AN UNUSUAL PRESENTATION OF COMPLEX RENAL CYST

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Abstract:
Renal cell carcinoma is the most common neoplasm of the kidney. It is a heterogeneous disease, comprised of different histological variants with a distinct clinical course, genetics and response to treatment. The various subtypes identified include clear cell, papillary and chromophobe, among others. Chromophobe renal cell carcinoma is a rare variant and accounts for 5 of all cases. The coexistence of different subtypes of renal cell carcinoma (RCC) within a single kidney is an extremely unusual entity. Presented herein is the case of a 60-year-old woman with RCC of chromophobe and papillary histology. Very few reports in the literature describe double or triple-synchronous renal neoplasms. To our knowledge this is the second report of this RCC subtype combination, which might trigger further investigation on the RCC pathogenesis theories. This patient underwent partial nephrectomy and on follow-up was found to be free of metastasis or local recurrence.

Keyword: Papillary, chromophobe, renal cell carcinoma, partial nephrectomy.

Case report:
A 60 year old lady presented to our institution with complaints of abdomen pain for one month. She did not have any other significant associated symptoms. She was seen initially at our medical gastroenterology department and had undergone upper GI Scopy which was normal and CT scan abdomen which demonstrated the presence of a renal tumor of the lower pole of the left kidney following which she was referred to our urology department.

Past history:
Patient had undergone appendicectomy 20 years ago, vaginal hysterectomy 5 years ago and cholecystectomy with mesh repair for ventral hernia (indication: cholelithiasis) 2 years ago, perioperative period uneventful.
On examination:
General examination was normal and vitals were stable
On per abdomen examination, abdomen soft scar present was healthy
No mass palpable renal angle free
B/L hernia orifices free
Bowel sounds present, no ascites
CVS & RS: Normal Spine & Cranium: Normal

Investigation: HB: 10.5gm
% , TC : 6000 cells / cumm , DC : P75% L20% E5% , ESR :6 / 15m m , Platelet – 2 lakhs / cumm
Blood sugar :120mg/dl, B. Urea:19mg/dl, Sr.Creatinine:0.6mg/dl
Urine Routine – alb/ sugar- nil , Deposits – pus cells – 1-2/ hpf
Urine c/s : No growth, Sodium: 142mEq/L , Potassium: 3.8mEq/L
LFT: Bilirubin (T): 0.8mg/dl, Protein: 7.6g/dl, Albumin: 4.9g/dl, SGOT:43 IU/L, SGPT:42IU/L, ALP: 90 IU/L
ECG , CXR - normal

USG KUB:

FIGURE 1: USG KUB: 4x3cm mixed echogenic lesion present at lower pole of left kidney

Angiogram:

Hypodense lesion 3.7 X 3.6 X 2.9cms with septations seen in the lower pole of left kidney with moderate enhancement
Renal artery: Right side double arteries, Left side single artery with no prehilar branching. Renal vein: single on both sides.

FIGURE 2a & 2b: CECT KUB showing complex cystic lesion in the lower pole of left kidney suggestive of Bosniak Class 4
FIGURE 3a & 3b: Renal angiogram
MANAGEMENT:
Left renal tumour (stage I – T1N0M0)
PLAN: Nephron sparing surgery (Left – Lower polar nephrectomy)
Procedure
Uder ETGA, through extra peritoneal left flank incision approach. Early vascular control was obtained. Complete tumor excision made along with perinephric fat with 1cm free margin. Proper closure of collecting system. Renorrhaphy done. Wound closure done.

FIGURES 4a, 4b, 4c & 4d: Left partial nephrectomy

HISTOPATHOLOGY:

FIGURES 5a & 5b: Papillary area. Infiltrating neoplasm composed of papillary area intervened by cyst

FIGURE 5c: Enclosing numerous macrophages, psammoma bodies along with nest of round to polyhedral cells. Some with granular & abundant pink cytoplasm, vesicular nucleus & prominent nucleolus.
DISCUSSION:
RCC is the most frequent neoplasm of the kidney. Renal cell carcinoma accounts for 2% to 3% of all adult malignant neoplasms, most cases being sporadic. All RCCs are, by definition, adenocarcinomas, derived from renal tubular epithelial cells. Approximately onethird of patients present with advanced disease at diagnosis, and recurrence occurs in 30-40% of cases treated for a localized tumor.

RCC is a heterogeneous disease, comprised of different histological variants with a distinct clinical course, genetic changes and responses to systemic treatment. The categorization of RCC is based on the World Health Organization (WHO) classification, which includes different subtypes based on morphology, including clear cell, papillary, chromophobe, granular, spindle cell, cystic-associated, translocation carcinomas and collecting duct carcinomas.

Distribution:
Clear cell RCC is the most common adult RCC, representing 70% of all RCCs. Papillary RCC accounts for 10%–15%, Chromophobe RCC for 4%–6%, Collecting duct carcinoma for less than 1%, and unclassified lesions for 4%–5% of RCCs.

Sarcomatoid dedifferentiation represents the high-grade end of all subtypes.

PAPILLARY RCC:
Second most common histologic type 10 to 15%. Two distinct variants of papillary RCC have been described by characteristic cytogenetics, immunostaining profiles and gene expression profiling. Type 1 papillary RCC, the more common form, consists of basophilic cells with scant cytoplasm; type 2 papillary RCC include potentially more aggressive variants with eosinophilic cells and abundant granular cytoplasm. The two subtypes of papillary RCC correspond with two familial RCC syndromes: hereditary papillary RCC syndrome (type 1) and hereditary leiomyomatosis and RCC syndrome (type 2).

Most commonly affects END STAGE KIDNEYS and ACQUIRED CYSTIC DISEASE. Lymph node involvement is most common in Papillary RCC. Macroscopically, typically appear hypovascular and homogeneous large tumors often contain areas of hemorrhage, necrosis, and cystic degeneration, fleshy tumor with fibrous pseudocapsule. Necrosis an...
Hemorrhage are common; Variegated appearance with dull yellow to dark brown. Bilateral and multifocal tumors are more common.

**CHROMOPHOBES RENAL CELL CARCINOMA**

Chromophobe RCC, is a distinctive histologic subtype of RCC that appears to be derived from the cortical portion of the collecting duct. It represents 3% to 5% of all RCCs. The tumor cells typically exhibit a relatively transparent cytoplasm with a fine reticular pattern that has been described as a “plant cell” appearance. The chromophobic nature a perinuclear clearing or “halo” is typically found and microscopic findings consist of numerous 150 to 300 nm microvesicles, which are the single most distinctive and defining feature of chromophobe cell carcinoma. These microvesicles characteristically stain positive for Hale colloidal iron, indicating the presence of a mucopolysaccharide unique to chromophobe RCC. Most studies of the clinical behavior of chromophobe RCC suggest a better prognosis for localized chromophobe RCC than for clear cell RCC. The coexistence of different subtypes of renal cell carcinoma (RCC) within a single tumor of RCC of chromophobe and papillary histology. Very few reports in the literature describe double or triple synchronous renal neoplasms. To our knowledge this is the second report of this RCC subtype combination, which might trigger further investigation on the RCC pathogenesis theories. This patient underwent partial nephrectomy and on followup was found to be free of metastasis or local recurrence.

**REFERENCES:**


