Metastatic Sarcomatoid differentiation of Papillary Renal Cell Carcinoma – A Rare Case

Dr. Pankaj Pande[1], Dr. Mahesh Kumar U[2], Dr. Yelikar BR[3], Dr. Kumar Sharad Sinha[4]

Abstract:

The aim of the article is to present a rare case of metastatic sarcomatoid differentiation of papillary renal cell carcinoma of left kidney in a 50 year old lady who presented with gross haematuria and left upper quadrant abdominal pain. Sarcomatoid differentiation in renal cell carcinoma (RCC) is a growth pattern characterized by malignant spindle-shaped cell histology, which arises in approximately 5% cases of Papillary R.C.C. It has a very aggressive disease pattern and in our case the patient succumbed to the disease within a month of diagnosis.

Key Words: Renal cell carcinoma, Papillary, Sarcomatoid.

[1] Associate Professor, [2] Assistant Professor, [3] Professor & Head, [4] Post Graduate resident

BLDE University's , Department of Pathology, Shri BM Patil Medical College, Bijapur, Karnataka

Corresponding author email: maheshdearmedico@yahoo.co.in

INTRODUCTION:

Sarcomatoid differentiation in renal cell carcinoma (RCC) is a growth pattern characterized by malignant spindle-shaped cell histology, which arises in approximately 5% cases of Papillary R.C.C. It is not a distinct histologic entity; rather, it can be observed across all RCC subtypes, including clear-cell, papillary, chromophobe, unclassified, and collecting duct carcinomas.[1]

Sarcomatoid tumors are characterized by a relatively high incidence of metastases to the lung and bone at presentation. Sarcomatoid differentiation is thought to represent transformation of the RCC malignancy to a higher grade, therefore Fuhrman grade 4 by definition. [1,2]

CASE PRESENTATION:

A 50 yr old female presented with gross haematuria and left upper quadrant abdominal pain. On general physical examination she appeared normal; there was no evidence of splenomegaly and lymphadenopathy. A pelvic computerized tomography (CT) scan showed a large heterogenous mass measuring 7x5 cm without any calcification at the upper pole of the left kidney. Routine hematological, biochemical and radiological investigations were within normal limits. Later she underwent left sided radical nephrectomy.

PATHOLOGICAL DISCUSSION:

Macroscopy:

Received, left sided radical nephrectomy specimen measuring 11x6x4 cms. External surface was bosselated, glistening without any breach in the capsule. Cut surface showed solid tumor tissue on upper pole which was light brown to grey colour. It was friable, soft to firm in consistency. Papillary fronds were seen grossly. Along the nephrectomy specimen we also received fibrofatty tissue measuring 8x6x4cms in length in which two lymph nodes were dissected.

Microscopy:

Multiple sections studied from the tumour tissue from the left kidney showed features of Papillary RCC. No psammoma bodies were seen. Sections from both the lymph nodes showed metastatic renal carcinoma cell deposits with sarcomatoid differentiation. On immunohistochemical

analysis tumour cells in lymph node tissue were positive for CD 10.

So based on histopathological and immunohistochemical analysis (IHC) a diagnosis of metastatic sarcomatoid differentiation of papillary renal cell carcinoma was confirmed.

Discussion:

Sarcomatoid dedifferentiation is seen in approximately 5% Papillary renal cell carcinoma and has been associated with both its type 1 and type 2 variants.[1] It is not a distinct histologic entity; rather, it can be observed across all RCC subtypes, including clear-cell, papillary, chromophobe, unclassified, and collecting duct carcinomas.[2]

Mutations of p53 may be associated with sarcomatoid differentiation.[3] With regard to immunohistochemical markers; these tumors are generally positive for AE1/AE3, epithelial membrane antigen, CD 10 and vimentin which support an epithelial origin.[4]

Staining for actin, desmin, and S-100 are usually negative. Vascular endothelial growth factor (VEGF), Kit, and S6 kinase have been expressed in the majority of sarcomatoid specimens.[5]

Patients who have metastatic sarcomatoid RCC have a poor prognosis and have a median overall survival (OS) of 3 to 10 months from the time of diagnosis. Patients who have localized disease have 2-year and 5-year survival rates of only 25% to 40%.[6]

Our patient has succumbed to the disease process within one month of diagnosis.

