Isolated renal hydatid disease (heterogeneous presentations)

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(Ann. Coll. Med. Mosul 2008; 34(1): 21-27).
Received: 24th Sept 2007; Accepted: 20th Feb 2008.

ABSTRACT

Objective: To demonstrate that isolated renal hydatid cysts have heterogeneous presentations.
Methods: a retrospective case series study during the period from Jan 2002 to Jan 2007 in the urosurgical department at Al-Jamhory teaching hospital. Five patients with different preoperative diagnoses were dealt with. After their initial clinical assessment and investigating their pathology by lab and imaging studies including IVU and MRI, provisional diagnoses were put and they were explored through lumbar approach and the pathology dealt with accordingly.
Results: there were three females and two males with six isolated renal hydatid cysts; mean age 28.8 years. Five cysts involving the left kidney. Their initial diagnoses were pyonephrosis, renal tumor, paracolic abscess, uremia due to large bilateral renal cysts and lower moiety hydronephrosis in a duplicated system. The preoperative diagnosis of renal hydatid was certain in two patients (40%) after cyst aspiration and hydatidiuria; the other three cases were diagnosed intraoperatively, and all cases proved by histopathology. Passing daughter cysts with the stool in a left side renal pathology may help in the diagnosis of a complicated renal hydatid cyst.
Conclusion: isolated renal hydatid is a rare disease, has heterogeneous presentations, and preoperative diagnosis may not exceeds 50%, so it needs to be kept in mind in the differential diagnosis of renal space occupying lesion.

الخلاصة:

اهداف البحث: اكياس العدارية لها تمثيل سريري متغاير.
التصميم: دراسة سلسلة من الحالات.
موقع الدراسة: دراسة مستعادة في قسم البولية بمستشفى الجمهوري التعليمي خلال الفترة من كانون الثاني ٢٠٠٢ الى كانون الثاني ٢٠٠٧.
المشاركون: خمسة مرضى مصابون بأعراض امراض مختلفة في الكلى تم التعامل معها، وبعد الفحوص المختبرية والتصويرية المتمثلة باشعة الكلية المدعمة بالصبغة الوريدية والمغنام وتثبيت التشخيص الاولي تم استكشاف الكلية عن طريق المنطقة القطنية للتعامل مع الحالة.
النتائج: خمسة مرضى (٣ اناث و٢ ذكور) وستة اكياس عدارية تم التعامل معها (معدل العمر ٢٨.٨ سنة). خمسة اكياس في الكلية اليسرى وواحدة في اليمنى. التشخيص الاولي للحالات كان تقيح الكلية، ورم الكلية، الفشل الكلوي بسبب ضغط الاكياس، استسقاء الجزء السفلي للكلية وخراج حول القولون. تم تشخيص الاكياس العدارية قبل اجراء العملية لمريضين (٤٠%) بعد بزل احد الاكياس والاخر بعد بيلة عدارية، وتم تشخيص بقية المرضى اثناء العملية. تم دعم التشخيص

بالفحص النسيجي للمرضى. ظهور الاكياس العدارية مع الغائط بوجود كتلة معقدة في الكلية اليسرى قد يساعد على تشخيص الاكياس العدارية في الكلية.
الاستنتاجات: ندرة حدوث اكياس العدارية في الكلية بصورة منفردة مع تغاير تمثيلها السريري، كما ان التشخيص النهائي قد لا يتعدى ٥٠% من الحالات قبل اجراء العملية الجراحية . الاكياس العدارية في الكلى يجب ان تضمن كأحد التشخيصات التفريقية الاولية للكتل المرضية في الكلى.

Renal Echinococcosis is a rare acquired disease of the kidney. The larval stages of Echinococcus granulosus, are responsible for cystic echinococcosis or hydatid disease. The kidneys are involved in less than 2 % of all human hydatidosis; isolated renal involvement is even rarer[1,2,3]. Hydatid cysts enlarge slowly and require 5-20 years before they are diagnosed. Developed cysts range from 1-15 cm in diameter, but may be larger. Unless it becomes infected giving the picture of renal abscess, renal hydatid cyst may be asymptomatic until it causes a noticeable mass effect; rarely hematuria or hypertension may be the presentation[1]. Signs and symptoms will vary according to cyst location, size, type and number. Rupture or leaking cysts can cause severe anaphylactoid reactions and may release protoscolices that can produce secondary echinococcosis. Cysts are typically spherical, thick-walled and unilocular, most frequently found in the liver (more than 70% in the right lobe) and lungs, but they may occur in other organs including the kidneys [4,5].

Unlike liver hydatid, the diagnosis of renal hydatid cyst is not easy (the only pathognomonic sign of renal hydatid is hydatidiuria (5-25%)[3,6,7], and the findings at imaging studies are frequently suggestive of hydatid disease but usually not conclusive, and differentiation from renal tumor or a complicated renal cyst may not be made without surgery. The combination of clinical history, imaging studies, serological and urine investigations yields reliable pretreatment diagnosis in only 50% [8].

