

## RHABDOID VARIANT OF CLEAR CELL RENAL CELL CARCINOMA- A CASE REPORT

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### INTRODUCTION

Renal cell carcinoma (RCC) accounts for 2% of all malignancies and over 90% of all renal malignancies. It is a tumour of adults with an average age at diagnosis of 55-60 years and a male to female ratio of 2:1. Of the various histological subtypes, clear cell type is the most common<sup>1</sup>. Rhabdoid tumour is a malignant tumour of the kidney which occurs typically in children, however only a few cases of pure rhabdoid tumours have been reported in adults<sup>2</sup>. Rhabdoid variant of clear cell renal cell carcinoma also known as adult clear cell renal cell carcinoma with rhabdoid features is an uncommon tumor in adults. Literature shows that about 5% of the renal cell carcinomas exhibit rhabdoid features. Moreover, it is a high grade neoplasm with aggressive behaviour and a dismal outcome<sup>3</sup>.

### CASE REPORT

A 62 years old male presented to the Armed Forces Institute of Urology with twenty days history of aching pain in both flanks, more marked on right side. It was associated with two episodes of haematuria. Upon investigation, his Renal Function Tests were within normal limits. Ultrasound abdomen and KUB (kidney, urinary bladder) revealed right renal mass while CT Scan showed a 12x10 cm mass in right kidney with extension into the renal vein. A provisional diagnosis of Stage T3 renal cell carcinoma was made. Right radical nephrectomy was performed and the specimen was received at Department of Histopathology, Armed Forces Institute of Pathology for histopathological examination.

On gross examination, the kidney measured 11x9x6.5 cm. The cut surface showed

a firm, gray white tumour measuring 5.5x5x4.5 cm, located in the middle and lower pole of the kidney. It was grossly involving the renal capsule. The renal vein was not grossly involved by the tumour (Fig-1).

Multiple sections were prepared from the tumour specimen. On light microscopy, the sections from the tumour revealed predominantly a rhabdoid component making



Figure-1: Gross appearance of rhabdoid variant of clear cell renal cell carcinoma.

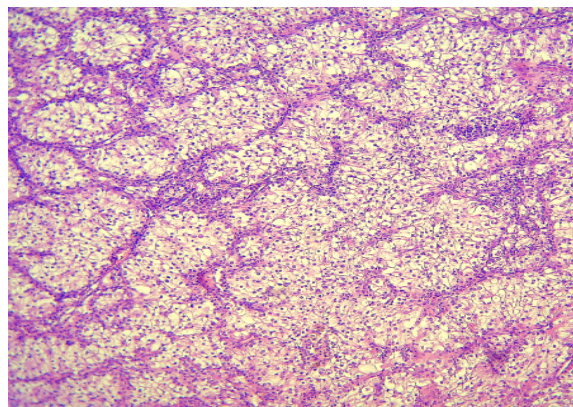


Figure-2: Photomicrograph of rhabdoid variant of clear cell renal cell carcinoma. Clear cell (Conventional) renal cell carcinoma component, showing cells having clear cytoplasm and Grade III nuclei (Hematoxylin-Eosin original magnification 10x).

upto 80% of the tumour while 20% of the tumour was composed of clear cell component. The clear cells with Nuclear Furhmann grade III nuclei, separated by arborizing capillaries,

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corresponded to clear cell (Conventional) renal cell carcinoma (Fig-2). The rhabdoid component was composed of sheets of epithelioid and polygonal cells having abundant eosinophilic cytoplasm with large intracytoplasmic hyaline globules, high grade eccentric nuclei with prominent nucleoli. (Fig-3a). Tumour necrosis was also present. The tumour was found to involve the renal pelvis, renal capsule and perinephric fat. The renal vein and ureter resection margin were not involved by the tumour.

Immunohistochemistry was applied on the paraffin sections. The rhabdoid component was positive for vimentin (Fig-3b), Neuron Specific Enolase, Pan Cytokeratin and negative for desmin and Cytokeratin<sup>4</sup>. The clear cell component was positive for vimentin and cytokeratin<sup>5</sup>. Therefore, the diagnosis of rhabdoid variant of clear cell renal cell carcinoma was made.

