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Case Report

Hydatid cyst of the kidney: a case report and review of the literature

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ABSTRACT

Primary involvement of the kidney is rare in case of hydatid disease. We present a case of primary left renal hydatid cyst in a 40 year old female patient admitted with left lumbar pain radiating to the back. Computed tomography of the patient was done and was suggestive of hydatid cyst involving the lower pole of the left kidney. The cyst was managed by left Nephrectomy with open surgical excision and preoperative course of Albendazole. This case emphasizes on better detection and evaluation of such rare cases to identify better treatment strategies.

Keywords: Cystic, Echinococcosis, Hydatidoses, Hydatid, Renal

INTRODUCTION

Hydatid disease is mainly caused by *Echinococcus Granulosus*.[1] It is a zoonotic disease. Liver is the most common site of involvement. Renal involvement is seen in 2% to 3% of patients, with isolated involvement of the kidney being even rarer.[2,3]

It often manifests as a slow growing cystic lesion. Patient may be asymptomatic or present with symptoms of lumbar region pain, haematuria and hydatiduria.

Computed tomography findings in renal hydatid typically include: a cyst with thick or calcified wall, unilocular cyst with detached membrane, a Multiloculated cyst with mixed internal density and daughter cysts with lower density than maternal matrix.

Rarely type IV hydatid cysts may mimic hypo vascular renal cell carcinoma. We present a rare case of primary left renal hydatid cyst with presenting feature of left lumbar pain.

CASE REPORT

A 40 year old female presented with left lumbar pain since 1 month. There was no dysuria, vomiting or fever. The patient had no history of cardiac disease and the pain was not associated with cough, wheezing, shortness of breath or hemoptysis. Routine laboratory investigations: complete haemogram show mild neutrophilia and Urine Routine examination show 18-20 pus cells/HPF.

X- ray KUB

Show enlarged left renal shadow with increased radio-opacity (35 mm) in pelvis.

Ultrasonography (USG)

Revealed a bulky left kidney, it is mal-rotated and shows multiple varying sized cysts in parenchyma. Dilated pelvis and upper ureter seen. Liver is normal in size with normal echo-pattern. Gall bladder, Pancreas, Spleen and right kidney are normal in size and echo-pattern.

CT scan abdomen- pelvis (Figure 1): Left kidney appears bulky and thinning of left parenchyma in the lower part is seen. Approximately 98 x 98 mm sized peripherally enhancing oval shaped lesion with multiple internal small non-enhancing cystic areas is seen arising from lower part of left kidney and extending into the adjacent prerenal and pararenal space and communicating with the left lower calyx through the defect in the left renal parenchyma.

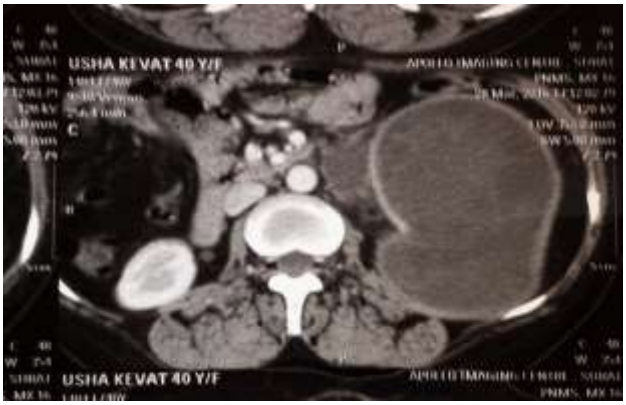

Figure 1: CT scan abdomen – pelvis: hydatid cyst involving the left kidney.

Gross hydronephrosis is seen in the left kidney with markedly dilated left ureter up to the vesico-ureteric junction with left side ureterocele. Non-enhancing areas are also seen in the left renal pelvis and upper ureter may represent extension of content of above described renal lesion. Left renal lesion is compressing the lower part of spleen, causing compressive superior, anterior and medial displacement of left kidney, distal body and tail of pancreas and peripheral displacement of adjacent bowel loops.

