Isolated renal hydatid disease in a non-endemic country: a single centre experience

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Abstract

Objective: Isolated renal hydatid disease (HD) is rare in non-endemic countries. Clinical and radiological suspicion warrants appropriate serological tests, preoperative treatment and intra-operative precautions. We present a tertiary care centre experience of isolated renal HD in a non-endemic country.

Methods: We reviewed the medical records of patients with HD treated in the past 20 years. We identified patients with the definitive diagnosis of isolated renal HD and described their management.

Results: Of the 119 cases with HD, 6 were found to have isolated renal involvement (5%). Their median age was 46.5 (28-70) years. Five patients presented with flank pain and 1 had an incidentally discovered renal mass. Radiologic investigations raised the suspicion of possible HD in 4 cases, while 2 cases were diagnosed as renal tumours. Computerized tomography showed complex renal cyst in 4, solid renal mass with heterogonous enhancement in 2 and calcification in 5**.** Eosinophilia and indirect hemagglutination test (IHA) were positive in 3 of the 4 suspected cases. Three cases were treated as renal tumours, while 3 were managed as HD. Four cases had total nephrectomy and 2 had partial nephrectomy. Histopathology revealed that all cases had renal HD. Patients were followed for a median of 7.3 (0.4-11.3) years with no evidence of recurrence.

Conclusions: Isolated renal HD is a challenging preoperative diagnosis in non-endemic countries. The definitive diagnosis is only possible by histopathology. Retrospectively, HD mimicked renal tumours in half the cases and should be considered in the differential diagnosis of renal space occupying lesions.

Résumé

Objectif : L'hydatidose rénale isolée est une maladie rare dans les pays où elle n'est pas endémique. Des éléments cliniques et radiologiques suspects justifient des tests sérologiques appropriés, un traitement préopératoire et des précautions peropératoires. Nous présentons l'expérience d'un centre de soins tertiaires avec l'hydatidose rénale isolée dans un pays où cette maladie n'est pas endémique.

Méthodologie : Nous avons passé en revue les dossiers médicaux de patients atteints d'hydatidose au cours des 20 dernières années. Nous avons cerné des patients ayant reçu un diagnostic final d'hydatidose rénale isolée et nous décrivons leur traitement.

Résultats : Sur les 119 cas d'hydatidose, 6 avaient une atteinte rénale isolée (5 %). L'âge médian de ces patients était de 46,5 ans (28 à 70). Cinq patients présentaient des douleurs au flanc, et un patient présentait une masse rénale découverte fortuitement. Des examens radiologiques ont fait hausser les soupçons quant à une hydatidose dans quatre cas, alors que dans les deux autres cas, on a diagnostiqué une tumeur rénale. La TDM a montré un kyste rénal complexe chez quatre patients, une masse rénale solide avec densification hétérogène dans deux cas et des calcifications dans cinq cas. L'éosinophilie et le test d'hémagglutination passive étaient positifs dans trois des quatre cas soupçonnés. Trois cas ont été traités comme des tumeurs rénales, alors que les trois autres ont été traités comme une hydatidose. Quatre patients ont subi une néphrectomie totale et deux, une néphrectomie partielle. L'histopathologie a révélé une hydatidose rénale dans tous ces cas. Les patients ont été suivis sur une période médiane de 7,3 ans (0,4 à 11,3) sans aucune donnée montrant une récurrence.

Conclusions : L'hidatidose rénale isolée est un diagnostic préopératoire difficile à poser dans les pays où elle n'est pas endémique. Le diagnostic final n'est possible que par histopathologie. De façon rétrospective, on note que l'hydatidose ressemblait à une tumeur rénale dans la moitié des cas et devrait être prise en compte dans le diagnostic différentiel de lésions occupant l'espace rénal.