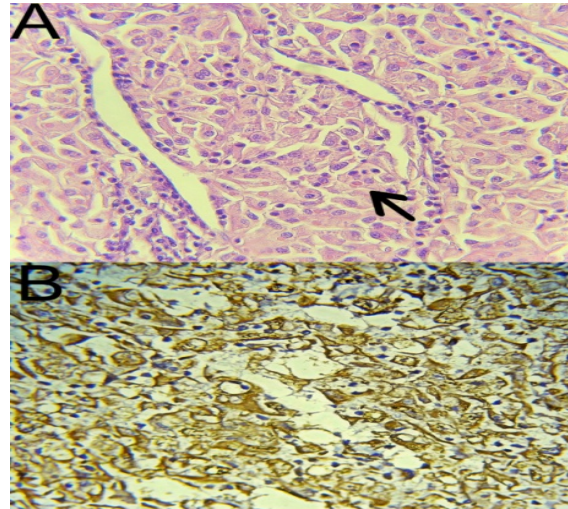
Upon follow up, the symptoms of our patient were found to have improved after the surgery. Staging investigations including CT scan chest and bone scan were carried out and they were negative for any metastasis.

## DISCUSSION

The term 'rhabdoid' refers to tumour cells that resemble rhabdomyoblasts morphologically but differ immunohistochemically and ultrastructurally<sup>6</sup>. Rhabdoid tumour was first described in kidney as a distinct entity in children in 1978 by Beckwith and Palmer as 'rhabdomyosarcomatoid variant of Wilm's tumor'<sup>7</sup>. Extrarenal sites of rhabdoid tumour include skin, liver, pancreas, stomach, intestine, urinary bladder, prostate, central nervous system, orbit, lacrimal gland and female genital tract<sup>8</sup>.

Rhabdoid tumours in the kidney are primarily found in children<sup>4</sup>. Pure renal rhabdoid tumours are reported rarely in adults. They usually present as composite neoplasms; occurring in transition with conventional clear cell renal cell carcinoma<sup>9</sup>. The median age of presentation is 62 years, with a male predominance<sup>10</sup>. It most commonly presents as

an abdominal mass, and occasionally with haematuria and flank pain. Metastases occur in 80% of the patients, mainly affecting lungs, liver and brain<sup>5</sup>. Grossly, the rhabdoid component of



**Figure-3. Photomicrograph of rhabdoid variant of clear cell renal cell carcinoma. Rhabdoid component, showing polygonal to epithelioid cells having eccentric nuclei and paranuclear hyaline globules (←) (Hematoxylin-Eosin original magnification 40x[A] and Vimentin 10x[B]).**

the rhabdoid variant of clear cell renal cell carcinoma appears as firm and white area within a yellow necrotic tumour<sup>11</sup>. Microscopically, rhabdoid cells make up 5-90% of tumour cells. The rhabdoid foci are composed of sheets and clusters of variably cohesive, large, round to polygonal to epithelioid cells having vesicular often eccentric nuclei, with high nuclear grade, prominent nucleoli and paranuclear, intracytoplasmic hyaline inclusions<sup>11</sup>. Ultrastructurally, rhabdoid cells have paranuclear aggregates of intermediate filaments or condensation of organelles, often along with peripheral vacuolization<sup>8</sup>. Immunohistochemically, there is positivity for vimentin (100%), NSE (79%), and Pan CK (56%). There is negativity for CK7, CK 20, SMA and desmin<sup>9</sup>.

Like rhabdoid tumours elsewhere, it behaves in a very aggressive fashion. The preferred treatment is radical nephrectomy followed by chemotherapy. The role of tyrosine

kinase inhibitors is also being investigated. Whether alone or in combination, rhabdoid features indicate a poor prognosis. These tumours are more likely to present at higher grades, exhibit extrarenal invasion, recurrence, distant metastasis and a median patient survival of about 8 months<sup>12</sup>. The mortality ranges from 40-50%<sup>10</sup>.

### CONCLUSION

In summary, we conclude that rhabdoid features, even if they are limited, are markers of poor prognosis. Therefore, renal cell carcinoma with rhabdoid features should be considered as a clinically important form of RCC and a pathologist should carefully search and report such features when present.

### CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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