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ISOLATED RENAL HYDATID CYST PRESENTING AS CYSTIC RENAL CELL CARCINOMA

Urology

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ABSTRACT

Introduction and objectives: Hydatid cyst/cystic echinococcosis is a parasitic infection caused by the larval stages of echinococcus granulosus .Definitive hosts are wild and domestic canine.Humans are aberrant intermediate hosts .2-4 % of such cases have Renal involvement. Isolated Kidney involvement is extremely rare (1.9%) and can mimic cystic RCC clinically and radiologically. Herein we discuss how to differentiate between Hydatid cyst and RCC and the management thereafter. **Methods:** Patient based in Tamil Nadu was admitted and worked up using various imaging tools such as USG, CT Scan and finally nephrectomy was done for histopathological analysis of the sample to confirm the diagnosis. **Results:** Based on past evidences and in the present patient, nephrectomy has to be carried to confirm and differentiate between the differential diagnosis of Cystic RCC and Isolated Renal Hydatid Cyst. Histopathological analysis of the specimen confirmed the same **Conclusion:** In spite of characteristic picture of cystic Echinococcosis on imaging isolated renal hydatid cyst is a very rare lesion and hence maybe misdiagnosed as a renal tumor. It should be considered a renal tumor until proven otherwise. Nephron sparing surgery is the primary treatment for isolated Renal Hydatid Cyst but Nephrectomy is warranted preoperatively misdiagnosed large cystic lesions and high grade lesions and for lesions involving collecting system.

KEYWORDS

INTRODUCTION

Hydatid cyst or Cystic Echinococcosis is a parasitic infection caused by the larval stages of Echinococcus granulosis. Cystic Echinococcosis is found in Africa, Europe, Asia, the Middle East, Central and South America, and in rare cases, North America(2). It has been found in all sheep raring countries, including India, with highest prevalence reported in Andhra Pradesh and Tamil Nadu(3).

The definitive hosts are wild and domestic canins. Humans are aberrant intermediate hosts, and become infected by ingesting eggs – most common mode of transmission is by consumption of soil, water, or food contaminated by fecal matter of infected dogs/sheep(2).

70% of the patients with Cystic Echinococcosis have liver involvement, while 25% have lung involvement, and 2-4% have kidney involvement(4).

Isolated kidney involvement is extremely rare – 1.9%(5), and it can mimic Cystic RCC clinically and radiologically(1). Clinical symptoms vary depending on the cyst's size, location and extension. Clinically, both Cystic RCC and Hydatid cyst can be asymptomatic for a long time, since their growth takes years. Once symptomatic, they can present with flank pain, hematuria, and abdominal mass.

Clinical Presentation

A 37 year old male, from Tamil Nadu, presented to the OPD with Bilateral loin pain, Left more than Right, of 4 years' duration and lower back ache, with no history of hematuria or mass per abdomen or significant weight loss.

Physical examination was unremarkable, with stable vital signs, average body build. Abdominal examination revealed no mass.

Renal Ultrasound revealed bulky left kidney with a defined heteroechoic lesion measuring 6.7x6.8x7.5cm, involving the lower pole with multiple cystic areas within, with peripheral vascularity [Fig 1a, 1b].

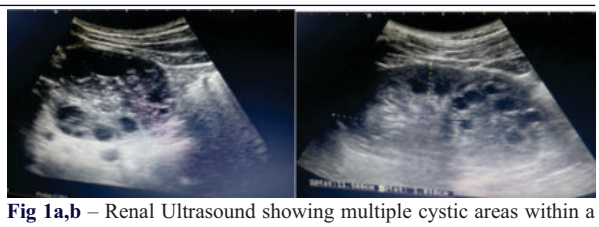
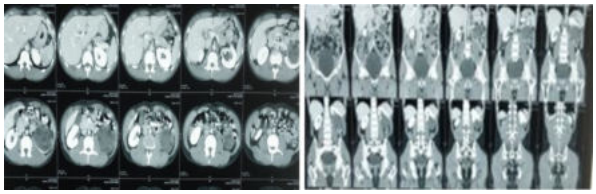


Fig 1a,b – Renal Ultrasound showing multiple cystic areas within a well defined lesion in the lower pole.

