Sarcomatoid Renal Cell Carcinoma: Case Report and Review of Literature

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ABSTRACT
Sarcomatoid renal cell carcinoma is a tumour with aggressive behaviour. It is a form of dedifferentiated carcinoma with high incidence of metastases to the lungs. We report a case of 55-year-old male who presented with complaints of loin pain and hematuria. The biopsy confirmed sarcomatoid variant of renal cell carcinoma.

CASE REPORT
A 55-year-old male presented with history of loin pain and hematuria. The Computerized Tomography (CT scan) showed a renal mass [Table/Fig-1a] and even bilateral lung metastasis. Laparotomy was performed and the nephrectomy specimen was sent for histopathology.

GROSS EXAMINATION
A radical nephrectomy specimen [Table/Fig-1b] consisted of right kidney surrounded by perinephric adipose tissue and Gerota fascia (300g), measuring 10.5 X 8 X 7cms. The ureter is dilated and measuring 3 cms in length. A 6 X 5.5 X 4.5 cms poorly defined, variegated mass occupied the centre and lower pole with areas of haemorrhage and necrosis. The renal pelvis is dilated. A defect is identified in the renal capsule near the hilus measuring 2 X 1 cms. The surrounding renal parenchyma is thinned out at the lower pole. The renal vein and adrenal gland is normal and free of tumour.

MICROSCOPIC EXAMINATION
The sections of kidney show a malignant tumour composed predominately of sheets of spindle cells with pleomorphic nuclei and prominent nucleoli [Table/Fig-2a]. This indicates sarcomatoid differentiation. Focally the tumour cells have a polygonal shape with granular eosinophilic cytoplasm and pleomorphic nuclei [Table/Fig-2b]. The Fuhrman nuclear grade was G4. In addition, bizarre tumour giant cells are seen along with extensive areas of necrosis and dense chronic inflammatory infiltrate [Table/Fig-2c]. The tumour is seen extending into the perirenal adipose tissue near the renal pelvis [Table/Fig-2d] and also into the adjacent renal parenchyma along with dilated tubules, few showing luminal cast and chronic inflammatory infiltrate. The ureter, renal artery and adrenal gland are free of tumour.

IMAGING
CT scan showed a heterogenous, exophytic, necrotic mass (5.7 x 6.1 x 5.5 cms) in the middle 1/3rd of the non - excreting right kidney extending into the pelvis and causing lower calyceal dilatation. The visualized lung bases showed multiple rounded lesions suggestive of multiple lung metastases.

Keywords: Laparotomy, Sarcomatoid carcinoma

[Table/Fig-1a]: CT scan shows a heterogenous, exophytic, necrotic mass in the mid 1/3rd of the non - excreting right kidney extending into the pelvis and causing lower calyceal dilatation
[Table/Fig-1b]: Cut section shows a variegated mass occupying the centre and lower pole with areas of haemorrhage and necrosis

[Table/Fig-2a]: Section shows sheets of spindle cells in whorled pattern (H&E 10X)
[Table/Fig-2b]: Section shows large polygonal cells with eccentric nuclei and abundant eosinophilic cytoplasm (H&E 10X)
[Table/Fig-2c]: Section shows extensive areas of necrosis and chronic inflammatory infiltrate (H&E 10X)
[Table/Fig-2d]: Section show tumor extending into the surrounding renal parenchyma (H&E 10X)
IMMUNOHISTOCHEMISTRY
MIB-1 immunohistochemical stain was done for confirmation which showed strong immunoreactivity [Table/Fig-3a,b].

DISCUSSION
Renal cell carcinoma (RCC) is the most common malignancy of the kidney and consists of multiple subtypes [1]. RCC with sarcomatoid differentiation is a tumour with aggressive behaviour [2]. They have a high incidence of metastases to the lung and bone at presentation [3]. On immunohistochemistry they are positive for MIB-1, AE1/AE3, vimentin, EMA which supports epithelial origin [4]. Sarcomatoid RCC was initially felt to represent a primary renal sarcoma, however, now it is considered a form of dedifferentiated carcinoma and is therefore not a distinct histologic entity. It is defined as any subtype containing foci of pleomorphic spindle cells and is seen in high grade RCC, at the end stages of disease progression. Sarcomatoid differentiation is reported to occur in approximately 1-8 % of RCC’s. Heterologous sarcomatoid transformation has been reported in other histologic subtypes of sarcomatoid RCC, and conventional RCC is the main type of tumour to undergo heterologous transformation [5]. On microscopic examination, two main histologic subtypes of sarcomatoid component have been described. A fibrosarcoma like appearance and malignant fibrous histiocytoma like pattern. Some tumours’ can represent as undifferentiated sarcoma. In our case it had a fibrosarcoma like appearance. Distant metastases were most frequent to the lungs as present in our case [6].

Investigation into the imaging features of sarcomatoid RCC has been limited. The current trend is to report any sarcomatoid component seen at pathologic analysis, regardless of what percentage of the total lesion comprises, as treatment and follow up protocols may be affected. Therefore, it would be beneficial if the presence of sarcomatoid dedifferentiation could be suggested based on imaging characteristics, potentially with MRI. On MRI, sarcomatoid RCC have an irregular or infiltrative morphology and demonstrate heterogeneous T2 signal intensity and enhancement. Internal necrosis and evidence of aggressive local or distant behaviour was frequently observed [7].

Sarcomatoid chromophobe RCC frequently have multiple gains (polysomy) of chromosomes 1, 2, 6, 10 and 17 [8,9]. Mutations of p53 tumour suppressor gene and frequent complex desmosomal junctions confirm the epithelial nature of the neoplasm [10].

REFERENCES