

**Primary Carcinoid Tumor of Kidney masquerading as Renal Cell Carcinoma****Dr. Sonia Chhabra<sup>1</sup>, Dr. Sunita Singh<sup>2</sup>, Dr. Mansi Agarwal<sup>3\*</sup>, Dr. Ashima Batra<sup>4</sup>**<sup>1,2</sup> Professor, <sup>3,4</sup> Resident; Department of Pathology, Pt. B.D. Sharma PGIMS, Rohtak, Haryana, 124001**\*Corresponding author**

Dr. Mansi Agarwal

Email: [agarwal.mansi610@gmail.com](mailto:agarwal.mansi610@gmail.com)

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**Abstract:** Carcinoid tumor of the kidney is rare. It is a tumor with low grade atypia having relatively low malignant potential and metastatic spread with a good prognostic index. In Case history A 31 year old male presented with painless gross hematuria and lumbar pain for 2 months. Renal cell carcinoma was suspected and left radical nephrectomy was done. It was diagnosed as primary carcinoid tumor of kidney on histopathology. In Conclusion the disease is rare and hence attains a low index of clinical suspicion. However, its good prognosis at early stages makes an early diagnosis even more important.**Keywords:** Primary renal carcinoid tumor, good prognosis.

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**INTRODUCTION**

The first ever case of carcinoid tumor was described histologically almost 100 years back by Lubarsch in ileum in 1888 [1]. About 70% of carcinoid tumors occur in the gastrointestinal tract and the rest arise mainly from the neuro endocrine cells in pancreas, gonads and pulmonary bronchi. Carcinoid tumor of the kidney is rare [2]. The clinical manifestations of carcinoid tumors may be limited to local symptoms or may present as features of carcinoid syndrome. [3]. However, it has been proposed that the absence of the later is associated with better prognosis [4].

Herein, we present a case of primary carcinoid of the kidney in a 31 year old male who had symptoms of lumbar pain and gross hematuria for 2 months with no features of carcinoid syndrome.

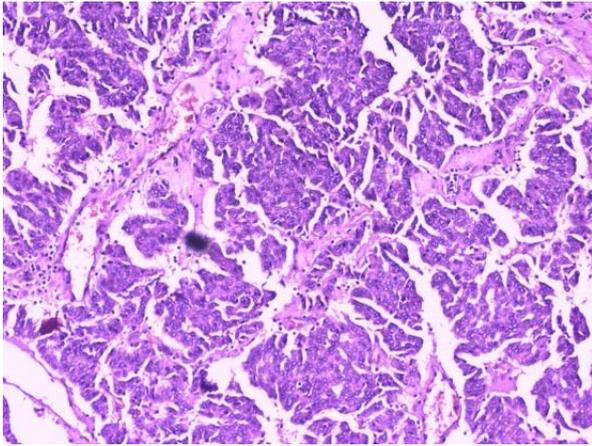
**CASE REPORT**

A 31 year old male presented in the out-patient department with complaints of painless gross hematuria and lumbar pain for 2 months. There was no history of sweating episodes, diarrhoea, flushing, paroxysmal asthma or psychiatric illness. Family history was unremarkable. Physical examination was normal. Blood pressure was 124/82 mm Hg. All the other routine investigations including blood and urine examination, erythrocyte sedimentation rate, serum electrolytes, blood urea nitrogen, serum creatinine levels and endocrine index were within normal limits. Ultrasonography abdomen revealed a 5 × 4.5 cm heterogenous mass in the medial side of the left kidney. Abdominal CT scan revealed a tumor compressing the renal parenchyma and displacing the left pelvicalyceal system without hydronephrosis. It had no extra-renal, ureteric or renal vein extension. No abdominal lymphadenopathy was noted on CT abdomen. Chest X-

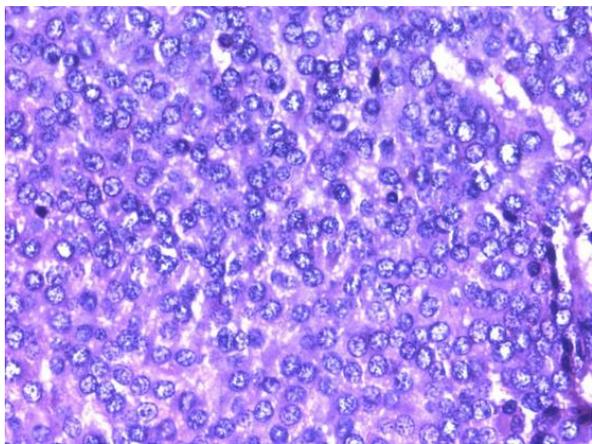
ray revealed normal lung parenchyma, soft tissue and bony structures. Renal cell carcinoma was suspected and the patient underwent left radical nephrectomy and the specimen was sent to the pathology department.

Gross examination of the left radical nephrectomy revealed a 4x3.5cms well-circumscribed grey-yellow firm growth involving the medial part of middle lobe and a part of the lower lobe of the left kidney. (Fig 1) H&E stained sections revealed a well-differentiated neuroendocrine tumor with typical pattern of carcinoid tumor. The tumor cells were arranged in cord-like or thin trabecular pattern having a granular eosinophilic cytoplasm with round to oval nuclei and fine stippled chromatin. Mitosis was rare. (Fig 2, 3) The renal pelvis, calyces and blood vessels were free from tumor. Immuno histo chemistry demonstrated strong cytoplasmic labelling for synaptophysin, and negative reactivity for CD10, hence, confirming the diagnosis. (Fig 4, 5).

