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Renal cell carcinoma with rhabdoid features: A rare aggressive and fatal variant

Abdikarim Hussein Mohamed *, Hussein Ali Mohamud

Department of Urology, Mogadishu Somali Turkish Training and Research Hospital, Mogadishu, Somalia

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ABSTRACT

Renal cell carcinoma with rhabdoid features is a rare histopathologic variant recently documented. It is a very aggressive tumor and associated with a higher mortality rate and poor prognosis. A 22 years old female patient presents with a rare case of clear cell renal cell carcinoma with rhabdoid features successfully managed with right radical nephrectomy and paracaval lymph node excision. The patient was alive, healthy and three years of flow up for the patient was free from metastasis. Despite rhabdoid features are lethal tumors and are associated with higher grades, radical nephrectomy with lymph node dissection increases survival rate.

Introduction

Clear cell renal cell carcinoma with rhabdoid features is a rare histopathologic variant of renal cell carcinoma (RCC) recently recognized. It is a very aggressive tumor and associated with a higher mortality rate and poor prognosis. Rhabdoid features are associated predominantly to clear RCC but may be rarely associated with the papillary type and collecting duct type. Rhabdoid tumors of the kidney are common in childhood (2% of pediatric tumors) and rare variant types of renal carcinoma in adults. About 3.2–7.4% of renal cell carcinoma has rhabdoid features [1].

There are fewer reports in the literature regarding this newly discovered histopathologic variant of renal cell carcinoma. We report a rare case in 22 years old female patient with clear cell renal cell carcinoma with rhabdoid features successfully managed with right radical nephrectomy and paracaval lymph node excision.

Case report

22 years old female ill patient presented with right flank pain and mass in the right side of the abdomen. Laboratory investigations showed normal parameters. Contrast-enhanced computed tomography of the thoraco-abdomen revealed a 10 cm sized mass lesion with necrotic changes and milimetrical calcifications in the right kidney (Fig. 1). There was no evidence of tumor thrombus in the renal vein and inferior vena cava as well as no evidence of metastasis to lung and liver. The patient has undergone right radical nephrectomy with paracaval lymph node excision. Pathology was reported large nucleus, abundant eosinophilic cytoplasm with a rounded eosinophilic cytoplasmic inclusion suggesting a variant of clear cell renal cell carcinoma with rhabdoid features grade IV (Fig. 2). The surgical margin of the renal vein and ureter were free of tumor. Gerota's fascia was intact, sinus hyperplasia was seen in two (2/20) lymph nodes and distant metastasis was not seen. The postoperative period was uneventful and 3 years follow up for the patient was alive and healthy.

Discussion

Rhabdoid features have linked to aggressive behavior types of tumors in general. Xavier Leroy Et al reported 14 cases of clear cell RCC with rhabdoid features, 43% of the cases died with 2 years [2]. 4/8 patients died within one year from the Kuroiwa K study. These studies are associated with higher mortality rates, radical nephrectomy with lymph node dissection may prevent reservoir of tumors cells and increases the survival rate. Clear cell renal carcinoma with rhabdoid features is associated with early metastasis (70% of the cases have distant metastasis at the time of diagnosis.) predominantly to the lungs and poor prognosis despite the size of the tumor and early intervention. Mortality from rhabdoid features (up to 50% and life expectancy reach 8 months in some studies.) may be associated with rapid cell proliferation. In the reporting case, the patient was alive, healthy and distant metastasis was not seen for three years follow up for patient.

A study from Jennifer R et al. noted that the dedifferentiation of renal cell carcinoma is related to rhabdoid features and differs from

* Corresponding author.
E-mail address: abdikarimgabeyre@gmail.com (A.H. Mohamed).

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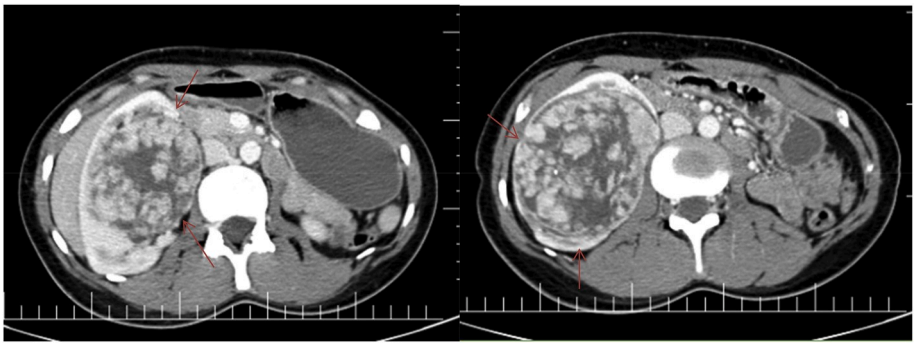

Fig. 1. Contrast-enhanced computed tomography of the thoraco-abdomen revealed a 10 cm sized mass lesion with necrotic changes and milimetrical calcifications in the right kidney (arrows).

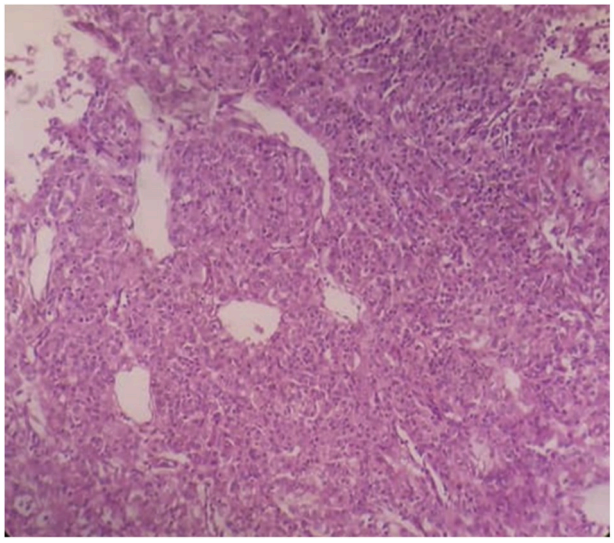

Fig. 2. Histopathologic features of clear cell renal carcinoma with rhabdoid.

sarcomatoid renal cell carcinoma due to lacking histopathologic features of sarcomatoid changes [3]. Rhabdoid features are associated with overexpression of P53 in which primary renal cell carcinoma is rarely mutated [2].

Rhabdoid features have linked to higher grades and stages of renal cell carcinoma as the current presenting case of clear cell renal cell carcinoma with rhabdoid features grade IV. Renal cell carcinoma with rhabdoid features is considered grade 4 by the International Society of Urological Pathology (ISUP) and recommended to report due to higher rates of metastasis, its aggressive pattern and related poor prognosis [4].

Renal cell carcinoma with a rhabdoid feature is managed mainly with radical nephrectomy and lymph node dissection as in our case. Anil Kapoor et al. and De Vincenzo F et al. reported treatment of tyrosine kinase inhibitors (sorafenib, sunitinib) for renal cell carcinoma with rhabdoid features respectively.

Conclusion

We conclude that rhabdoid features are lethal tumors and are associated with higher grades and very poor prognosis. It is managed mainly with radical nephrectomy and lymph node dissection.

Declaration of competing interest

The authors declare no conflict of interest and this study received no financial support.

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