Case Report:

A rare case of chromophobe renal cell carcinoma with focal sarcomatoid differentiation.

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Abstract:

Chromophobe renal cell carcinoma (CRCC) is a variant of renal cell carcinoma having relatively good prognosis. Sarcomatoid differentiation is found in 9% of CRCC and its presence is associated with poor prognosis. We are reporting it because of its rarity and prognostic importance.

KEY WORDS: Sarcomatoid differentiation, Chromophobe renal cell carcinoma

INTRODUCTION:

Chromophobe Renal Cell Carcinoma is the third most common histological subtype of Renal Cell Carcinoma (RCC), accounting for less than 5% of RCCs.[(i)] It originates from intercalated cells of Cortical collecting duct.[(ii)] Men and women of age around 6th decade are the commonest patient population.[(iii)] The tumour is usually unilateral. Most patients are symptomatic at diagnosis; abdominal pain and haematuria are commonly observed. [(2)] The tumour is generally non-aggressive with very good prognosis. Sarcomatoid differentiation of Chromophobe RCC is a rare finding and is characterized by a relatively high incidence of metastases to the lung and bone with worst prognosis.[(iv)]

CASE SUMMARY:

A 56 year old male presented with the history of intermittent abdominal pain and haematuria since one week. Physical examination revealed firm, slightly tender palpable lump in the left lumbar region that did not move with respiration. Contrast- enhanced Computed Tomography and the ultrasound revealed a well defined, heterogeneously-enhancing mass, measuring 21.2x11.3x10.7 cm in the left kidney.

We received left sided nephrectomy specimen measuring 22 x15x10 cm; external surface bosselated. Cut surface of the kidney showed a large circumscribed gray tan tumour mass measuring 20x12x12cm. The tumour is variegated with solid areas, necrosis, haemorrhage and cystic changes (Fig. 1). Normal kidney identified at the lower pole. A part of ureter identified 3cm in length. Ureteric and vascular surgical margin were unremarkable. We also received fibrofatty tissue measuring 7x4x3cm with 18 lymph nodes ranging in size from 0.5 cm to 2 cm. Histological examination showed kidney with a well demarcated tumour composed of lobules separated by fibrous septae (Fig. 2). Lobules were composed of solidly packed, round to polygonal cells

with well-defined cell borders and eosinophilic cytoplasm. Nuclei were round with regular nuclear membrane and Perinuclear halo was seen (Furhman grade I-II). Few small foci of pleomorphic, bizarre spindle cell were seen, arranged in fascicles and diffuse sheets (Fig 3). Large areas of necrosis and haemorrhage were seen. Sclerosis and calcification were noted (Fig 4). Tumour had invaded the capsule. Pelvicalacyal system was also involved. However, part of distal pelvis and ureter were free of tumour. Vascular/lymphatic/perineural invasion were not seen. Gerota's fascia was free of tumour. There was no evidence of renal vein metastasis. Surrounding kidney showed secondary pyonephrosis. 18 lymph nodes were dissected from the lymph node mass. All showed only congestion and reactive hyperplasia. No metastasis detected.

DISCUSSION:

Renal cell carcinoma being the most common neoplasm of the kidney, has been classified by WHO (2004) into various subtypes. These are: clear cell RCC (70%), papillary RCC (10-15%), chromophobe RCC (4-6%), collecting duct carcinoma (about 1%) and unclassified RCC (4-5%).[v] Distinct characteristic of chromophobe variant were first described by Theones et al in 1985.[vi] There is no gender preponderance and it is seen mostly around sixth decade.[vii] It is non aggressive having only 6-7% incidence of metastasis.[viii] Gross appearance is generally well circumscribed, solitary tumour with homogenous grey tan cut surface.[ix] CRCC has been classified into various subtypes as: Type I (eosinophilic variant) - Small cells with granular, eosinophilic cytoplasm. Type II (mixed variant) - larger cells with a peri-nuclear halo. Type III (classical variant) - Thick, well defined borders, 'raisinoid' nuclei and abundant, pale, granular cytoplasm.[x] In this case, the tumour was composed of, round to polygonal cells with well-defined cell borders and round nuclei with perinuclear halos. So, a diagnosis of Mixed (Type II) chromophobe renal cell carcinoma was made. Differentiation between eosinophilic variant of CRCC and Oncocytoma can be a diagnostic dilemma. Eosinophilic variant shows tumour cells in sheets with reticulated cytoplasm, irregular cleaved nuclei with perinuclear halo while Oncocytoma has nested architecture with densely granular cytoplasm and round nuclei.[8] In CRCC, Hale's colloidal Iron will show a diffuse cytoplasmic staining while Oncocytoma shows focal positive staining. Immunohistochemically, CRCCs are positive for CK-7, CD-117 and epithelial membrane antigen.[xi]