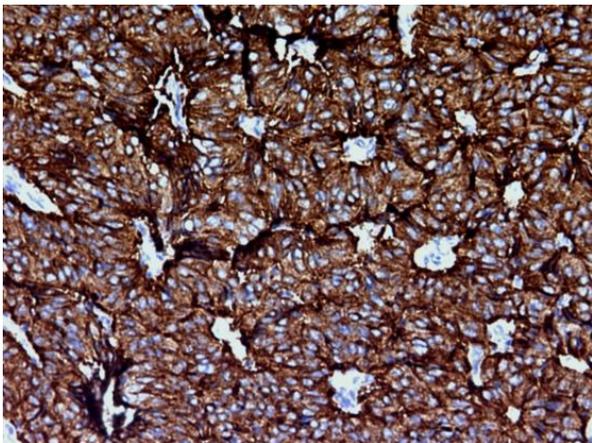
**Fig 1: Gross specimen of Kidney showing medial growth.**



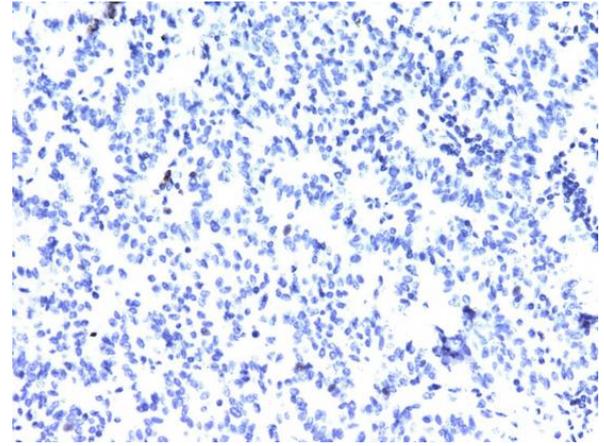
**Fig 2: H&E stained section showing trabecular pattern of tumor. (H&E, 100X)**



**Fig 3: H&E stained section showing 'salt and pepper' chromatin pattern. (H&E, 400X)**



**Fig 4: IHC marker: Strong synaptophysin positivity. (IHC, 200X)**



**Fig 5: IHC marker: CD10 negative. (IHC, 200X)**

## DISCUSSION

The 2010 WHO classification of neuroendocrine neoplasms divided them in following grades: Grade 1 well differentiated NE tumor (carcinoid), Grade 2 well differentiated NE carcinoma (Atypical carcinoid), Grade 3 poorly differentiated (small and large cell type), mixed adenoneuroendocrine carcinoma (MANEC), hyper plastic and pre neoplastic lesions [5].

Most commonly, they occur in gastrointestinal tract followed by lungs. Genitourinary system involvement is unusual with testis and prostate being maximally affected. Kidney involvement is extremely rare due to absence of entero chromaffin cells in renal parenchyma. Few theories suggested that the probable cells of origin in kidneys are from intestinal metaplasia of the pyelocalyceal urothelium, metastases from undiscovered primaries, or entrapped neural crest cells or pancreatic cells [6]. An increased incidence of carcinoid tumor has been seen in horse-shoe kidneys [7].

Romero *et al.*; [8] demonstrated the predilection for right kidney more than the left with equal occurrence in both the sexes. Carcinoid tumor associated with horse-shoe kidney has a male preponderance probably due to the later occurring more often in males than females. According to shurtleff *et al.*; [9] the most common presenting symptom is flank/abdominal pain and hematuria occurring in nearly 56% of the patients. Some patients may present with features of carcinoid syndrome or other constitutional symptoms or may be diagnosed as asymptomatic renal masses.

The radiological features of renal carcinoid are variable and histology forms the definite tool of diagnosis. Mckeown *et al.*; [10] described 10 cases of primary renal carcinoid; out of which five were predominantly solid on imaging, one was a complex cystic tumor, four arose from the teratoid malformation of kidney and one was lacking the radiological details. Romero *et al* collected 56 cases out of which 26%

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showed calcifications on imaging studies. Some authors are of the view that renal carcinoid mimics renal cell carcinoma and other kidney lesions radiologically[6,11]. However, Somatostatin receptor scintigraphy is known to be the most useful tool for finding the mass and determining the stage[9].

Metastases is not much common and if it occurs, hilar lymph nodes followed by liver are the most common sites[8]. Surgery forms the gold standard treatment for renal carcinoid tumor. Radical nephrectomy with adjacent lymph node dissection is the treatment of choice if the tumor is confined to the kidneys. In cases of distant metastasis, chemotherapy may be considered although no definite proven benefits have been yet reported. [11, 12]. Prognosis is relatively optimistic with an average post-operative survival of 40 months in the patients with localised tumor and 27.6 months in patients who had distant metastasis [12].

### CONCLUSION

Primary renal carcinoid tumor is rare and diagnosis is usually made on biopsy due to its tendency to mimic renal cell carcinoma on imaging. Also, a low index of clinical suspicion renders a late diagnosis which is dismal owing to its good prognostic results if diagnosed early. Therefore, it becomes important for the clinician to analyse all the clinical features along with a confirmatory histopathological assessment to provide an early definitive diagnosis.

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