In this paper we present five cases of isolated renal hydatid cyst with different or heterogeneous presentations.

Patients and methods:

A retrospective study of five patients with six isolated renal hydatid cyst were managed at Mosul teaching hospital, urology department during the period from Jan 2002 to Jan 2007. All cases were referred from medical or surgical departments for urosurgical advice and intervension. History, clinical examination and imaging studies including ultrasound, intravenous urography, and MRI were done for all patients in addition to urine examination, renal function tests and hematological tests (complete blood picture and ESR).

Pre operative renal cyst aspiration was done under local anesthesia and ultrasound guidance in one patient. All patients underwent renal exploration through lumbar approach and the pathology dealt with either by hydatid cystectomy or nephrectomy and the

diagnosis was confirmed histopathologically. All patients were put on albendazole postoperatively.

Results:

Five patients and six renal hydatid cysts were dealt with. There were three females and two males, their age range from 11 – 40 (mean age 28.8 year). All patients were from rural areas. Five cysts were found in the left kidney and one in the right. All cases presented with complications of the cyst. The presentations or preoperative diagnosis were renal failure due to large bilateral renal cysts, fig (1&2), pyonephrosis, renal tumor, abdominal mass due to left lower moiety, hydronephrosis, fig (3). Two cases were diagnosed preoperatively, one after cyst aspiration in order to alleviate the pressure created by the cysts. The aspirate revealed crystal clear fluid (contains hydatid sand on microscopical examination), and the aspiration was followed by hypotension and urticaria (1st patient) and the other after hydatidiuria (4th patient, fig 4&5). The other three cases were diagnosed intra operatively. One of the cases gave past history of similar attack of loin mass, fever and diarrhea then the patient passed different sizes cystic structures with the feces followed by disappearance of the loin mass and resolution of the condition. Eosinophilia was seen in one patient only. In three cysts (50%) there were no daughter cysts inside them, fig (6). The summery of the cases is presented in table 1.

Table 1: Summery of the cases and their presentation and treatment.

Name	*Sex*	*Age*	*Cyst site*	*Pre operative diagnosis*	*Diagnostic finding*	*Treatment*
1	**F**	**35**	**Bilateral**	**Renal failure**	**Crystal clear aspirate**	**Bilateral cystectomy**
2	**F**	**28**	**Left**	**Diarrhea & loin mass**	**Daughter cyst passage through the rectum**	**Cystectomy. (Fig 7)**
3	**M**	**40**	**Left**	**Renal tumor**	**Post nephrectomy mass bisection**	**Nephrectomy**
4	**M**	**30**	**Left**	**Pyonephrosis**	**Hydatidiuria**	**Nephrectomy**
5	**F**	**11**	**Left**	**Abdominal mass. (lower moiety hydronephrosis)**	**Intra operative cyst aspiration**	**Nephrectomy**

Table2: summarizes the differences between simple and hydatid renal cysts.

Character	**Simple renal cyst**	**Hydatid renal cyst**
Wall thickness	**Thin**	**Thick, may be thin**
Ultrasound	**Echo-free**	**Echo-free or daughters echos**
Plain X ray	**No calcification**	**Partial or complete calcification**
Aspirate	**Amber (hydatid sand -ve)**	**Crystal clear (hydatid sand +ve)**
Pericystic adhesions	**No adhesions**	**Dense adhesions**

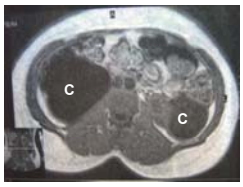

Fig 1:MRI,large bilateral hydatid cysts (C).

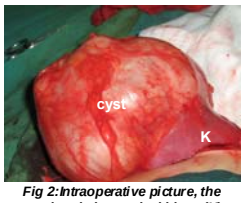

Fig 2:Intraoperative picture, the cyst in relation to the kidney (K).

Fig 5: hydatid daughter cysts

Fig 6: renal hydatid cyst without daughter cysts

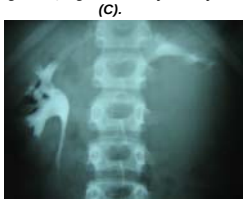

Fig 3: IVU, left soft tissue shadow.

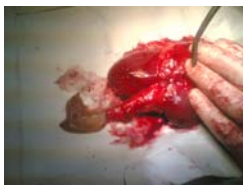

Fig 4:Daughter cysts filling the collecting system (clinically hydatidiuria).

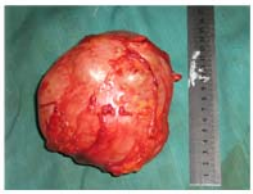

Fig 7: hydatid cystectomy

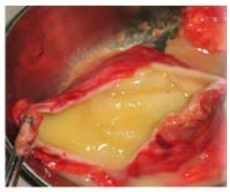

Fig 8: infected hydatid

Discussion

Isolated renal hydatid cysts are rarely encountered, we report only five patients in five years period, nearly the same incidence as it was reported by Gugus et al (20 patients in 25 years) and Yilmaz et al (7 renal cysts out of 372 hydatid cysts treated in 10 years) [7, 9].