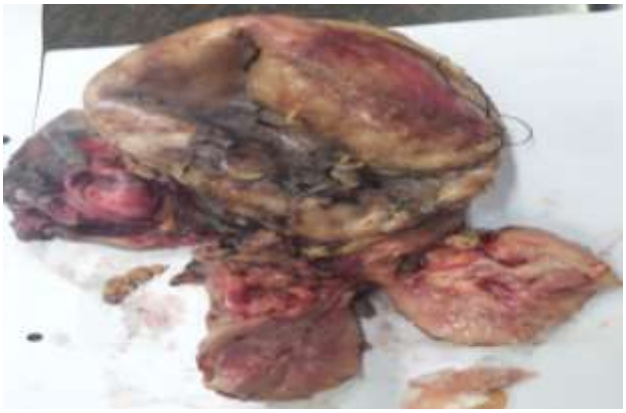

Figure 2: Left nephrectomy specimen: outer surface.

This may represent Hydatid Cyst of Left Kidney. There is delayed excreting left kidney. Right kidney is normal in size with homogenous cortical density. No calculus or hydronephrosis seen on right side. On administration of IV contrast, right kidney shows normal cortical enhancement and prompt excretion. The right ureter is not dilated and no evidence of ureteric calculus on either side. Bladder is normal.

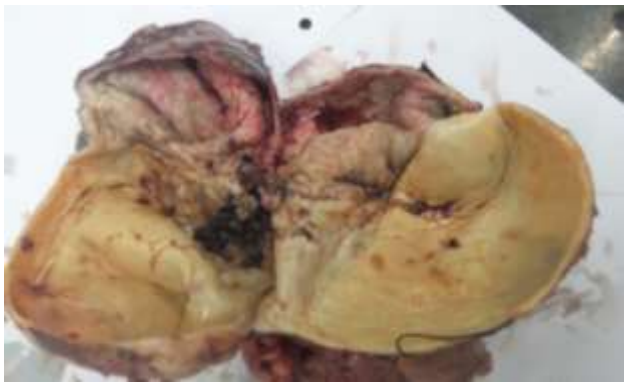

Figure 3: Left nephrectomy specimen: cut surface (with already drained cyst).

Surgery was done under general anesthesia via a left lumbar incision. The left kidney was identified after reflecting the peritoneum upwards. A cystic lesion was seen at the lower pole of left kidney. Nephrectomy was done. Incision was closed in layers. Patient was given a perioperative course of Albendazole with a view to sterilize the cyst preoperatively and to decrease the risk of recurrence of the cyst post operatively. Whole specimen is sent to the laboratory (In formalin).

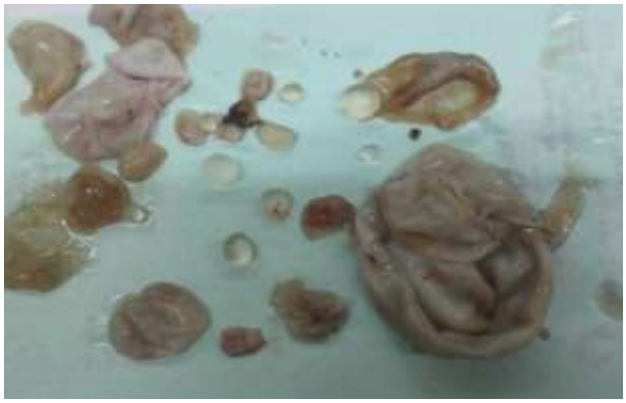

Figure 4: Gross image: multiple hydatid cysts with daughter cysts.

Gross findings: Left Nephrectomy specimen with multiple cysts is received. Upper pole is 8.5 x 5.0 x 6.5 cms, capsule is easily stripped off, outer surface is irregular with multiple scarring, on c/s, there is thinning of cortex. Lower pole is replaced by large cyst, size 16.0 x 10.0 x 6.0 cms, wall thickness is 0.3-0.4 cms. Ureter is 16.8 x 0.8 x 0.8 cms, proximal portion of ureter is dilated (Figure 2-4).

