the complaints



Fig. 1. CT scan of the abdomen showing thin walled multiloculated cyst in the upper pole of the right kidney measuring 5.5×5.2×5 cm in size

of vague flank pain, mild fever and hematuria. The patient was of average built and nutrition. Physical examination revealed a smooth non-tender mobile mass in his right hypogastrium. Systemic examination revealed hypertension only. IVU displayed the area as a filling defect, while ultrasonography and CT scan of the abdomen picked up thin walled multiloculated cyst in the upper pole of the right kidney measuring 5.5×5.2×5 cm in size (Fig. 1), rest of the urinary system was unremarkable. No cystic or solid lesions were noticed in other organs as liver, lungs or spleen. The left kidney and the ureter were normal and there was no ascites or lymphadenopathy. CT findings suggested a differential diagnosis of renal cell



Fig. 2. Multiple small cysts taken out of the larger cyst

carcinoma and cystic nephroma. Right sided nephrectomy was performed on account of suspected malignancy and the resected kidney was sent for histopathological examination. On gross examination of the specimen, cut section showed multiple small cysts (Fig. 2) encased in a large cyst located in the upper pole of the kidney (Fig. 3). Microscopic examination revealed a thin nucleated inner germinative membrane along with few scolices (Fig. 4), a non-nucleated laminated membrane composed of innumerable delicate laminations (Fig. 5) and an outermost thick fibrous membrane (fibrocyst). Cytospin smear from cyst fluid consisted of many hooklets. No features of malignancy was detected. Finally on the basis of histopathological examination diagnosis of isolated echinococcosis was made.

The postoperative period was uneventful, however the patient was prescribed albendazole 400 mg twice daily for 4 weeks to prevent metastatic cyst formation. The patient's follow-up with abdominopelvic CT scan and chest radiography was normal after a period of 6 months.

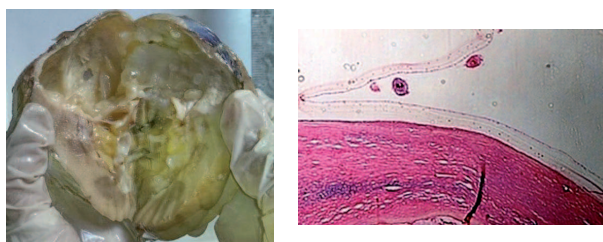


Fig. 3. Multiple small cysts encased in a large cyst located in the upper pole of the kidney

Fig. 4. Thin nucleated inner germinative membrane along with few scolices (H&E, $\times 100$)

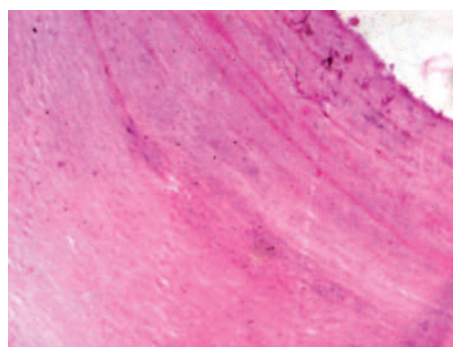


Fig. 5. Non-nucleated laminated membrane composed of innumerable delicate laminations (H&E, $\times 100$)

Discussion

Hydatid disease is a cyclo-zoonotic parasitic infection prevalent worldwide. Larval stage of the cestode *Echinococcus granulosus* is responsible for the disease in human, which includes predominantly two species: *Echinococcus granulosus*, the causative agent of cystic hydatid disease (or cystic echinococcosis, CE); and *Echinococcus multilocularis*, which causes alveolar echinococcosis, AE [3]. The recent studies of *Echinococcus* genetic diversity has led to taxonomic revisions and the genotypes G1-G3 are now grouped as *E. granulosus* sensu stricto, G4 as *E. equinus*, G5 as *E. ortleppi*, and G6-G10 as *E. canadensis*. Genotype G1 is cosmopolitan and the major causative agent of human cystic echinococcosis but not the only one. Liver is the most common site of echinococcal infection (54% to 77%) [4], because the liver acts as the initial filter for the organisms playing an important defensive mechanism. Those larvae that escape filtration by the liver subsequently gets filtered by the lungs (9% to 30%), spleen (0.9% to 8%), kidney (2% to 3%), and brain (1%) are the other organs involved however to a much lesser extent [4,5]. When the genitourinary tract is affected, the site is incorrigibly renal, but prostatic, bladder, paravesical, ureteral, epididymal, and testicular involvement have also been proclaimed.

