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Case Report

Surgery Section

Long-standing Renal Hydatid Cyst Mimicking an Angiomyolipoma: A Rare Case Report

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ABSTRACT

Primary hydatid cyst of the kidney is very rare and accounts for 1-4% of all hydatid disease. Authors hereby reports an unusual case of an isolated, long-standing renal hydatid cyst, mimicking an angiomyolipoma on imaging. A 72-year-old female, from a rural area, presented with pain in the right flank since two years. She also complained of mild fever off and on, since one year. A 10×10 cm, non tender lump was palpated in the right flank. Ultrasonography and computerised tomography showed a large right-sided renal mass measuring 10 cms and a diagnosis of angiomyolipoma was rendered. A right-sided open/simple nephrectomy was done. The kidney was large, cystic, firm and was sent for histopathological examination. On gross examination, a large, unilocular, thick-walled, cystic lesion was seen. Cut-surface showed heterogeneous, creamy coloured necrotic contents, with few nodular gelatinous masses. On microscopy, characteristic acellular, laminated structure of the ectocyst of *Echinococcus granulosus* was seen. Extensive calcification and ossification of the cyst wall with bone marrow production was also seen. Contents of the cyst were necrotic and no daughter cysts were seen. Adipose tissue of the bone marrow was mistaken on imaging for the lipomatous component of an angiomyolipoma. Isolated renal hydatid cyst is very uncommon and though active hydatid cysts do have characteristic radiological findings, they can be misdiagnosed as a benign renal tumours, renal cysts or abscesses. Old cases show degenerative changes as well as calcification and are difficult to diagnose preoperatively. Surgical treatment of renal hydatid cyst depends on the size, number of cysts, exact intrarenal location and residual renal function.

Keywords: Albendazole, Calcified cysts, *Echinococcus granulosus*, Hydatidosis, Renal cyst

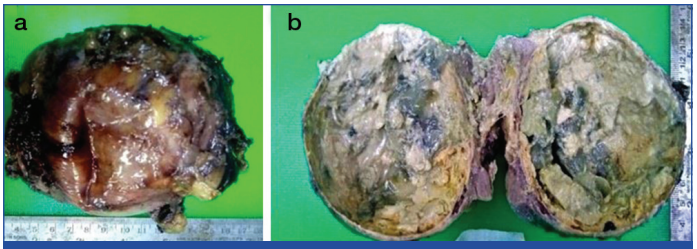
CASE REPORT

A 72-year-old female, from a rural area, presented with pain in the right flank since two years and mild fever off and on since one year. There was no past history of renal disease, parasitic disease, diabetes and hypertension. On examination, a 10×10 cm, non tender lump was palpable in the right flank. Haemoglobin was 11 gm/dL, total count 6300/cmm with neutrophils of 70%, lymphocytes of 28% and eosinophils of 2%. Peripheral blood smear showed normochromic normocytic morphology of red blood cells and adequate platelets. Fasting blood sugar was 116 mg/dL, blood urea nitrogen was 20 mg/dL and creatinine was 1 mg/dL. Urine analysis showed 2-3 epithelial cells per high power field, few oxalate crystals and no proteinuria. Ultrasonography revealed a large, right-sided renal mass measuring 9.3×8.5×8.4 cm in the upper pole and the interpolar region, and was reported as an angiomyolipoma with haemorrhage within. A computerised Tomography (CT) scan showed a 10.7×9.5×8 cm mass in the midpole of the right kidney with exophytic extension in the perirenal space and acute hemorrhage within fat planes, and a diagnosis of angiomyolipoma was given.

An open, simple nephrectomy was done via a retroperitoneal approach. The kidney was replaced by a large spherical, cyst-like, firm structure which was excised and sent for histopathological examination. No other intraabdominal pathology was noted.

On gross examination, a large, thick-walled unilocular cyst measuring 10×9×7 cm was received [Table/Fig-1a]. The cut surface showed heterogenous, creamy-coloured necrotic contents and multiple nodular gelatinous masses. Scanty residual renal parenchyma was seen at the hilum [Table/Fig-1b].

On microscopy, the cyst wall was composed of dense fibrous tissue, within which, was seen characteristic, eosinophilic, laminated, acellular structure of the ectocyst of *Echinococcus granulosus*. No daughter cysts with protoscolices were seen [Table/Fig-2a,b]. The residual renal parenchyma at the hilum showed dialated renal tubules with colloid casts [Table/Fig-2c]. The cyst wall also showed extensive calcification and ossification with bone marrow production while the contents of the cyst were necrotic material [Table/Fig-2d]. Calcified areas were seen as dark blue amorphous, clumped material formed by deposition of calcium salts while ossification shows formation of new bone tissue with bony trabeculae enclosing bone marrow. The inner surfaces of these trabeculae were lined by osteoblats and osteoclasts.

