

# INCIDENTALLY DETECTED PRIMARY GIANT RENAL CYSTIC ECHINOCOCCOSIS IN A YOUNG PATIENT: AN UNDERESTIMATED ENTITY?

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**Disclosure:** No potential conflict of interest.

**Received:** 13.10.14 **Accepted:** 24.11.14

**Citation:** EMJ Urol. 2015;3[1]:42-45.

## ABSTRACT

Echinococcosis is a parasitic infestation caused by *Echinococcus granulosus* and is an endemic disease in many parts of world. The symptoms and signs depend on the location and size of the cyst. Renal cystic echinococcosis or hydatid cyst (HC) disease of the kidney is extremely rare and constitutes only 2-4% of all cases of hydatid disease (HD). We present a 39-year-old male patient who was referred to our outpatient clinic with cystic right kidney mass that was incidentally diagnosed during hepatobiliary ultrasound for chronic hepatitis B evaluation. Routine blood tests were normal without eosinophilia. Indirect haemagglutination test was negative. Abdominal kidneys, ureters, and bladder X-ray showed an 83x95 mm sized curvilinear calcification in the right upper abdominal quadrant. Abdominal computed tomography scan and magnetic resonance imaging demonstrated a 10x9x10 cm sized cystic mass arising from the middle pole of the right kidney, destructing the whole upper pole and extending into the liver. Daughter vesicles were present in the cystic lesion suggesting renal HD. Right retroperitoneal exploration with flank approach and right radical nephrectomy was performed without any complications. Pathology confirmed HC lesion. Following surgery, albendazole 400 mg per os twice daily for 4 weeks was suggested.

**Keywords:** Renal cystic echinococcosis, renal hydatid cyst, kidney, diagnosis, management.

## INTRODUCTION

Echinococcosis is a parasitic infestation caused by *Echinococcus granulosus*. Infestation occurs as a result of ingestion of the larval form of a cestode and humans are intermediate hosts. Cestode larvae reach the liver with portal system, while infection of the kidneys occurs with systemic circulation. Echinococcosis is an endemic disease in many parts of world such as South America, Mediterranean countries, New Zealand, Australia, and South Africa.<sup>1</sup> Keeping in mind that dogs are main hosts and sheep are intermediate hosts; a high index of suspicion for echinococcosis for patients coming from rural areas is mandatory. Laboratory workers

who are handling hydatid disease (HD) specimens are also in need of following biosafety level 2 practices for preventing accidental contacts.<sup>2</sup>

Renal cystic echinococcosis (RCE) or hydatid cyst (HC) disease of the kidney is extremely rare and constitutes only 2-4% of all cases of HD.<sup>3</sup> There are no specific clinical symptoms or signs but hydatiduria as a pathognomonic symptom was reported in literature.<sup>4</sup> Routine blood tests are generally within normal limits except for eosinophilia which is observed in 20% of patients.<sup>5</sup> Diagnosis is usually incidental or the consequence of high suspicion index. Radiologic evaluation is the mainstay of diagnostic modalities for HD. In endemic countries, echinococcosis should be included in

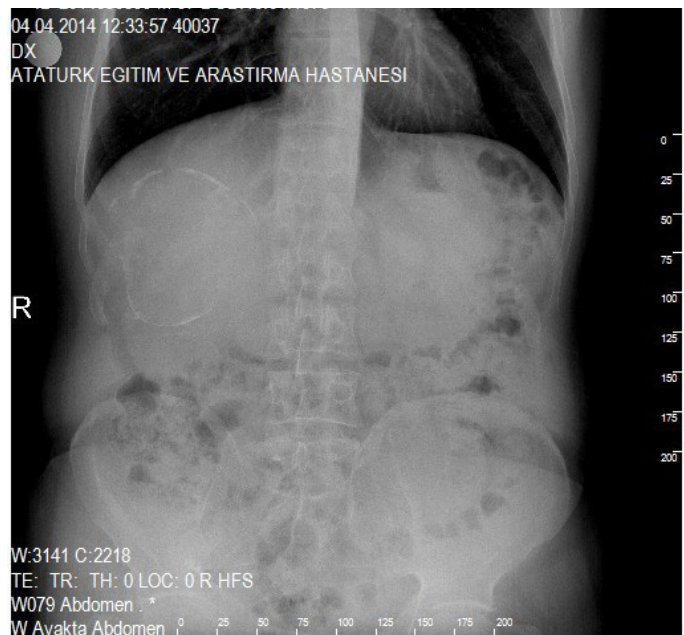
the differential diagnosis of lesions in solid organs, especially for liver and kidneys. Herein, we present a young male patient with isolated giant right renal HC disease that was diagnosed incidentally, and discuss its management.

