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A CASE REPORT- RENAL CELL CARCINOMA-RHABDOID VARIANT

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ABSTRACT

Renal cell carcinoma also known as (Hypernephroma, Grawitz tumor, Renal adenocarcinoma) is a kidney cancer that originates in the lining of the proximal convoluted tubule. Adult clear cell carcinoma with extensive rhabdoid features is a rare clinical entity. Rhabdoid morphology in renal cell carcinoma resembling malignant rhabdoid tumor of kidney was first described in 1991, but this phenomenon should be strictly distinguished from extrarenal malignant rhabdoid tumors because the prognosis and therapeutic strategies differ significantly. Rhabdoid features have been recently identified as a morphological variant of renal cell carcinoma associated with poor prognosis and a marker of high risk for metastasis. Rhabdoid cells are characterized by a large abundant eosinophilic cytoplasm with an irregular eccentric nucleus and a rounded eosinophilic cytoplasmic inclusion. A 45 years old male came with a history of pain in left lumbar and epigastric region since 4-5 months.

INTRODUCTION

RCC is the most common type of kidney cancer in adults, responsible for approximately 90-95% of cases. The 2004 World Health Organization classification for renal neoplasms recognizes several distinct histologic subtypes of RCC. These subtype include clear cell RCC, papillary RCC, chromophobe RCC, hereditary cancer syndromes, multilocular cystic RCC, collecting duct carcinoma, medullary carcinoma, mucinous tubular and spindle cell carcinoma, neuroblastoma-associated RCC, Xp11.2 translocation-TEF3 carcinoma, and unclassified lesions. Rhabdoid features have been recently identified as morphological variant of renal cell carcinoma associated with aggressive behavior and poor prognosis. Rhabdoid cells are characterized by abundant eosinophilic cytoplasm with an irregular eccentric nucleus and round eosinophilic cytoplasmic inclusion.

Case Report

A male Patient 45 years old came to MGM Hospital, Kamothe OPD with history of left lumbar pain 4-5 months radiating to back and epigastric pain which was off sudden onset and progressive in nature. It was associated with on and off fever since 4-5 months. USG Abdomen-Left side enlarged bulky kidney with diffuse increased cortical echotexture of size - 97x99x109 mm echocystic mass present in left renal area arising from mid pole of left kidney suggestive of RCC?? Abscess. CT Scan for abdomen and pelvis suggested large necrotic exophytic mass involving left renal mid polar and part of upper polar region with zorotous fascia involvement with upper and mid calyceal cupping with cortical obstruction and calcification without renal vein or artery involvement without thrombosis, suggestive of left renal cell carcinoma involving zorotous fascia with upper and mid calycealcupping.

Gross

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Microscopic Examination

H&E stained section studied shows tumor cells arranged in monolayered sheets, tubular, acinar and pseudopapillary pattern. Individual tumor cells are highly pleomorphic with abundant clear to granular eosinophilic cytoplasm (rhabdoid cells), hyperchromatic nuclei, irregular nuclear membrane and few prominent nucleoli. Some of the tumor cells are bizarre and highly anaplastic. Also seen are tumor giant cells and presence of large areas of necrosis and hemorrhage. Tumor emboli are also noted. Adrenal gland showed infiltration with tumor.

DISCUSSION

Renal cell carcinoma with rhabdoid features is an uncommon and highly aggressive malignancy. Although only few cases of adult pure rhabdoid renal tumors are reported, Gokden and colleagues found that 5% of RCC exhibited rhabdoid features. Despite the relative rarity of rhabdoid features, they are universally indicators of aggressive neoplasm. In the kidney, RCCs with rhabdoid features are considered as high grade and high stage tumors with frequent extra renal extensions, widespread distant metastasis.

CONCLUSION

RCC with rhabdoid component is an uncommon and highly aggressive malignancy often found in paediatric age group. Few cases are reported in adults.

Rhabdoid features are an indicator of aggressiveness in the tumour with high risk of distant metastasis to lung, bone, liver, soft tissue, skin and adrenal upto 70% of cases and 20% metastasis to lymph nodes. Hence pathologists should carefully search for a rhabdoid component in RCC, and when present it should be reported because it is an indicator of dedifferentiation and is related to sarcomatoid transformation and very poor prognosis.

References


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