Echinococcus, commonly called as flat worm, is a member of the Taeniidae family. It is approximately 5 mm long and the adult inhabits the bowel lumen of the definitive host. The adult tapeworm residing in the intestine of the dog, produces larvae that cause echinococcosis. The infection occurs when humans are exposed to contaminated faeces. The adult worm abides in the proximal jejunum of the definitive host by attaching itself to the intestinal mucosa using hooklets. The

eggs are excreted in the host's feces and when it is ingested by an appropriate intermediate host, such as sheep, cattle, or humans, the embryos hatch out from the eggs, penetrate through the intestinal mucosa and enter into the systemic circulation and can reach the portal venous or the lymphatic system. The patients with renal echinococcosis mostly present with vague pain in the lumbar region [7]. Less commonly they may present with a palpable mass in the loin and rarely some present with a history of passing whitish material in urine also known as "grape skin" [7]. Two variants of the disease occur, classic hydatid disease caused by *Echinococcus granulosus* and the rarer aggressive, multilocular variant caused by *Echinococcus multilocularis* [8], which infiltrates the organ inhabited by it and hence poses difficulty in its elimination. Germinal layers of *Echinococcus granulosus* responsible for the generation of brood capsules and protoscoleces into a central cavity filled with clear "hydatid" fluid. It is surrounded first by a acellular laminated layer, then by the host response, a peripheral zone of fibroblasts. "daughter" vesicles of variable size may present inside or outside the "mother" cyst [9]. The growing cyst causes a mass effect upon the affected organ. Radiologic imaging techniques help in the diagnosis of renal hydatid disease, but they may be inconclusive at times, and the diagnosis is possible at the time of the cyst removal. Ultrasonography US is particularly useful in detection of the cystic membranes, septa, and the hydatid sand while CT demonstrates the cyst wall calcification and the cyst infection. There is no specific diagnostic test of renal hydatid disease. In 20% to 50% of cases, moderate eosinophilia is present [6]. The Casoni and Weinberg tests have been annihilated in many centers due to their limited efficacy [10]. Although serologic and hemagglutination tests have low reliability, still their positivity confirms the disease. Immunoelectrophoresis against arch-5 is however a highly specific test [11]. Surgery is usually the best treatment for renal hydatid disease, and if possible kidney-sparing procedure is more rational. In cases where preoperative diagnosis of a hydatid cyst is made, the area around the cysts can be carefully isolated by gauze packing and initial cyst aspiration and infiltration of scolicide agent can be done. Partial cysto-pericystectomy is another available option where the cyst, including the hydatid membrane and the daughter cysts are opened, the laminated membrane is removed and scolicide-

soaked swabs are kept into the cavity, the margin of the remnant pericyst tissue is then sutured by running absorbable sutures. Total nephrectomy is recommended only in the cases of non-functioning kidneys. Chemotherapy with antihelminthics of the benzimidazole family, shows a poor compliance due to low accessibility of the cyst to the drug but the outcome can be better if it is used in conjunction with the surgery which helps in preventing recurrences.

Conclusions

1. Preoperative diagnosis of renal hydatid disease is difficult even in an endemic zone.
2. Imaging studies are suggestive but usually inconclusive, and the differential diagnosis with a renal tumor or complicated cyst may not be made without surgery.
3. Complicated cysts in renal echinococcosis can mimic renal malignancies or a benign ureteropelvic junction obstruction.
4. Although hydatid cysts of the kidney are relatively rare, this disease must be considered in people with renal cystic masses from endemic countries.
5. Veterinary interventions to prevent and control the extent and intensity of infection in definitive host populations is a must in endemic areas, by controlling the prevalence in animal intermediate hosts also.

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