Multiple flat membranous and oval cysts seen, grayish white (Figure 3), translucent and soft with smooth surfaces largest is 9.0 x 6.0 x 1.0 cms, collectively 12.0 x 10.0 x 1.5 cms, cyst wall is 0.1 – 0.2 cms thick, contains gelatinous yellowish material.

Microscopic findings

Sections from cyst show lamellated cyst wall (Figure 5), cyst wall is dense fibrous at places with focal calcification, hooklet-like structure is identified in the cyst contents in Haematoxylin and Eosin (H.E) slides after carefully examining throughout the specimen. The hooklet-shaped structure is best seen with the condenser down.

Occasional proto-scolices seen (Figure 6) - consistent with morphology of hydatid cyst.

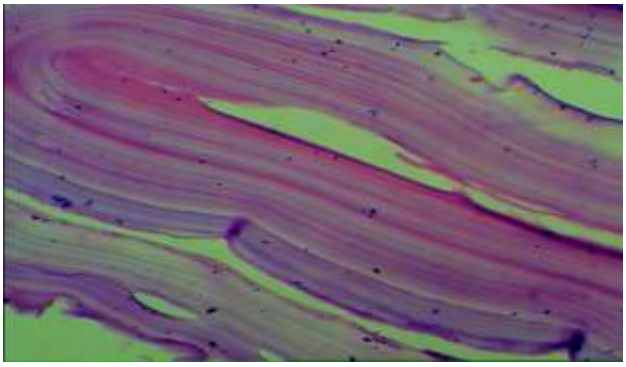

Figure 5: Microscopic image: revealed characteristic hydatidcyst with lamellated wall.

Section also show numerous glomeruli with changes of glomerulosclerosis, Tubules are atrophic, lumen containing colloid casts (thyroidisation of tubules) and interstitial tissue is heavily infiltrated with mixed inflammatory cells predominantly lymphocytes.

Sections also reveal arteriosclerosis. – Changes of chronic Pyelonephritis.

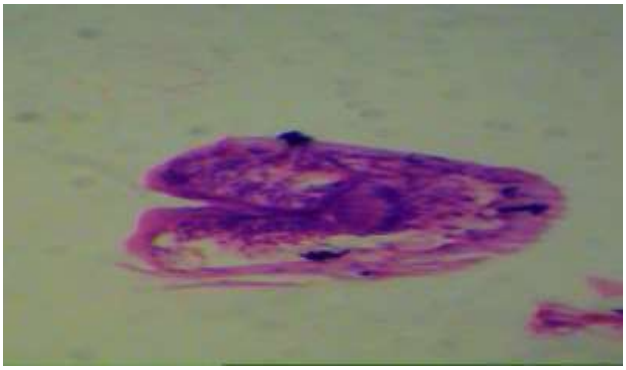

Figure: 6 Microscopic image: Hydatid cyst- Scolex.

Dilated portion of ureter reveals slight thinning of wall, congestion and mixed inflammatory cells. Distal portion of ureter reveal unremarkable pathology.

The case was reported as a parasitic infestation of Echinococcus Granulosus (Hydatid cyst) of left kidney with changes of Chronic Pyelonephritis.

Follow up

The patient remained asymptomatic. The right kidney and bladder appeared normal without hydronephrosis. No lesions were detected in the liver or spleen. The chest radiograph was unremarkable.

DISCUSSION

Echinococcosis is a zoonotic disease and caused by the tapeworm of the genus *Echinococcus*. It is endemic in Mediterranean and other sheep rearing countries. However, due to increasing travel and tourism it may be found even in developed countries. In India, annual incidences of Hydatid disease per 100000 persons vary from 1 to 2001. The liver is most common site of involvement2. Renal involvement is seen in 2% to 3% cases.[2,3]

Despite multimodality imaging, in a subset of patients, no definitive diagnosis can be made and the differential diagnosis often includes infected renal cysts, abscess, pyonephrosis and neoplasm. Rarely Type IV hydatid cysts may mimic hypo vascular renal cell carcinoma. On the other hand, in endemic countries, unusual neoplasm such as mucinous cyst adenoma/carcinoma may be misdiagnosed as renal hydatid.