Single or multiple small simple renal cysts are most common benign renal lesions, representing more than 70% of all asymptomatic renal masses. Occasionally the cyst may reach a very large size causing sensation of heaviness in the loin or the mass may be palpable where aspiration under ultrasound guide and injection of seclrosing agent (95% alcohol) may result in cyst cavity ablation in more than 90% of cases[10]. In the areas where hydatid disease is an endemic disease, renal hydatid cyst is suspected when there is personal or family history of hydatid disease or when there is ultrasound, or imaging evidence of daughter cysts. As isolated renal hydatid cyst is rare, and to have bilateral isolated renal hydatid is very rare, a thin walled hydatid cyst is difficult to differentiate from simple renal cyst specially in the absence of calcifications[3] or daughter cyst, as daughter cysts were found in three cysts out of six similar to a report by Makki[11]. Aspiration of the crystal clear fluid (containing hydatid sand) followed by hypotension and urticaria was the first clue for diagnosis in one of our cases. Table 2 summarize the differences between simple and hydatid renal cysts.

According to the site of hydatid cyst, daughter cysts could escape through the site of rupture giving rise to a variety of manifestations, biliary colic, ureteric colic or sudden death as it open to biliary system, ureter or a major vein respectively[12]. The history of passing small size cystic structures (hydatidiuria) with the urine associated with severe ureteric colic and dysuria is the only known pathognomonic sign of renal hydatid[3,6,7]. It was seen in one of our patients. The passage of variable size cystic structures with fecal material as a symptom of renal hydatid has not been reported. One of our case had this

history, the left renal hydatid cyst ruptured to the descending colon and evacuated its contents (associated with disappearance of the left loin mass) keeping the endocyst intact to repeat the cycle of cyst growth and present again after a variable duration. When the left renal hydatid cyst opens to the descending colon, the daughter cysts will be discarded with the feces without digestion.

Eosinophilia is of limited help in the diagnosis of renal hydatid, because less than 50% of patients will have elevated eosinophil count in their blood[3,6,7,13]; (20% in our series). Although Casoni's, complement fixation and indirect hemagglutination tests are helpful in some patients with renal hydatid cysts, their positive results may not exceed 25-50%, 40% and 75% respectively with high incidence of false positive and false negative results[3,7]. On the other hand a recent counter immunoelectrophoresis against arc-5 gained wide acceptance with higher specificity and sensitivity rates[7,10].

The T2- weighted MR imaging usually reveals a high-signal- intensity mass consisting of multiple thin-walled lesions and outlined by a thick, hypointense rim. The high intensity is due to the characteristic high fluid content of the mass. The signal intensity of the small cysts is similar to that of the mass. T1-weighted MR imaging demonstrates a thick wall that is nearly isointense relative to the septa and central part of the mass. The small peripheral cysts are hypointense relative to the central component and they are surrounded by a zone of tissue that demonstrates enhancement after the administration of gadolinium-based contrast material, whereas the mass itself did not enhance[14]. Lesions with a solid appearance may be encountered. Lack of internal contrast enhancement allows one to classify them as hyper attenuating or hyperintense cystic lesions and misinterpreting them as tumors[15,16,17]. Angulo et al reported in his series that renal tumor was the initial diagnosis in 56% of his 34 cases of renal hydatid[8].

The diagnosis of renal hydatid preoperatively was 40% in our study, Angulo et al, reported that the diagnosis of isolated renal hydatid disease may not exceed 50%, while Zargar et al couldn't diagnose any one out of 11 cases of renal hydatid cysts preoperatively[18]. other preoperative differential diagnoses include complicated renal cyst, cystic renal cell carcinoma, calcified renal cell carcinoma, hypovascular renal tumor, cystic nephroma, lobar nephronia and hemorrhagic pyelonephritis [8,17,18,19]. Renal hydatid should be born in mind in the differential diagnosis of space occupying lesion of the kidney[2,13].

At surgical exploration, dense pericystic adhesions are usually encountered in all cases due to the cytotoxic effects of the vesicular fluid causing exuberant granulomatous response by the host immune system; there is no such reaction in simple renal cysts. This reaction has two main consequences: fibrosis and necrosis. Complete calcification of the wall of a hydatid cyst can be considered an

indication of quiescence or perhaps death of the parasite[20]; these radiological and intraoperative findings help in the diagnosis of renal hydatid.

Conclusion

As isolated renal hydatid cyst is very rare, it causes difficulty in the differentiation from the more common simple renal cyst specially in the absence of daughter cysts, and from other renal pathologies in complicated cysts. The pre-operative diagnosis of isolated renal hydatid usually doesn't exceed 50%. The exact diagnosis is mainly surgical and should be kept in mind in the differential diagnosis of renal space occupying lesion in the endemic area.

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