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CASE REPORT

RENAL CELL CARCINOMA IN UNILATERAL AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE AN UNUSUAL PRESENTATION: A CASE REPORT

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ABSTRACT

Introduction: Adult polycystic kidney disease is characterized by bilateral multiple renal cysts which is often accompanied by cyst formation in liver and pancreas. Although the disease is bilateral, renal involvement may be asynchronous and asymmetrical. There are some reports on unilateral ADPKD in literature. Risk of renal cell carcinoma in patients with ADPKD is controversial and reportedly <1%. We are reporting a case of unilateral ADPKD with focus of RCC presenting as a huge mass.

Case presentation: Seventy years old male patient from South India was admitted with the history of abdominal distension, dyspnoea, cough with expectoration, fever and weight loss with a positive family history of ADPKD. On examination, a 30 × 25 cms mass was palpable in right hypochondriac, right lumber, right iliac, umbilical, left iliac and hypogastric areas. The mass was crossing the midline. Computerized tomography scan of abdomen showed a huge, predominately cystic, well defined mass occupying right hypochondriac, renal, iliac and umbilical regions. Computerized tomography guided Fine needle aspiration cytology showed moderately differentiated renal cell carcinoma. Intravenous pylography showed a grossly enlarged right polycystic kidney with multiple calculi (an incidental finding). Right radical nephrectomy was done, and about a 5 kg, hugely enlarged polycystic right kidney mass was removed. Patient recovered well postoperatively.

Conclusion: Bilateral ADPKD is common finding, but there are only a few reports on unilateral ADPKD; still rare is development of RCC in ADPKD. Our case adds to our knowledge; of unilateral presentation of ADPKD with the focus of RCC; which presented as a huge mass which was crossing the midline.

INTRODUCTION

ADPKD is the most common cystic disease of kidney and is seen in 1:500-1000 [1], characterized by bilateral multiple renal cysts often accompanied by cyst formation in liver and pancreas. Although disease is bilateral, renal involvement may be asynchronous and asymmetrical [2]. Unilateral cystic kidney posses a diagnostic dilemma although there are some reports on unilateral ADPKD in literature. [3] Walters and Bruasch first reported association of ADPKD and RCC in 1934. Risk of renal cell carcinoma in patient with ADPKD is controversial and reportedly <1%. [1] We observed one such puzzling case, with surprising explanation of unilateral ADPKD; presenting as a huge renal mass crossing midline with focus of RCC being diagnosed incidentally.

Case History

A 70 year old male patient from South India was hospitalized with the history of abdominal distention since 6 months, H/o dyspnoea and cough with expectoration since 4 months, H/o fever and weight loss was also present. Patient is a known asthmatic and a known history of being a smoker. The patient's mother expired due to renal disease and on retrospective screening among family members, bilateral renal cysts were found in the patient's daughter.

General Condition: Good.
Temp: Febrile
Pulse Rate: 76 beats per min and regular.
Blood Pressure: 110/80 mm Hg.
Respiratory Rate: 16 cycles per minute.
Patient was anemic.
CVS, CNS: Normal
RS: Generalized wheeze present.

Per Abdomen: Fullness of Right hypochondriac, Right lumbar, Right iliac, Umbilical, Left iliac and Hypogastric areas. Right renal angle was full on palpation: 30×25 cm mass palpable involving the Right hypochondrium, Right lumbar, Right iliac fossa, Umbilical region, Hypogastric area and the Left iliac fossa. The mass was crossing the midline. Fingers could be insinuated between the costal margin and the mass on Right side. It was firm in consistency, bimanually palpable and the renal angle was dull on percussion. [Refer Fig. 1]

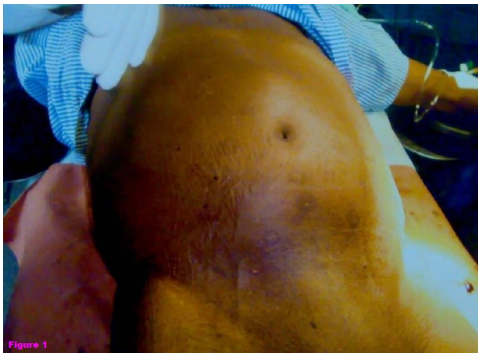

Figure 1. Clinical photograph of patient's abdominal lump crossing midline

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Investigations

Haemogram was normal, except Hb = 6.4 g % Blood glucose level, renal and liver function test were within normal limits USG abdomen showed right sided cystic disease of the kidney. CT scan of abdomen revealed huge, predominantly cystic, well defined mass occupying Right hypochondriac, lumbar, iliac, umbilical regions (having solid enhancing component, fat and calcified component, mass was seen separately from liver, Right adrenal gland with no obvious invasion or encasement of adjacent organs or vessels. No obvious lymphadenopathy or metastatic foci, Left kidney normal). Impression: Right renal polycystic mass or possibility of retroperitoneal teratoma with absent right kidney should be considered. [Refer Fig. 2] IVP: Grossly enlarged right polycystic kidney with multiple calculi-incidental finding. CT guided FNAC showed moderately differentiated renal cell carcinoma. Preoperatively chest physiotherapy and 3 units of blood transfusion was given and then patient was taken for surgery under epidural anesthesia.