Three of the four *Echinococcus* species are of medical importance in the human beings, including *Echinococcus Granulosus* that causes cystic Echinococcosis, *Echinococcus multilocularis* that causes alveolar Echinococcosis, and *Echinococcus* vogeli. *Echinococcus Granulosus* is the most common of the three. *Echinococcus* multilocularis is rare but is the most virulent and *Echinococcus vogeli* is the rarest.

The life cycle of *Echinococcus* involves two hosts, one definitive and the other intermediate. Humans act as an accidental intermediate host. The life cycle of *Echinococcus* has three developmental stages: (1) The adult tapeworms in the definitive host, (2) Eggs in the environment, and (3) The metacestodes in the intermediate host. The dog usually acts as the definitive host for this species and many mammals act as intermediate hosts, particularly sheep and horses.

After ingested by the definitive host, the metacestodes mature into the tapeworm in the definitive host and then release eggs into the environment. The egg ingested by the intermediate host hatches in the intestine, penetrates the gut wall, and travels through the lymphatic or blood vessels to the liver, lungs and other organs.

The liver acts as a first line of the defense and is most commonly involved (75%), followed by involvement of the lungs (15%) which act as the second site of filtration of the hydatid cyst. Haematogenous dissemination may lead to secondary involvement of almost any anatomical location.[4] Involvement of the kidneys is extremely rare

(2-3%).[2,3] Renal hydatid cysts usually remain asymptomatic for many years. It is postulated that the cysts pass through the portal system into the liver and retroperitoneal lymphatics to reach the kidneys.

The hydatid cyst of the kidney is considered closed if all three layers of the cysts i.e. pericyst, ectocyst and endocyst are intact. When the cyst is no longer protected by the third layer i.e. pericyst or by the lining of collecting system it is considered to be an exposed cyst. If all the three layers of the cyst have ruptured resulting in free communication with the calyces and pelvis, it is called an open or communicating cyst. Cystic rupture into the collecting system, causing hydatiduria is pathognomonic of renal Hydatidoses, though it is usually microscopic and is seen in only 10-20% of renal Hydatidoses.[5]

Table 1: Sonographic appearance of hydatid cysts in Gharbi classification.

Type	Sonography appearance
I	Well-defined, purely anechoic lesions that may be indistinguishable from simple renal cysts. Multiple echogenic foci due to hydatid sands may be seen in the cyst (22%)
II	Focal or diffuse detachment of the inner germinal layer results in a floating membrane inside the cyst (4%)
III	Multi-septated cysts with multiple daughter cysts (54%)
IV	Heterogeneous, solid appearance with infolded membranes, and internal echoes (12%)
V	Solid appearance, calcifications in the cyst wall, and germinative membranes (8%)

Gross hydaturia is uncommon, but diagnostic of the condition. The cysts passed in the urine are daughter cysts; hence they lack the third layer pericyst, which is contributed by the host around the mother cyst. Eosinophilia is noted in about 50% cases. Serological tests in primary renal hydatidoses are usually negative. The mainstay of diagnosis is by advanced radiological techniques like CT scan and magnetic resonance imaging.[6,7] The stage of cyst growth (i.e., whether the cyst is unilocular, contains daughter cysts, or is partially or completely calcified) determines the findings on radiological imaging studies.

Difference in the cyst content leads to difference in attenuation and signal intensity between the fluid in the central portion of the cyst and in the peripheral cysts. This is a typical finding in Echinococcosis. When daughter cysts are separated by the hydatid matrix, they demonstrate a "wheel spoke" pattern. The matrix represents hydatid sand containing membranes of broken daughter vesicles and scolices. The cytotoxic effects of the vesicular fluid may result in an exuberant granulomatous response by the host's immune system resulting in 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.

