ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 03 | March - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

POLYCYSTIC KIDNEY DISEASE ASSOCIATED WITH RENAL CELL CARCINOMA: CASE REPORT

Pathology

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ABSTRACT

Adult Polycystic Kidney Disease is the most common cystic disease seen in 1: 500-1000 patients. Co-existence of RCC and renal cyst in same kidney has been reported in only 1-2% of cases of RCC. We present a case of a 57-year old male who presented to the Department of Urosurgery with complaints of left loin pain for the past 6 months which was radiating to left leg. Detailed history, examination and imaging studies suggested a mass in the upper pole of left kidney, multiple cortical cysts, non obstructing calculi noted in bilateral kidneys associated with simple hepatic cyst. Per operative findings revealed a renal mass at the upper pole of left kidney which was sent for histopathological examination that showed features of Tubulopapillary variant of Renal Cell Carcinoma. Details will be presented.

KEYWORDS

renal mass, renal cell carcinoma, Polycystic Kidney Disease, cortical cysts.

INTRODUCTION

Adult Polycystic Kidney Disease is the most common cystic disease seen in 1: 500- 1000 patients. Co-existence of Renal Cell Carcinoma and renal cyst in same kidney has been reported in only 1-2% of cases of RCC.

Case Report

A 57 year old male presented to Urology OPD with complaints of left loin pain radiating to leg since 6 months, intermittent type of fever since 15 days. He is a known case of Hypertension and Type II Diabetes since 4 months using regular medication. He is a alcoholic and non-smoker. On general examination left loin tenderness present.

Laboratory workup: Complete Blood Picture - Normocytic normochromic anemia (Hb: 4.92g/dl) ; Leucocytes are within normal limits (7330 cells/microlitre); Platelets adequate(1.78 Lakhs). FBS-103mg/dl. CUE - Blood- Negative; Bilirubin –negative; Urobilinogen- negative; Ketones- Trace; Glucose- negative; Proteins-30mg/dl; Ph-5; Nitrites- negative; Leucocytes-negative; Specific gravity- 1.025. Serum Creatinine- 0.9 mg/dl Blood Urea – 37.4mg/dl , HbA1c – 6.6%

CT Abdomen – 1.Bilateral Polycystic kidney disease
2.Bilateral non obstructing renal calculi
3. Simple hepatic cyst
4.Evidence of well defined lobulated, exophytic soft tissue density lesion measuring 62.6 x 5.6 x 48.8 mm (CC x T x AP) noted in upper pole of left kidney involving cortex and medulla – likely benign lesion (Oncocytoma). Lesion showing homogenous enhancement on post contrast. No evidence of calcifications. Lesion is maintaining fat planes with perinephric tissues.

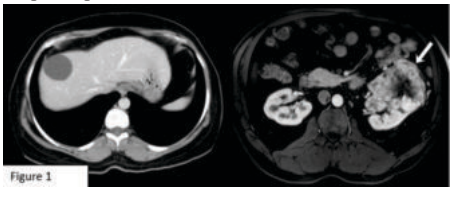


Figure 1

Patient diagnosed as left renal mass and underwent laparoscopic left partial nephrectomy and specimen sent for histopathological examination. We received Left Partial nephrectomy specimen measuring 7x6x4 cm, weighing 110 gms. Hilar structures and ureter are not visualized. External surface- Capsule and perinephric fat present, areas of congestion seen. Cut surface- solid grey white to grey brown to grey tan areas, soft, friable necrotic areas seen.

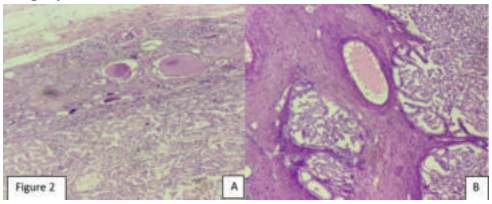
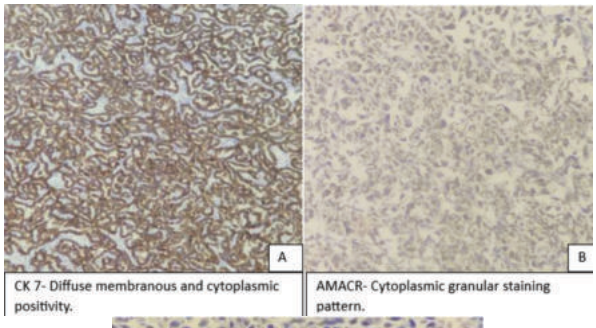
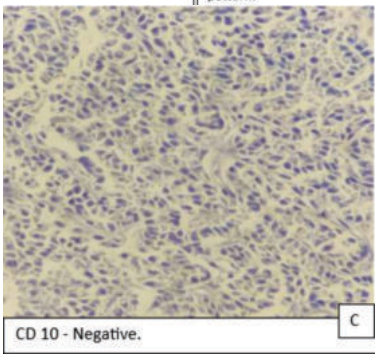


Figure 2 A B

A= 10x H&E section showing normal kidney with capsule and tumor tissue with cells arranged in tubulopapillary pattern. Cells show moderate eosinophilic cytonuclear pleomorphosis, irregular nuclear borders.

B= 40x H&E section showing capsular invasion.


A
CK 7- Diffuse membranous and cytoplasmic positivity.
B
AMACR- Cytoplasmic granular staining pattern.


CD 10 - Negative.
C

DISCUSSION

The association of renal cell carcinoma and polycystic kidney disease was first described in 1934 by Walters and Braasch[1]. Since then, over 60 cases of this association have been published in English literature. The diagnosis of RCC in a background of PKD poses several challenges. An evolving focus of RCC in a background of multiple renal cysts and distorted architecture may be difficult to discern in early stages even on regular follow-up. Symptoms from malignancy such as pain, hematuria, mass effect, or hypertension may be confused with cyst rupture or hemorrhage[2]. A literature review of 25 cases of RCC in background of ADPKD compared to RCC in general population reported no gender predilection, earlier age at presentation (45 vs. 61 years), higher occurrence of fever as a presenting symptom (32% vs. 7%), with higher incidence of bilaterality, multicentricity, and sarcomatoid features[3]. Surgical series in ADPKD have also documented multifocality and bilateral renal involvement with RCC. Papillary and clear cell RCC were the most common reported pathologic diagnoses in these series and nearly a third of patients had more than one histologic subtype[4,5]. Metastatic disease is noted in

nearly 20 to 23% patients with RCC in the setting of ADPKD, possibly due to a delay in diagnosis[3,6]. The postulated hypotheses of RCC development in ADPKD include chronic renal injury favoring renal parenchymal genetic mutations with consequent malignant change, or hyperproliferation in ADPKD acting as a precursor to RCC[7,8]. None of these theories have been substantiated yet. Some studies also demonstrate increased apoptosis in cystic and non cystic structures in ADPKD, negating the malignant potential in this condition.

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