Introduction

Cystic echinococcosis (hydatidosis) is a parasitic disease caused by the larval form of *Echinococcusgranulosus*. It is a common disease in specific regions. It is endemic in most sheep-raising countries in Asia, Europe, South America, New Zealand and Australia where sheep, dogs and humans live in close contact.[1,2] The guts of dogs and other carnivorous animals represent a definitive host for the adult worm. Humans become an accidental intermediate host by ingesting Echinococcus eggs. The liver is the most commonly involved organ, followed by the lungs. Isolated renal involvement is very rare affecting up to 1.9% of patients.[3] No report in the English literature has been made from a

Table 1. Patients' characteristics; clinical, laboratory and radiological findings, treatment and pathological features

			Laboratory									
Case	**Gender**	**Symptoms**	**Esinophilia**	**IHT**	**Radiological**	**Procedure done**	**Gross features**	**Laminated membrane**	**Germinal layer**	**Scolices**	**Hooklets**	**Calcification**
1	Female	Right flank pain	Positive	Positive	Aus- 6 cm x 6 cm x 5 cm mass lower pole right kidney with internal echoes .CT - 6 cm x 6 cm x 6 cm solid mass lower pole right kidney	Right partial nephrectomy	Right kidney, cystic, 9 cm, calcification	YES	YES	NO	YES	YES
2	Male	Accidentally discovered	Positive	Positive	CT- Left upper pole cyst with calcification	Left partial nephrectomy	Left kidney (upper pole), cystic, 7 cm	YES	NO	YES	NO	YES
3	Female	Right flank pain	Negative	Not done	Aus - 6 cm x 5 cm well defined mass with mixed echogenecity IVP - Curvilinear calcification, distortion of right pelvicalcial system, CT-upper pole mass right kidney with mixed heterogenous enchancement	Right nephrectomy	Right kidney (upper pole), cystic, 6 cm	YES	NO	NO	NO	YES
4	Male	Left flank pain	Negative	Negative	CT - Complex cystic mass lower pole left kidney with peripheral calcification and enhancing solid components	Left nephrectomy	Left kidney (upper pole), cystic, 8 cm	YES	NO	NO	NO	YES
5	Male	Right flank pain	Negative	Not done	CT - There is 5.2 x 4.9 x 4.7 cm thin rim enhancing low density structure with internal heterogenecity arising from the right mid renal pole extending up to the renal pelvis. No enhancing intramural nodule or septa. It splaying the pelvicalyceal system	Right nephrectomy	Right kidney (lower pole), cystic, 4.2 cm	YES	YES	NO	NO	NO
6	Female	Left flank pain	Positive	Positive	CT - left kidney is replaced by large well defined multiseptated 13x11x8 cm cystic mass with mild enhancement of its with calcification in the upper part	Left nephrectomy	Left kidney, cystic, 18 cm, calcification	YES	YES	YES	YES	YES

non-endemic country of isolated renal hydatid disease (HD). Due to its infrequent occurrence, a preoperative diagnosis is often not considered. However, a preoperative accurate diagnosis is important to provide appropriate antihelmentic treatment, take intraoperative precautions preventing parasite dissemination and possibly avoid unnecessary nephrectomy. The combination of clinical history, laboratory and imaging studies offer a reliable pre-treatment diagnosis in only 50% of cases and a presumptive diagnosis in 71%.[4] We report 6 cases of isolated renal HD managed in a single centre in a non-endemic country in the past 20 years. We aim at raising the awareness of the urologists in non-endemic

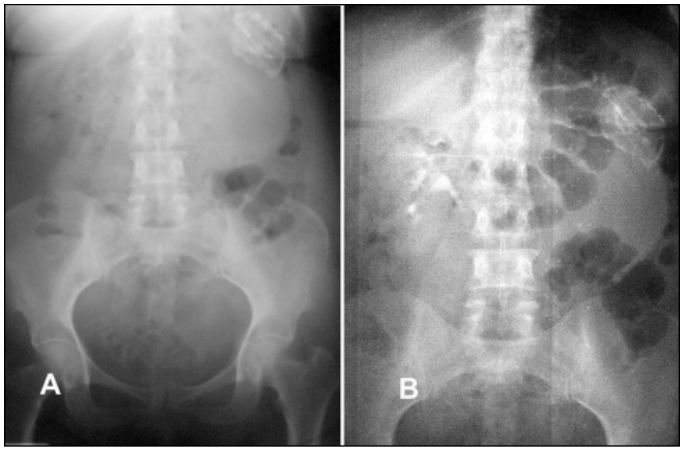

Fig. 1. A. Plain x-ray reveals curvilinear calcifications; B. Excretory urography showed non excretion of contrast by the left kidney.

countries to consider isolated renal HD in the differential diagnosis of space-occupying lesions of the kidney.