## CASE REPORT

A 39-year old male patient was referred to our outpatient clinic with cystic right kidney mass that was incidentally diagnosed during hepatobiliary ultrasound (US) examination for chronic hepatitis B (CHB) evaluation. He had renal cyst history 20 years ago, of which we had no exact radiological report, but verbal history from the patient suggest a small benign cyst. The patient had CHB infection since his childhood but he was excluded from medical follow-up due to his poor compliance. He is a temporary employee in the construction industry. He had no known history of contact with dogs or sheep and has been living in central Turkey.

An abdominal kidneys-ureters-bladder X-ray showed an 83x95 mm sized curvilinear calcification observed in the right upper abdominal quadrant (Figure 1). Abdominal computerised tomography (CT) scan and magnetic resonance imaging (MRI) demonstrated a 10x9x10 cm sized cystic mass arising from the middle pole of the right kidney, destructing the whole upper pole and extending into the liver (Figures 2a and 2b). Daughter vesicles were present in the cyst mass lesion, suggesting renal HD. Chest X-ray and blood tests including complete blood count, blood biochemistry, urine culture, and microscopy were within normal limits. Blood tests for echinococcosis including indirect haemagglutination were negative. Physical examination revealed no additional abnormal findings but a palpable right renal mass.

Right retroperitoneal exploration with flank approach and right radical nephrectomy was performed. Estimated blood loss was 80 cc. Histopathology confirmed hydatid cyst of the right kidney (Figures 3a and 3b). A mass in kidney with 10 cm in diameter, surrounded by vicious capsule that included cystic lesion was reported that involved the superior and middle parts of the kidney. The inferior pole of kidney was normal. Postoperative follow-up was uneventful and the patient was discharged on postoperative Day-3. Albendazole 400 mg per os twice daily for 4 weeks was prescribed. No perioperative (0-30 days) complication was detected.

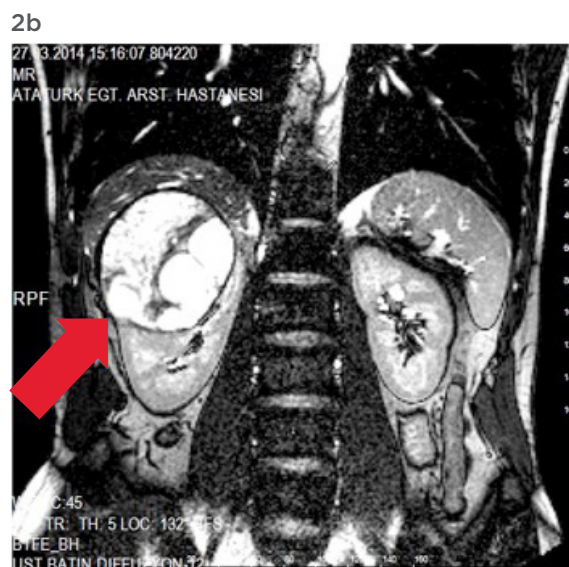


**Figure 1: Abdominal X-ray imaging showing a curvilinear calcification on the right upper abdominal quadrant.**

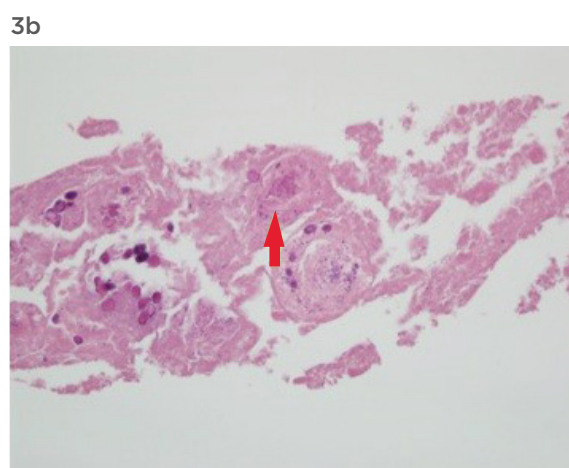
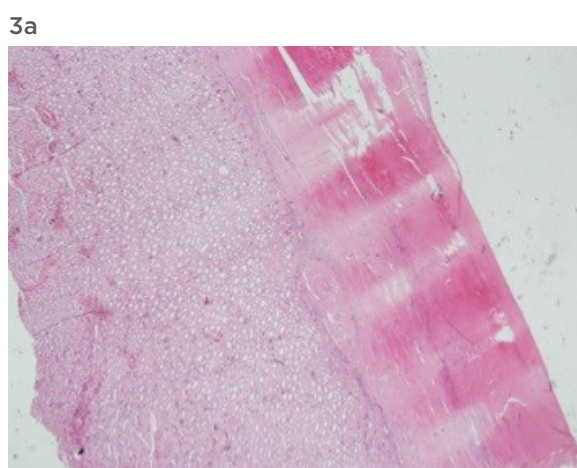
## DISCUSSION

