

Well-Differentiated Neuroendocrine of Left Kidney

Neuroendocrine tumours (NETs) of primary renal origin are quite rare neoplasms, and the majority of published literature refers to them as sporadic. Hence, there are no details about the epidemiology and aetiological characteristics about them.¹ Majority of them occur on the right side as reported in the literature.^{2,3} Herein, we present a young female with well-differentiated NET of the left kidney.

A 31-year female presented with intermittent left flank pain and burning micturition for 2 months. CT scan of the abdomen revealed a large circumscribed exophytic mass arising from the upper and interpolar region of the left kidney medially. It showed solid and cystic components within it and measured 4.7 x 4.8 x 7.2 cm in AP, TS, and CC dimensions. Its solid enhancing component measured 4.7 x 4.8 x 4.3 cm in similar dimensions and showed areas of hyperdensity at the periphery likely representing haemorrhage. It was causing anterior displacement of the left renal vein and artery without definite evidence of infiltration. Superiorly, it abutted the spleen and the tail of the pancreas with focal indistinct fat planes. Fat planes with the left adrenal gland appeared intact. Posteromedially, it abutted the left psoas muscle without infiltration (Figure 1). Furthermore, there was no evidence of metastasis. Subsequently, she underwent radical nephrectomy and the histopathology was consistent with well-differentiated NET with a size of 6.5 x 5 x 4 cm limited to the upper pole of the left kidney with lymphovascular invasion.

Immunohistochemically, the tumour cells were positive to CKAE1/AE3, CK LMW, synaptophysin, and CD56, whereas S-100 and chromogranin were focal positive. However, CK7, PAX8, CD10, CD117, inhibin, Mart-1, HMB-45, TTF-1, CDX2, and GATA-3 were negative and Ki-67 was 1-2%. Post-surgery period was uneventful and imaging on surveillance after 3 months revealed no recurrent or metastatic disease.

Well-differentiated NETs of the kidney may have no symptoms on initial presentation. Therefore, these often present during routine examination in almost 50% of patients; however, 20% of patients have abdominal pain and/or low back pain and less than 10% may present with haematuria. Carcinoid syndrome is a rare occurrence and is seen in less than 4% of patients with carcinoid tumours.³

According to Romero *et al.*, renal carcinoid tumours are found more often on the right side (60.9%) than on the left.² This was also consistent with another study.³ Average diameter of well-differentiated NETs of renal origin is almost 5.9 cm. Furthermore, gross examination of well-differentiated NETs of the kidney shows solid and cystic components of variable sizes with components of internal haemorrhage and occasional necrosis in the majority of cases.^{2,3}



Figure 1: CT abdomen scan revealed large circumscribed exophytic mass arising from the upper and midpolar region of the left kidney medially. It shows solid and cystic components within it and measures 4.7 x 4.8 x 7.2 cm in AP, TS, and CC dimensions. Its solid enhancing component measures 4.7 x 4.8 x 4.3 cm in similar dimensions and show areas of hyperdensity at periphery likely representing haemorrhage.

On immunohistochemistry, they stain positive for synaptophysin more than chromogranin, CD56, and neuron-specific enolase (NSE).³ In a study, the sensitivity of synaptophysin was 100%, followed by chromogranin, NSE, CK, and vimentin at 97.2%, 93.5%, 88.2%, and 40%, respectively, in renal carcinoids.² In another study, the sensitivity of synaptophysin was also 100%, and that of chromogranin, NSE, and Vimentin was 92.3%, 100%, and 40%, respectively. In the present case, the surgical specimen was immunohistochemically positive for synaptophysin, NSE, and CD56.^{3,4}

The prognostic value of histology parameters is still not clear, but some studies have demonstrated that mitosis and cytological atypia have a certain prognostic value.⁵ Radical nephrectomy is the mainstay of treatment for localised NETs of the kidney.

COMPETING INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

AHO: Conceived the idea, conducted the literature search, and drafted the manuscript.

AHJ: Conceived the idea, provided overall supervision, and managed the study.

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