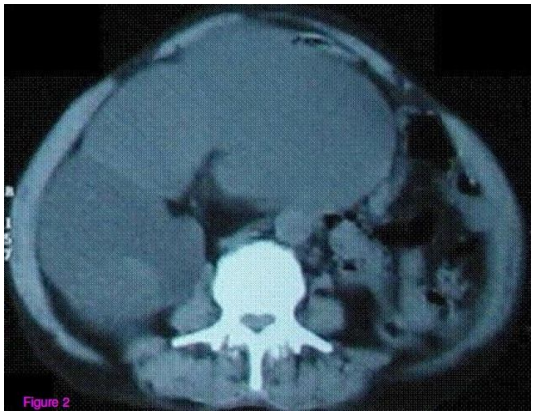

Figure 2. Abdominal CT Scan shows cystic renal mass crossing midline

Intra-operative findings

Hugely enlarged right polycystic kidney mass measuring about 30 x 25x 15cm, weighing about 5 kg, crossing the midline extending into right hypochondriac, right lumbar, right iliac, Umbilical, left iliac and hypogastric areas. Prominent enlarged veins on the surface. Cysts contained hemorrhagic fluid. There was involvement of Gerota's fascia in lower pole but no involvement of Gerota's fascia in upper pole and adrenal. Renal vein was normal. [Refer Fig. 3] Right radical nephrectomy done. Specimen was sent for HPR. Patient recovered well post-operatively.

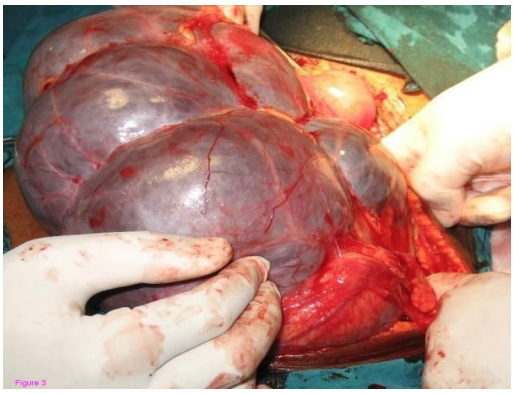

Figure 3. Intra-operative photograph, huge mass external surface shows typical polycystic appearance

Histopathological report

External Surface

Enlarged, cystic, but reniform in shape, grayish white in colour with congested blood vessel. [Refer Fig. 3]

Cut Section

Multiple cyst of varying size with translucent cyst wall was seen. It also showed a solitary grey white area measuring about 3 x 2 cm. Cyst showed serous and hemorrhagic fluid. [Refer Fig 4]

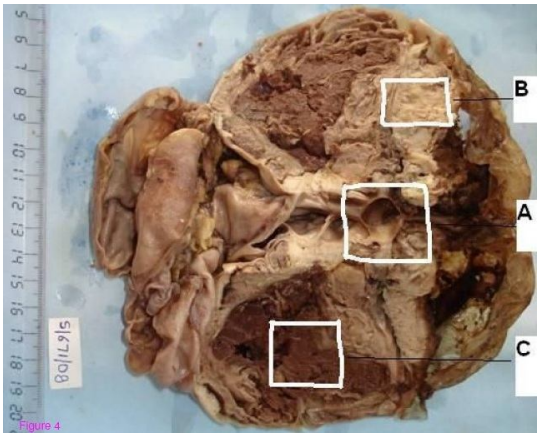

Figure 4. Cut surface shows multiple cysts with a solid component

Microscopy

Section from area (A) shows – cyst were lined with a single layer of cuboidal epithelium with hyperplastic epithelial cells were seen focally. Section from area (B) shows - tumor tissue composed of cells with moderate anisonucleosis, hyperchromatism and inconspicuous nucleoli, arranged in a papillary-trabecular, papillary solid pattern with fibro vascular core. Section from area (C) shows hemorrhage and necrosis. [Refer Fig.5 and 6] Impression: Papillary renal cell carcinoma (type 2) with polycystic kidney.