Materials and methods

We searched the surgical pathology database for the diagnosis of HD. The medical records of these patients were reviewed to identify patients with isolated renal involvement. We collected data on residence, clinical presentation, laboratory investigations, radiological studies, treatment and follow-up. Data included urinalysis, blood count, indirect haemagglutination test (IHA), abdominal ultrasonography (AUS), intravenous urography (IVU) and computerized tomography (CT). Pathology reports and available glass slides of isolated renal lesions were re-examined by the pathologist.

Results

In the 20-year period from 1989-2009, 119 cases with the pathological diagnosis of HD or hydatid cyst were found. Out of these, 10 cases had renal involvement, of which only 7 cases were isolated. One patient was excluded because he was referred from an endemic country. We report on the remaining 6 patients with isolated renal HD.

The median patient age was 46.5 years (range 28-70). All patients had a lifetime residency within the Kingdom of Saudi Arabia (KSA), which is classified as a non-endemic country for HD. Half of the patients were born and lived in the Central Region where the hospital is located; the other 3 patients were from the West, North, and East areas of the KSA. Their history of travelling to endemic areas could not be known from the charts. Half of the 6 patients were male. Five cases presented with flank pain (83%), while the 6th case was incidentally discovered. Only one side was involved, with the right or left kidneys being affected

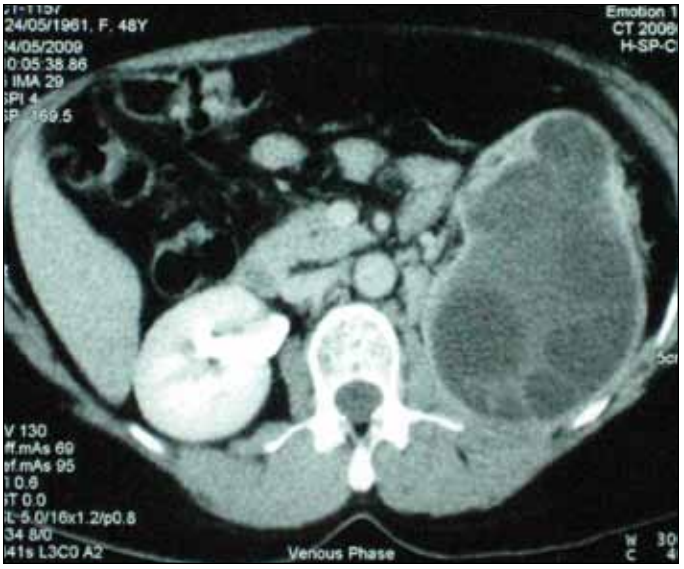

Fig. 2. A computed tomography scan showing a large hydatid cyst of the left kidney.

equally. The entire kidney was involved in 1 case, upper pole in 2, and lower pole in 3 (Table 1).

A plain abdominal x-ray revealed a ring-shaped or curvilinear calcification in 3 cases (50%) (Fig. 1, part A). An IVU was done in 3 patients and demonstrated space-occupying lesions and caliceal distortion in 2 patients and a non-functioning kidney in 1 patient (Fig. 1, part B). A CT was performed in all cases and showed complex cystic mass with calcifications and peripheral enhancement after contrast injection in 4 cases (67%). In 2 cases (33%), a CT demonstrated soft tissue mass with calcifications and heterogonous enhancement after contrast injection mimicking a renal tumour (Fig. 2). Based on imaging, a presumptive diagnosis of renal tumour was made in 2 cases. The remaining 4 cases were suspicious for having HD. Eosinophilia and IHA test were positive in 3 of these 4 suspected cases. The fourth case showed no evidence of eosinophilia and IHA test was not done. In summary, 2 cases were treated as renal tumours, 1 case was considered a renal tumor with suspicion of being HD, and 3 cases were considered isolated renal HD. Preoperative antihelminthicalbendazole treatment and preparation for intraoperative hypertonic saline and surgical field isolation were carried out in the 4 HD suspected cases. Four patients underwent nephrectomy, while partial nephrectomy was performed in 2 patients. All patients continued postoperative albendazole or started the treatment upon histopathological diagnosis.