Conclusion:

Metastatic sarcomatoid differentiation of papillary renal cell carcinoma is an aggressive disease that is associated with poor outcome. So, early

diagnosis and treatment is essential in reducing the morbidity and mortality.

References:

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Conflicting Interest : Nil

Source of financial support: Nil

Figure 1: Gross photograph with cut surface showing solid tumor tissue on upper pole of left kidney.

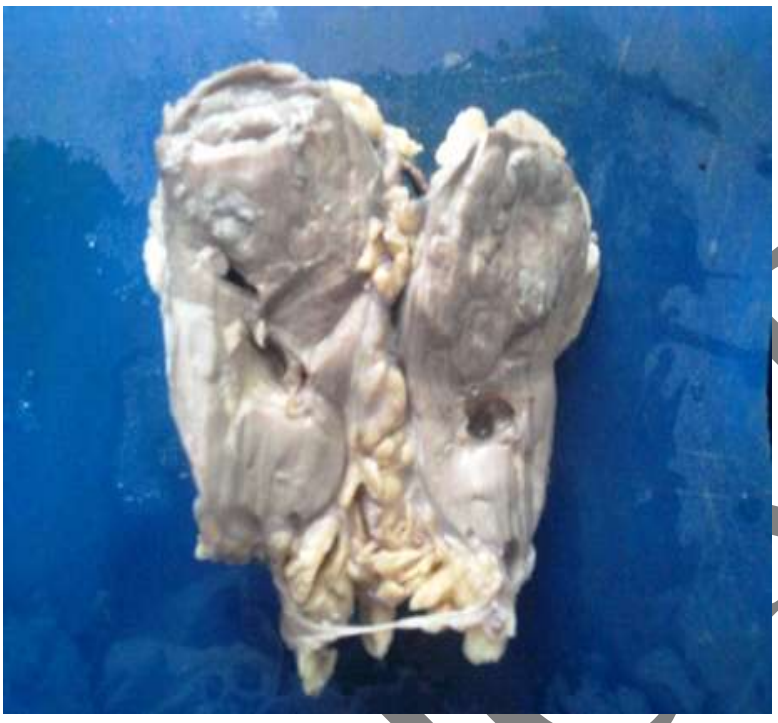

Figure 2: Sections from left kidney shows tumor tissue arranged in papillary pattern (Papillary RCC). H&E stain, 40X.

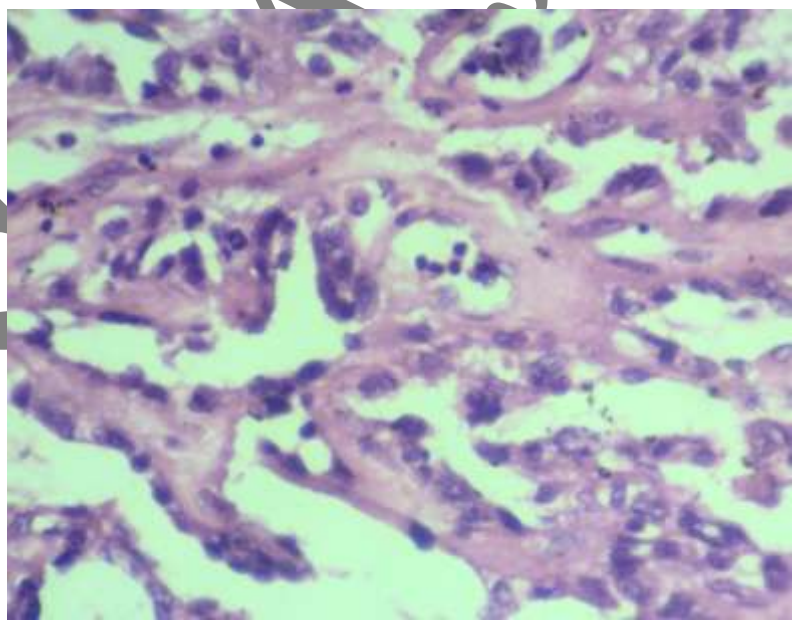

Figure 3: Photomicrograph showing effaced architecture of lymph node tissue with tumour tissue. H&E stain 10X.