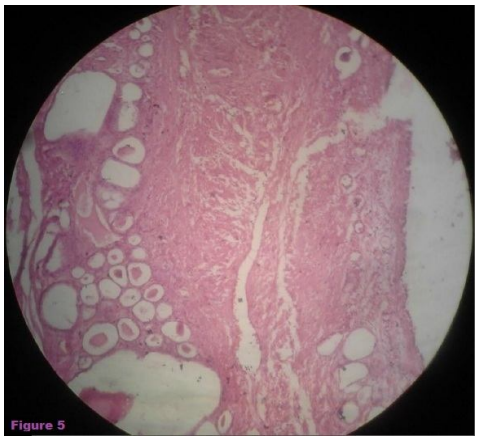

Figure 5. Histopathological section of cyst wall. From area A on cut section

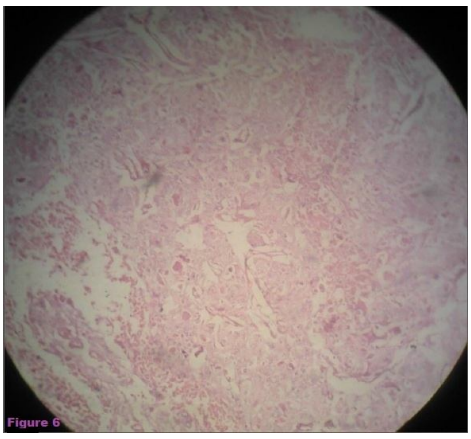

Figure 6. Histopathological section showing Papillary Renal cell carcinoma of type II variant. From area B on cut section

DISCUSSION

When patients with ADPKD present clinically, they usually are found to have bilateral cysts. However the disease can manifest asymmetrically with cyst on one side at first or with an unilateral renal mass, 20 to 30% patients with ADPKD develop stones. [4] In adult unilateral polycystic kidney disease has been rarely described in urological literature. Kossow and Meck reported a 79 year old woman with unilateral cyst but these were apparently of multi-cystic variety. The patient of Lee *et al* was aged 72 years but there was no family history of APDKD and there were multicystic cysts, in one kidney without cyst formation in contralateral kidney. Here the diagnosis is made by Histopathological similarities to ADPKD. Ohno *et al.* described a case of newborn with unilateral cyst formation, normal contralateral kidney and healthy parents here based on histology, a diagnosis of ADPKD was made. [2] In both cases based on histology a diagnosis of ADPKD was made. In our case diagnosis is confirmed by family history and histopathological similarities with ADPKD. Walter and Bruasch first reported association of ADPKD and RCC in 1934. Risk of renal cell carcinoma in these patients is controversial and reportedly <1%. [1] Majority of the reported cases have been shown to be an incidental finding, either during surgical exploration or at autopsy. [5] Most reports of renal cell carcinoma in ADPKD have been of patients not under treatment for chronic renal failure. However, the risk may be increased in patients who retain their native kidneys during long term treatment with haemodialysis. [6] There are no clinical features which could indicate the presence of RCC in ADPKD. [7] Clinical recognition of the renal cell carcinoma in ADPKD is often difficult because of preexisting renal cell mass and occasional haematuria. Renal cell carcinoma should be suspected when there is abdominal pain, back pain, severe haematuria and a new renal mass in an adult with ADPKD. [8] Renal cell carcinoma in ADPKD may be bilateral and multicentric. [9] Ultrasonography often fails to reliably differentiate between benign hemorrhagic cysts from complications with the ADPKD. [10]

CT scan characterizes any malignant mass on ADPKD cyst and CT guided FNABC of cyst of interest would certainly give reliable information. [7] Differential diagnosis of our case are Giant hydronephrotic kidney and large cystic RCC. Giant hydronephrosis in the presence of more than one liter of fluid in the collecting system is the rare urological entity in adults. [11] Cysts of hydronephrosis communicate with the pelvis. [12] Which is not seen in our case. Regarding Large Cystic RCC – Apart from being usually focal and unipolar, the rate of occurrence of cysts in RCC is variable. Cystic areas are rare in chromophobe RCC. [13] Cystic changes occur in up to 15% cases in clear cell renal cell carcinoma. Cystic changes are common and rarely may produce a predominately cystic lesion. [14] These are differentiated from ADPKD by microscopy of the tumor cells lining the cysts, cyst wall being thick and fibro-collagenous. In clear cell RCC tumor cell have structure less cytoplasm with well defined cell border. Nuclei round to oval and regular although heterogeneity can exist in a single tumor. The third type of RCC, papillary RCC, in some instance, this tumor presents as cystic mass. The cut surface is friable giving impression of extensive necrosis. [15] In our case the kidney had cysts involving the kidney diffusely and not unipolarly (like in a large cystic RCC) and translucent cyst were lined by single layer of cuboidal epithelium and there was evidence of normal renal components surrounding the cyst which is unlikely in large cystic RCC.

Conclusion

ADPKD are usually bilateral with only some reports on unilateral ADPKD in literature. To add on, ADPKD is rare to develop renal cell carcinoma and majority of reported cases are incidental findings either during surgical exploration or autopsy; our case is unusual as it is unilateral ADPKD with focus of RCC and the mass is crossing the midline.

Abbreviations

RCC, renal cell carcinoma; ADPKD, autosomal dominant polycystic kidney disease

Consent

Written informed consent was obtained from the partner of the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

NN and RK performed the radical nephrectomy. NN and JA analyzed and interpreted the patient clinical data and was also major contributor in writing the manuscript. NB helped in overall pathological study. AZ, SI, SD and VB assisted in the overall study.

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