Contrast Computed Tomography was performed before presentation of the patient to our center and described a cystic lesion measuring 7.3x6.8cm, partially exophytic, in the lower pole in the left kidney, with multiple internal daughter cysts [Fig 2a,2b, 2c, 2d]. On CT imaging there is no metastatic lesion.



CECT Abdomen and Pelvis showing a cystic lesion in the lower pole of left kidney, with multiple internal daughter cysts

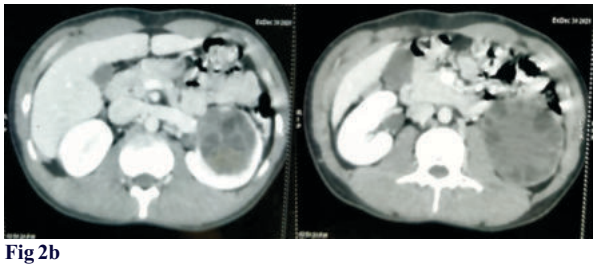


Fig 2b

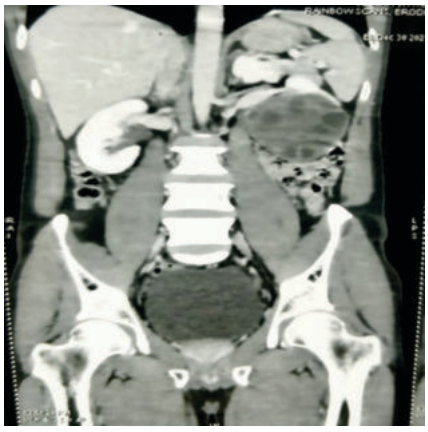


Fig 2c

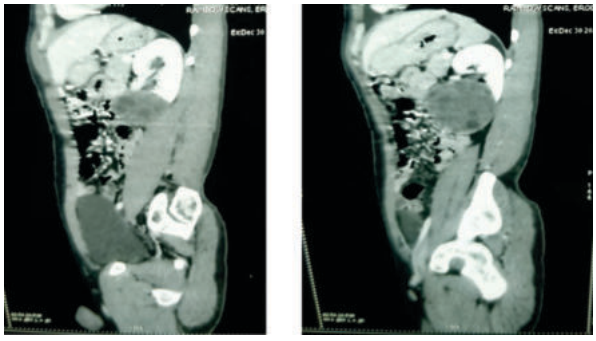


Fig 2d Fig 2b,c,d – Transverse, coronal and saggital sections of the Left kidney on CECT showing a cystic lesion in the lower pole of the left kidney with multiple cystic areas within.

A urine analysis and culture, renal function tests and routine tests were unremarkable, with raised ESR. Owing to the suspicion of malignancy, open left radical nephrectomy was performed through a Chevron incision. Left kidney tumor mass measuring 8x7cm was present involving the inter polar and lower pole found. Specimen was retrieved intact, without rupture and sent for Histopathological examination. The operation was completed in 3 hours, with minimal blood loss. The patient's Serum Creatinine was raised in the post-operative period (POD1) but returned to normal on POD3. The post-operative period was otherwise uneventful.

The external surface of the mass was cystic. On sectioning, the renal parenchyma was thinned out at the lower pole. Corticomedullary differentiation was lost. The pelvis and lower half of kidney was replaced by multiple translucent cystic structures largest measuring 2.5x2cm [Fig 3a, 3b]. Although the cyst was quite large, it was found that the interpole and the upper pole parenchyma of the kidney was preserved. Histopathological examination described and confirmed the diagnosis of renal hydatid cyst, and excluded malignancy. Clear gross and histopathological pictures reduced the necessity of post-operative serological testing for Echinococcus. A course of Tablet Albendazole 400mg twice daily, was prescribed for 4 weeks to reduce the possibility of relapse.