In an appropriate clinical scenario, detection of a cystic lesion with internal septations and sand, wall calcifications, or the rosette sign is usually suggestive of renal Hydatidoses. In the human beings, hydatid cyst disease most commonly occurs in the liver (55-70%) followed by the lung (18-35%), and these two organs can be affected simultaneously in about 5-13% of cases.[8,9]

Gharbi et al, classified hydatid cysts based upon sonographic morphology into five types (Table 1). Pre-operative diagnosis of hydatid cysts can be made ultrasonically and confirmed by a CT scan.[10] The CT scan has an accuracy of 98% to demonstrate the daughter cysts, and it is the best test to differentiate hydatid cysts from amoebic and pyogenic cysts in the liver.[11] A thin rim of calcification delineating a cyst is suggestive of an echinococcal cyst. MRI offers no real advantage over CT scan. Several serological tests can be used for diagnosis, screening, and post-operative follow-up for recurrence. These include the hydatid immune-electrophoresis, Enzyme-linked immunosorbent assay (ELISA), Latex agglutination and Indirect haemagglutination (IHA) test.[12]

The morphological diagnosis by the pathologist is frequently made by the presence of hydatid elements, especially hooklets. In our case, the hooklets are very difficult to find under transmitted light in H.E slides even by carefully examining through the specimen. With the use of special stain like Ziehl - Neelson stain or Trichrome stain, the hooklets are stained purple-blue and pink-red respectively and many hooklets are easily to identify over the background. These two stains offer significant advantage in morphological diagnosis of hydatid cysts, especially very old lesions, where hydatid components may be very difficult to identify.

Histologically, Hydatid cyst comprises of pericyst or fibrous layer, middle lamellated membrane and inner germinal layer which produces scolices. In patients with ruptured cysts and hydatiduria the membranes do not reveal pericyst.[6] Isolated renal hydatid involvement presents a further diagnostic challenge (as occurred in our patient). In our opinion, hydatid cyst should always be considered in the differential diagnosis of isolated complex cystic renal lesion. Absence of relevant history or hepatic involvement should not prevent diagnosticians from entertaining this rare diagnosis.

Surgery is the treatment of choice in cases of renal hydatid cyst. Kidney sparing hydatid cyst removal is possible in most cases (75%). Conservative surgery included simple internal endocyst excision plus drainage, internal capsule excision and external pericyst wall resection. The management of simple cyst is entirely for its symptoms or complications.

During kidney-sparing surgery scolicidal solutions such as hypertonic saline should be used before opening the cavities to kill the daughter cysts and therefore prevent further spread or anaphylactic reaction.

Pre and postoperative course of Albendazole is recommended to sterilize the cyst, decrease the chance of anaphylaxis and decrease the tension in the cyst wall thus reducing the risk of spillage during surgery and recurrence, post operatively. Results of simple aspiration are associated with very high recurrence rate (upto 90 %).[13-16]

Total or partial Nephrectomy is recommended when the hydatid cyst lesions breaking into the collecting system, rupture, infection and a serious kidney injury.[17-19] Radical surgery included pericystectomy for the renal hydatid and partial Nephrectomy.[20-22] Incidence of hydatid in left kidney is high than the right kidney (Left renal artery is shorter than the right renal artery, so there is high chance of hydatid larvae spread to left kidney first).

The procedure of preference is the simple cyst excision. When the kidney is damaged, Nephrectomy is necessary. Medical management of renal hydatidosis should considered as adjuvant therapy.[23-25] Chemotherapy, as an adjuvant therapy, with or without puncture aspiration-injection-reaspiration is suitable for in-operable renal hydatid disease (PAIR).[26-27] Surgery may cure the patient completely but may not totally prevent recurrence.

The surgical treatment principle of renal hydatid should be based on residual renal function, hydatid cyst size, number, location and surgical techniques to determine the surgical plan.

CONCLUSION

If the kidney is badly damaged (as in this case) by lesion such as chronic Pyelonephritis, it is better to dealt with Nephrectomy along with cystectomy. Most cases of renal Hydatidoses are dealt with open surgery i.e. cystectomy with pericystectomy. More study is needed on such rare cases of primary renal Hydatidoses to define a standard treatment modality after comparing the various available options. Also minimally invasive approach needs to be studied in comparison with the open approach.

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