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Tubulocystic renal cell carcinoma in the right kidney – a case report

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ABSTRACT *Tubulocystic renal cell carcinoma is a rare type of renal cell carcinoma. About 100 cases have been reported till date. Here we report a case of Tubulocystic renal cell carcinoma in a young male who presented with abdominal pain and intermittent hematuria . Ultrasonographically, a diagnosis of right renal mass was made. Patient underwent right radical nephrectomy . A diagnosis of tubulocystic renal cell carcinoma was made based on gross finding of multiple cysts and histopathological finding of variable sized tubules and cysts lined by cuboidal cells having eosinophilic cytoplasm, nearly spherical nuclei with hobnailing at some places and prominent nucleoli .*

KEYWORDS : Tubulocystic renal cell carcinoma, Hobnail appearance, Bubble-wrap appearance.

INTRODUCTION:

Tubulocystic renal cell carcinoma of the kidney is a recently known distinct type of renal cell carcinoma. Tubulocystic renal cell carcinoma was first described by Pierre Masson in 1956 who believed it to be originated from collecting ducts of Bellini and he named it "Bellinian epithelioma" or "carcinoma of the collecting ducts" (1). It was also termed as "low-grade collecting duct carcinoma". In 2004, Amin et al. named the tumor "Tubulocystic carcinoma" in a series of 29 cases (2) .

Tubulocystic renal cell carcinoma was not included in the WHO 2004 classification. However, it was recognized as a distinct entity in 2010 by the American Joint Committee on Cancer. In 2012, it was included in the Vancouver Classification of renal cancer (3) . About 100 cases have been reported till date. Here We report a case of Tubulocystic renal cell carcinoma in the right kidney of a young male.

CASE REPORT :

A 20 -Year -old male came to surgery OPD of RIMS , Ranchi with history of dull right flank pain and intermittent hematuria for last 5-6 months. There was no history of fever, significant weight loss and loss of appetite. General physical examination revealed mild pallor. Abdominal examination revealed mass in the right lumbar region which was non tender, firm, well defined borders and moving minimally with respiration . There was no palpable superficial lymph node. Laboratory investigations were with in normal limits except blood hemoglobin was 10 gm/dL . Abdominal ultrasound showed right renal mass with cystic lesions. Contrast enhanced computed tomography (CECT) scan of abdomen showed heterogeneously enhancing well defined mass with cystic changes in the right kidney. There was no involvement of adjacent organ or retroperitoneal lymph node. Patient underwent right radical nephrectomy .

Grossly the external surface of the tumor was smooth and cut surface revealed multiple cysts having clear serous fluid (Fig.1). Tumor was confined within the kidney . The overlying renal capsule was intact and there was no vascular invasion.

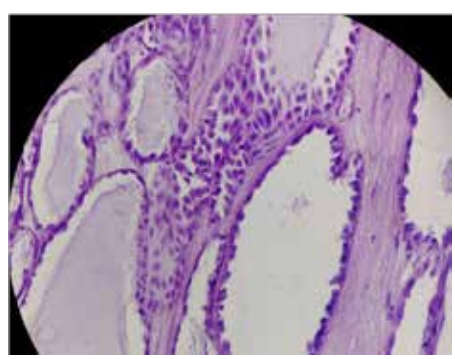

Fig 1: right nephrectomy specimen showing multiple cysts

As shown in Figure 2 and 3, Histopathology revealed tubules and Cysts of variable sizes lined by cuboidal cells having eosinophilic cytoplasm. Nuclei were nearly spherical showing mild pleomorphism and hobnailing at places. Prominent nucleoli were seen . Cysts were separated by fibrous stroma . The fibrous stroma separating the tubules and cysts were pauci cellular. A final diagnosis of Tubulocystic renal cell carcinoma was made on the basis of histomorphological features.

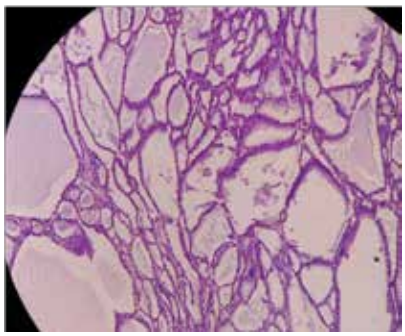

Fig 2. low power view

Fig 3: high power view

DISCUSSION:

Kidney cancer is among the ten most common causes of cancer related death in adults. Among the renal cell carcinoma, Clear cell carcinoma is the most common type of renal cell carcinoma. Tubulocystic renal cell carcinoma is a rare type of Renal cell carcinoma.

Tubulocystic renal cell carcinoma is usually discovered incidentally. Sometimes it presents with nonspecific abdominal pain, flank mass , gross hematuria and weight loss . Tubulocystic renal cell carcinomas of the kidney occur most commonly in the fifth and sixth decades of life and it shows a strong sex association with the male to female ratio being 7:1 (4) . Majority tumors involve the renal cortex or the cortex and medulla , not the medulla alone, which would be the typical location of a collecting duct carcinoma .

Grossly ,the Tubulocystic renal cell carcinoma is usually solitary and well circumscribed and composed of multilocular cystic spaces, some containing serous fluid. Cut surface is characteristically described as"-bubble-wrap", "spongy" or "Swiss cheese" in appearance due to the multiple cystic spaces (4). Tumors typically do not show areas of solid growth. Hemorrhage or necrosis is uncommon.

Histopathologically , tumors are composed of closely packed tubule**s** and cyst**s** separated by fibrous stroma and lined by a single layer of cuboidal cell(5). Cytolplasm are eosinophilic. Nucleoli are prominent . Hobnail appearance are seen at some places. Cysts range from 0.05 to 2mm, but can be as large as 1 cm. Mitoses are rare or absent**.** Necrosis or lympho vascular invasion is usually not identified. Common findings associated with Tubulocystic renal cell carcinoma is the coexisting Papillary renal cell neoplasms.

Differential diagnosis for Tubulocystic renal cell carcinoma include Multilocular renal cyst , Cystic nephroma, Multilocular cystic Clear renal cell carcinoma , Mixed epithelial and stromal tumor of the kidney and collecting duct carcinoma.

Tumors are positive for CD10 and AMACR (P504S). Weak or heterogeneous staining for CK7 and Strong staining is seen for CK8, CK18 and CK19. It is also positive for Pax2, kidney specific cadherin, carbonic anhydrase IX, and parvalbumin . Chromosome 7 gain is specific for Tubulocystic renal cell carcinoma.

Tubulocystic renal cell carcinoma has favourable prognosis. The biological behavior of this neoplasm is generally indolent and the stage is typically low at presentation .

Overall, Tubulocystic renal cell carcinomas have a small but definite risk for metastasis, and are best considered a low malignant tumor. This risk is markedly increased when Tubulocystic renal cell carcinoma is associated with high grade papillary renal cell carcinoma.

Radical nephrectomy is generally recommended, but partial nephrectomy may be performed for small tumors located in the superficial renal cortex. Sunitinib, a tyrosine kinase inhibitor, may exhibit a partial response or temporary effect for this tumor (6). Tubulocystic renal cell carcinoma of the kidney with sarcomatoid change has responded poorly to sorafenib (7).

CONCLUSION:

Tubulocystic renal cell carcinoma is a rare type of renal cell carcinoma. Although this type is more common in the fifth and sixth decades of life ,here this type was reported in a young male patient. Tubulocystic renal cell carcinoma has specific macroscopic, microscopic and immunohistochemical findings. Study of more cases is required to better understand the biological behaviour of this neoplasm as well as to know the prognosis and treatment of this rare entity.

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