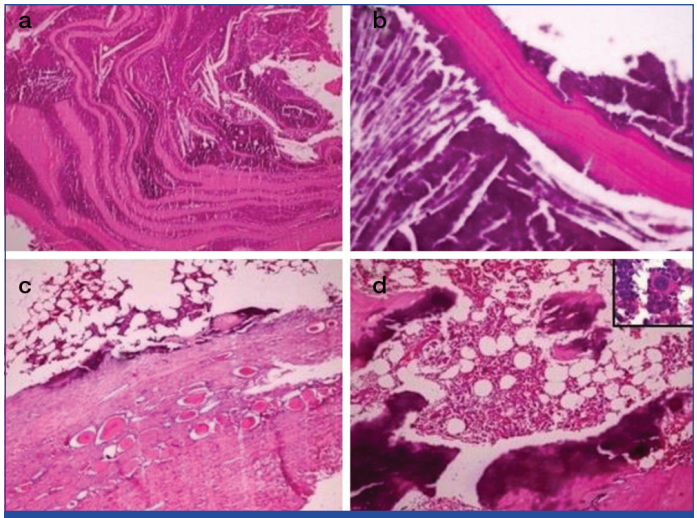


[Table/Fig-1]: a) Cyst-like kidney measuring 10×9×7 cms; b) Cut surface showing creamy- coloured necrotic contents with gelatinous nodules.

The patient was treated postoperatively with Albendazole 400 mg, twice daily, for four weeks. She was asymptomatic and disease free at a six month follow-up.

DISCUSSION

Primary hydatid cyst or echinococcosis of the kidney is very rare and is caused by the larval stage of the parasite *Echinococcus granulosus (E. granulosus)*. Humans are accidental dead end hosts. It occurs worldwide but is endemic in countries with sheep-rearing. Renal hydatid disease is rare and accounts for 1-4% of all hydatidosis [1]. Humans are accidental, intermediate, dead-end hosts, infected by consumption of water and food contaminated by the fecal matter of dogs. The eggs get converted into larvae in the gastrointestinal tract, these larvae migrate into the wall of the small intestine and enter the mesenteric circulation, and hence hepatic infection is very common followed by the lungs.



[Table/Fig-2]: a) Multiple, laminated ribbon- like structures,which are the ectocyst of *E. granulosus* seen in the cyst wall (H&E, 100X); b) High- power view of the characteristic laminated ectocyst of *E. granulosus* (H&E, 400X); c) Cystically dialated renal tubules with colloid casts seen in the residual renal parenchyma (H&E, 100X); d) Cyst wall showing calcification and bone marrow formation, surrounded by atrophic renal parenchyma (H&E, 100X). Inset shows a megakaryocyte (H&E, 400X).

The clinical presentation of the hydatid cyst disease of the kidney is usually a palpable mass with pain, in the lumbar or flank region or the patient may be aymptomatic for years. Hydatiduria is seen in 10-15% of the cases and is a pathognomic sign of renal hydatid disease [2]. Eosinophilia is present with active cysts. Serological tests in primary renal hydatid disease are often negative and not helpful. Clinical diagnosis requires a high index of suspicion. Diagnosis of the hydatid cyst is mainly by imaging with CT scan offering excellent diagnosis.

Imaging has a prime role to play in the diagnosis, classification and treatment of hydatid cysts. The Gharbi-World Health Organisation (WHO) radiological classification of the liver hydatid cysts with characteristic and diagnostic radiological features is commonly used. This classification though formulated for hepatic cysts can be used for hydatid cysts at other sites too. The Gharbi-WHO classification of Cystic Echinococcosis (CE) classifies hydatid cysts and into five classes as follows: CE1 and CE2 include active cysts; CE3 transitional cysts; CE4 and CE5 old degenerated and calcified cysts [3].

The cyst wall comprises three layers: the ectocyst which is white, translucent and has the appearance of the kernel of a tender coconut; the middle layer or the ectocyst which is acellular and laminated; the inner layer is called the endocyst or the germinative which has daughter cysts with protoscolices.

Complications of hydatid cyst are infection and rupture of the cyst leading to disseminated hydatidosis. Old, long standing hydatid cysts undergo degenerative changes with calcification. The differential diagnosis of hydatid cysts includes benign cysts, benign tumours and abscesses [1,4-6]. In the present case, the hydatid cyst was diagnosed on imaging as an angiomyolipoma, as the adipose tissue of the bone marrow was mistaken for the lipomatous component of an angiomyolipoma. Extensive ossification and bone marrow formation was seen in our case, and has not been reported in literature. Preoperative diagnosis of hydatid cysts is best made on imaging techniques like CT scan or Magnetic Resonance Imaging (MRI), but old long-standing hydatid cysts can difficult to diagnose radiologically. Angiomyolipomas are hypervascular lesions on radiology and often show abundant fat. Serological tests can also be used in the diagnosis of active hydatid disease but they are not reliable in old calcified hydatid cysts.

Treatment of hydatidosis involves both medical and surgical modalities. Treatment depends upon the location of the cysts and whether the cysts are active or inactive [6,7]. Albendazole is the drug of choice and it can be given preoperatively, during surgery and postoperatively. Surgical treatment of renal hydatidosis includes renal sparing surgeries, cystectomy with pericystectomy and extra peritoneal nephrectomy for destroyed or infected kidneys [7,8]. Laparoscopic nephrectomy involves risk of cyst rupture and dissemination.

CONCLUSION(S)

Isolated, old renal hydatid cyst is very rare, and though there are pathognomic radiological findings, it is often misdiagnosed as a benign renal tumour or a benign renal cyst or an abscess. Long standing hydatid cysts can undergo extensive ossification and calcification. Abundant adipose tissue of the bone marrow (ossification) can lead to a mistaken diagnosis of adipocytic lesions like angiomyolipoma, as was seen in the present case report.

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