All tissues were fixed in 10% neutral buffered formalin and subjected to standard tissue processing, after which the tissues were embedded in paraffin, cut at 3- to 5-micrometre sections and stained with the hematoxyline and eosin stain. Pathological diagnosis was made by identifying the finely laminated membrane in all cases (Fig. 3, part C). Germinal

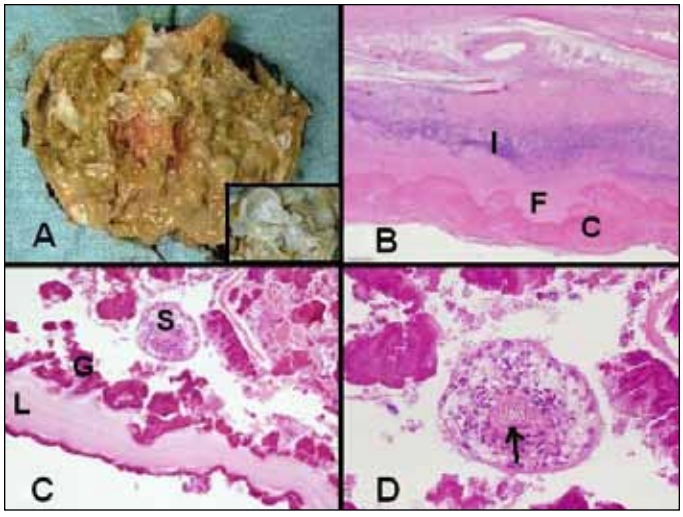

Fig. 3. A. Opened renal hydatid cyst (case 6) showing multiloculation. A close up view of some daughter cysts is depicted in the inset. B. Low magnification microscopic view of a calcified cyst where host inflammatory reaction (I), fibrosis (F) and calcification (C) are evident (hematoxylin and eosin [H&E] original magnification 40×). C. The characteristic laminated membrane (L) and germinal layer (G) are illustrated; a scolex (S) is noted in the center of the field. (H&E original magnification 400×). D. The scolex is seen in high magnification including the hooklets (arrow). (H&E. original magnification 400×).

layer, scolices and hooklets were variably present (Table 1) (Fig. 3, part C and D). There was also variably prominent host reaction surrounding the lesion, including foreign body giant cell reaction, lymphoplasmacytic response, fibrosis and calcification (Fig. 3, part B).

Patients were followed up for a median of 7.3 (0.4-11.3) years. All patients had no evidence of recurrence during the follow-up period, as shown by postoperative serological and radiological studies.

Discussion

Hydatid disease is a parasitic infestation of sheep and dogs caused by the tapeworm *E. granulosus*. Humans can be accidental intermediate hosts. It is endemic in sheep- and cattle-rearing parts of the world.[4] The eggs in the dog's feces contaminate grass, farmland and water, which are ingested by sheep (the usual intermediate host). Humans can also be the intermediate host. The larvae hatch, penetrate venules in the wall of the duodenum and are carried by the portal circulation to the liver, which becomes infected most frequently (70%). The larvae that enter the systemic circulation may lodge in the lung (25%), kidney (2-4%) or other organs.[4,5]

In endemic countries, isolated HD of the kidney is a rare (2%) and challenging condition to diagnose.[3] The kidneys are the most commonly affected urinary organs, but bladder, prostate, seminal vesicles and testis can also be involved.[6-9] The exact incidence of HD in non-endemic regions, like the KSA in which this study was done, is unknown. Over the 20-year period, we identified 119 HD cases in different body sites. Out of these, 6 had isolated renal HD (5%), which is higher than previously reported.[3] This relatively high number might be explained by the fact that our hospital is a tertiary centre, to which cases are referred from different regions.

The clinical picture of our cases is not specific to suggest the diagnosis of urinary tract HD and is similar to that of the published series from endemic areas. Hydatid disease is usually encountered between the ages of 30 and 50 years, and is rarely seen in children.[5] Our patients' ages ranged from 28 to 70 years (median 46.5 ± 13.7). Patients may present with a flank mass (84%), pain (73%), hydatiduria (5%-29%), and rarely acute retention of urine or anuria.[4,10-14]

In our series, 5 cases presented with flank pain (83%) and 1 case was incidentally discovered. Hydatiduria is a pathognomonic sign due to rupture of the cyst into the collecting system. We had no case presenting with hydaturia.