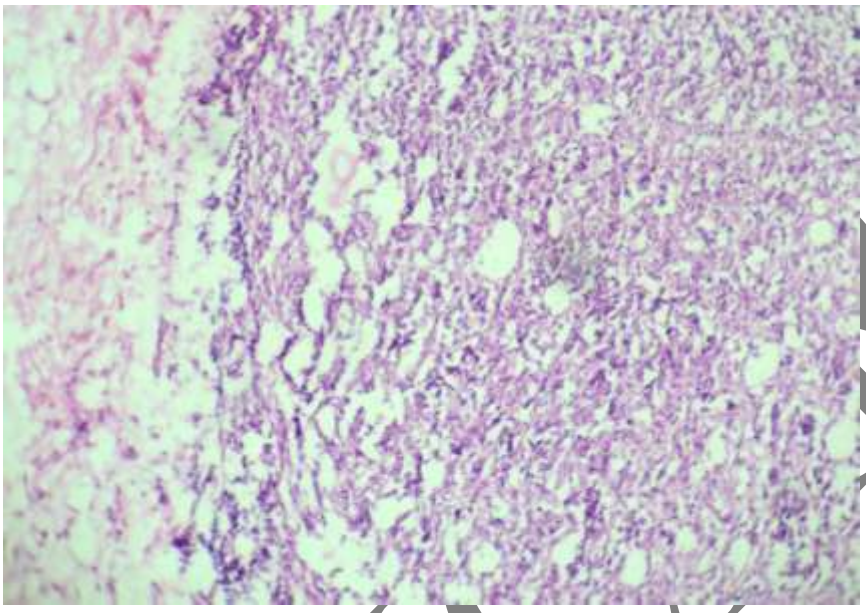

Figure 4- Photomicrograph showing Sarcomatoid differentiation of renal cell carcinoma in lymph node. H&E, 20X

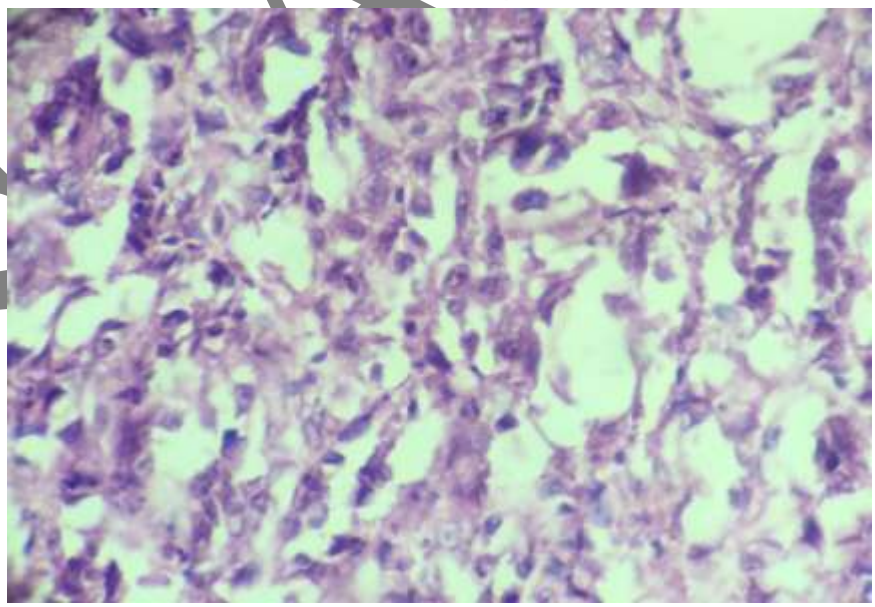

Figure 5: Photomicrograph showing tumour cells positive for CD 10 (IHC).