Renal echinococcosis accounts for approximately 3% of all HDs and is the third most common localisation of HD.<sup>1,6</sup> The liver and lungs have been reported to be most frequently affected by HD.<sup>1</sup> Renal echinococcosis may remain asymptomatic for a long period of time.<sup>7</sup> Abdominal pain and hydaturia, which occurs due to a ruptured cyst in the collecting system and leads to passing collapsed daughter cyst-like material in the urine, have been reported as the most frequently reported clinical findings.<sup>8</sup> Fever, lumbar mass, and haematuria might also be present.<sup>8</sup> Renal failure and ureteropelvic junction obstruction due to renal HD were also reported in the literature.<sup>9</sup>

The pathognomonic sign, consisting of hydaturia, should indicate the rupture of cyst and the diffusion of the content in the excretory tracts. While microscopic hydaturia could be seen in 10-20% of patients, macroscopic hydaturia is a rare symptom.<sup>7</sup> Radiological investigations allow clarification of the diagnosis and provide the most interesting evidences for hydatidosis diagnosis. The X-ray allows the display of a thin arc-shaped calcification that is characteristic of HC compared to heterogeneous and more or less diffused calcifications.<sup>10</sup> However, the plain abdominal X-ray could also be normal.<sup>4</sup>



**Figure 2: Abdominal computed tomography scan (2a) and magnetic resonance imaging scan (2b) showing a 10x9x10 cm sized cystic mass arising from the middle pole of the right kidney, destructing the whole upper pole and extending into the liver (arrows).**



**Figure 3: a) Hydatid cyst (HC) involving kidney tubules with cyst wall (haematoxylin and eosin x20); b) HC of the kidney - appearance of the hooklet forms, in the form of birefringent (arrow) (haematoxylin and eosin x200).**

In our patient, curvilinear calcification was present in the right upper abdominal quadrant of the X-ray that suggested renal cyst HD.

The diagnosis of HC using US is reliable and it has been reported to be specific in up to 80%<sup>6</sup> of cases, and sensitivity is reported in up to 95%.<sup>11</sup> This technique provides an accurate size of the cyst, its topography, structure, and diagnoses any associated abdominal lesions. Advanced radiologic techniques, such as CT, play an important role in the diagnosis. CT scan shows a spectrum of findings from unilocular cysts, which may have thick

calcified walls, to a multiloculated cystic mass with heterogeneous density, and daughter cysts.<sup>4</sup> In our patient, abdominal CT scan and MRI demonstrated a 10x9x10 cm sized cystic mass arising from the middle pole of the right kidney, destructing the whole upper pole and extending into the liver.

There is no specific laboratory finding for RCE. Eosinophilia may be present.<sup>10</sup> Casoni and Weinberg tests have 'historical' importance in cyst hydatid.<sup>1</sup> Counter immunoelectrophoresis against arch-5 antigen test was suggested as highly specific for HD.<sup>7</sup> However, in our patient the serological results

were all negative. In the epidemiological context, the radiological and biological data allow the final diagnosis. Huang and Zheng<sup>11</sup> have reported a retrospective analysis of 19 renal HD patients and the preoperative diagnostic accuracy for US, CT, and serology were 66.7%, 88.2%, and 92.3%, respectively.<sup>11</sup> Other cystic masses of the kidney, such as cystic renal cell carcinoma, nephroblastoma, and abscess should also be considered in the differential diagnosis. Particularly history of the patient and the area of his/her living should alert the clinician about the possibility of the presence of renal HD.

Surgery is the main treatment for the RCE. Medical management with albendazole shows limited effect with potential side-effects.<sup>1</sup> We administered albendazole, which was suggested following a consultation with the Infectious Diseases Department due to the very large size of the cystic renal lesion. Nephron sparing surgery (NSS) or partial nephrectomy should be applied whenever possible and has been reported to be possible in 75% of cases.<sup>12</sup> Laparoscopic cyst excision with irrigation-aspiration was also reported.<sup>13,14</sup> In our

case, we performed open retroperitoneal approach due to the large size of the renal cyst that involved most of the kidney. We did not consider transperitoneal laparoscopic approach in order to avoid possible cyst rupture; due to its large size it may have led to intra-abdominal dissemination that could lead to serious complications. However, minimal invasive NSS was also reported that depended on the size and location of the renal hydatid mass lesion.<sup>15</sup> Following surgical resection, excellent outcomes have been reported with complete or partial surgical resection, with most patients having no recurrences.<sup>6,10</sup>

## CONCLUSION

In conclusion, renal HD is a rare but significant aetiology of renal cystic lesions and it can mimic renal masses. Disease course is usually silent, common presentation is asymptomatic and a high suspicion index is mandatory for clinical diagnoses. The definitive diagnosis is only possible after histopathological examination. Surgery is the mainstay of treatment in contemporary practice and NSS is feasible in the majority of the cases.

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