No laboratory test is specific to HD.[6] Eosinophilia is reported in 25% to 50% of HD cases and may occur in other parasitic diseases.[15,16] In our series, esinophilia was reported in 3 out of 4 suspected cases (75%). Many immunodiagnostic tests were reported to have overall sensitivity against the hydatid crude antigen. Negative tests do not exclude HD and positive results neither confirm the diagnosis nor correlate with the pathological stage of renal HD.[13,15] Positive tests may suggest further workup for HD. The IHA test is positive in about 75% ofcases,[4,6] which is similar to our results. False-positive reactions are present in areas where *E. granulosus* and *E. multilocularis* coexist, and in areas where other parasitic diseases are endemic. Negative results do not rule out the diagnosis due to circulating immune complexes.[6] Serology may be beneficial after surgery to exclude recurrence.

Radiologic findings of HD, although not specific, may provide a clue towards a correct preoperative diagnosis. The plain x-ray may show a soft-tissue mass or a ring-shaped calcification in the renal region. Excretory urography may demonstrate caliceal distortion, caliectasis and a non-functioning kidney, possibly caused by the mass effect of cystic lesions.[17] Although abdominal sonography is helpful in the diagnosis, a CT is more accurate and sensitive.[6] The sonographic findings may show anechoic lesions with well-defined margins, while a CT may demonstrate a cyst with a thick or calcified wall, a unilocular cyst with a detached membrane, a multiloculated cyst with mixed internal density, and daughter cysts with lower density than the maternal matrix.[18-20] In 4 (76%) of our cases, a CT was helpful to suggest HD in the differential diagnosis. The other 2 cases were diagnosed as follows: complex renal cyst in 1 case and a solid renal mass managed as a renal tumour in the other case. Maintaining a high index of suspicion and being familiar with the characteristic clues in imaging may provide a correct preoperative diagnosis and, consequently, appropriate management.

Our series shows that, in a non-endemic country, careful observation provided suspicion of isolated renal HD in two-thirds of patients. Nevertheless, half the patients went into surgery with the primary diagnosis of renal tumours. A correct preoperative diagnosis protects against dissemination of the parasite and avoids unnecessary radical surgery.

Treatment of renal hydatid cysts is essentially surgical. Kidney-sparing surgery is performed whenever possible.[21-23] The surgeon must be careful to totally remove the cyst and avoid spilling its contents. Spilling the cyst contents may cause severe anaphylactic reaction and dissemination of the disease. If a cystectomy is not feasible, partial or total nephrectomy is required. A preoperative diagnosis is important to take the necessary intraoperative precautions, such as delicate manipulations, use of abdominal pads soaked with hypertonic saline to reduce the risk of dissemination during surgery and to prevent recurrence.[24] Four of our patients underwent nephrectomy (2 patients in view of massive parenchymal replacement and the other 2 with suspicion of malignancy). Partial nephrectomy was feasible in 2 patients. Four patients received perioperative antihelminthic chemotherapy (albendazole), as HD was suspected preoperatively; the other 2 received the medication once the diagnosis was histologically confirmed. Preoperative treatment with albendazole renders the cyst nonantigenic and decreases the tension in the cyst wall. As a result, the risk of spillage and anaphylaxis are reduced. None of our patients experienced intraoperative or postoperative complications; they were completely cured with no evidence of disease recurrence.

The technique of percutaneous aspiration injection and reaspiration has been described as a safe and effective treatment modality for renal HD.[25,26] However, other reports have shown that none of the cysts disappeared completely using this technique; the technique could be reliable for risky patients with symptomatic hydatid cysts.[27] We believe that this technique carries the risk of dissemination and anaphylactic reactions. Further studies in more patients would elucidate the usefulness of this technique in renal HD.

Conclusions

Although isolated renal HD is a rare entity in our region, it must be considered in the differential diagnosis of renal lesions. Clinical, serologic and radiological findings can suggest, but not confirm, the diagnosis. Isolated renal HD is a challenging preoperative diagnosis in non-endemic countries. The definitive diagnosis is only possible by histopathology. Retrospectively, HD mimicked renal tumours in half the cases and should be considered in the differential diagnosis of renal space-occupying lesions. Surgical excision of renal HD is successful in providing a long-lasting cure.

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