Fig 3a. Cut surface of the left kidney showing multiple daughter cysts within. The mid and upper pole parenchyma is preserved.
Fig 3b. Daughter cysts

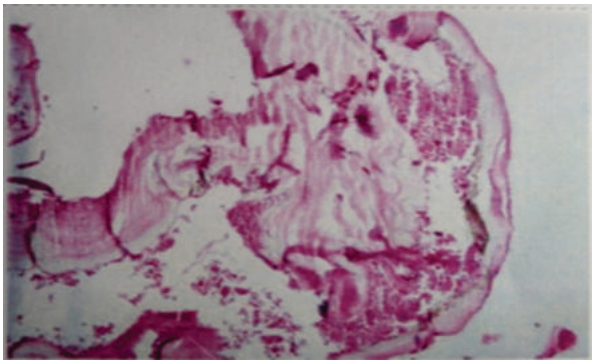


Fig 4. Histopathological examination of the renal hydatid cyst composed of outer adventitial layer made of fibrocollagenous tissue admixed with lymphoplasmacytic infiltrate and an inner laminated membrane. Germinative layer and protoscoleces are not seen.

DISCUSSION

Cystic Echinococcosis is a worldwide parasitic infection, seen mostly in sheep-raring countries, including India, with highest prevalence reported in Andhra Pradesh and Tamil Nadu(3). The definitive hosts are wild and domestic canids. Humans are aberrant intermediate hosts, and become infected by ingesting eggs in contaminated soils or food(2). The current case did not have a clear history of contact with animals. Thus, infection may have occurred via contaminated food.

Although it has no specific common clinical presentation, renal hydatid cyst has a suggestive characteristic appearance on imaging, surgical cut surface, and histopathological examination. The most common complaint is flank pain, due to cyst compression. Although hydatiduriais pathognomic finding of renal Cystic echinococcosis, it is extremely rare(4). The combination of a relevant clinical history, imaging tools, and laboratory investigations, provides a reliable preoperative diagnosis in only half of all cases.

On retrograde revision of the imaging studies in the current case, a well demarcated renal cyst with multiple cystic areas within, was observed. The multiple cystic areas were the daughter cysts within the Hydatid cyst. Cystic Echinococcosis lesions have been classified by the World Health Organisation into 5 grades. Grade 1 and 2 represent the early stage of disease where the production of scolices is active. Grade 3 is a transitional stage that represents the start of degeneration of the cyst with detachment of the germinal membrane from the cyst wall. It has been differentiated into Grade 3a for a solitary cyst with multiple septa and Grade 3b for multiple small cysts in solid parenchymal tissues. Grades 4 and 5 refer to old and degenerative stages of the disease with calcification. Based on this classification, imaging findings of the current case refer to a Grade 3b hydatidcyst(6). In the current case, this finding was confirmed by the cut surface of the mass after nephrectomy. A typical radiological picture of a renal hydatid cyst has been previously described as a single unilocular lesion with multiple daughter cysts or vesicles, like in our current case. Atypically, daughter cysts may be absent(7). An isolated renal hydatid cyst should be treated by nephron sparing surgery. However, nephrectomy is recommended for non-functioning kidneys, large cysts thought to be connected with the collecting system, and cysts with suspicion of tumor(4). The surgical aim is to remove the cyst intact, without rupture, to prevent anaphylactic shock and subsequent death. The current case warranted nephrectomy due to clinical and radiological suspicion of renal tumor pre-operatively, and the specimen was retrieved intact.

CONCLUSION

In spite of the characteristic picture of Cystic Echinococcosison imaging, an isolated renal hydatid cyst is a very rare lesion, hence may be misdiagnosed as a renal tumor. Therefore, this should be taken into consideration in the differential diagnosis of single or multiple renal cystic lesions. It should be considered a renal tumor until proven otherwise, even in countries and regions endemic for Echinococcosis(8). Nephron sparing surgery is the primary treatment for isolated renal HC, but Nephrectomy is warranted for pre-operatively misdiagnosed large cystic lesions and high grade lesions, and for lesions involving the collecting system(4).

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