According to study done by Parada et al, sarcomatoid differentiation in CRCC has been reported worldwide in only 16 cases; signifying its rare occurrence.[xii] Immunohistochemical and ultrastructural studies on the sarcomatoid component have demonstrated that the sarcomatoid portion is derived from metaplastic transformation of carcinoma.[xiii] This tumour has highly malignant biological behavior.[4] Sarcomatoid renal cell carcinomas with zero or minimal necrosis yield a favorable prognosis, whereas those with moderate or massive necrosis yield a significantly poorer survival time.[xiv] In our case the tumour showed foci of sarcomatous change with extensive necrosis and calcification indicating poor prognosis.

CONCLUSION:

We conclude that Sarcomatoid differentiation of CRCC adversely affects the prognosis, hence screening for its presence with extensive sampling is necessary for proper diagnosis.

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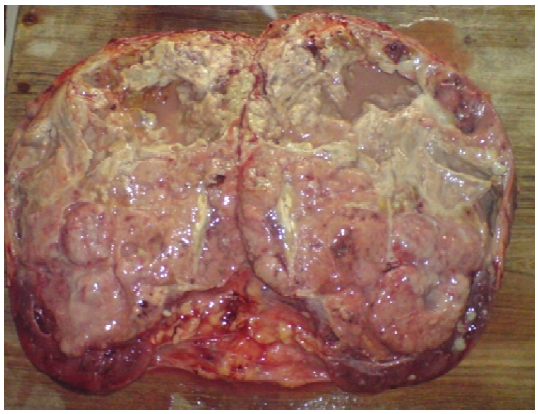
1 Gross specimen shows gray tan variegated tumour mass with areas of necrosis, haemorrhage and cystic change.

2 Microphotograph of tumour cells with eosinophilic cytoplasm and perinuclear halos (H & E, 100X)

3 Microphotograph of spindle shaped cells in the foci of sarcomatous differentiation (H&E, 400X)

4 Microphotograph showing calcification in the foci of sarcomatous differentiation (H&E,)



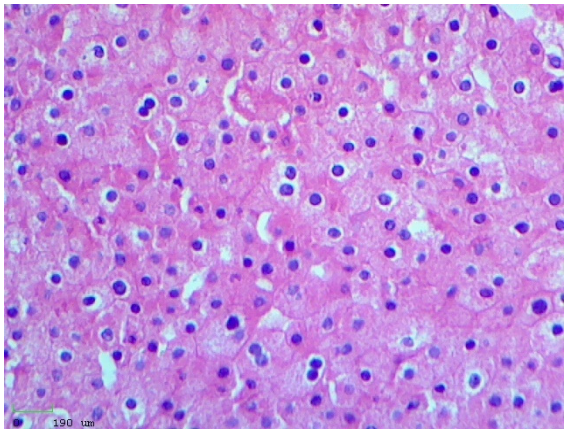
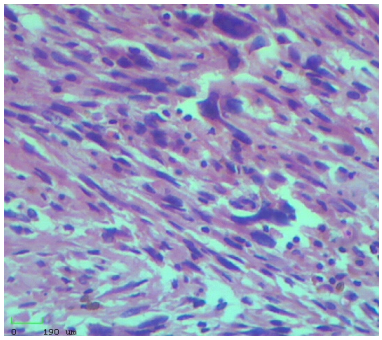


Figure 1: Gross specimen shows gray tan variegated tumour mass with areas of necrosis, haemorrhage and cystic change.

Figure 2: Microphotograph of tumour cells with eosinophilic cytoplasm and perinuclear halos (H & E 100X)



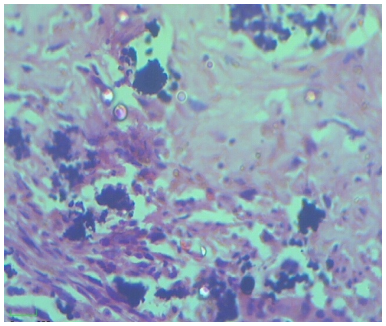


Figure 3: Microphotograph of spindle shaped cells in the foci of sarcomatous differentiation (H&E, 400X)

Figure 4: Microphotograph showing calcification in the foci of sarcomatous differentiation (H&E, 400X)

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