

Sarcomatoid Renal Cell Carcinoma With Distant Metastasis- An Incidental Autopsy Finding

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Abstract: Sarcomatoid Renal Cell Carcinoma is an aggressive variant of renal cell carcinoma. It is not a distinct histologic entity and represents high grade transformation in different subtypes of renal cell carcinoma. It is a rare entity constituting about 1-8% of all renal malignant neoplasms and is more commonly associated with conventional (clear cell) carcinoma. It is not known whether any particular histologic type has a predilection for sarcomatoid change or whether the primary histologic type of renal carcinoma undergoing sarcomatoid change affects prognosis. We report a case of an incidental autopsy finding of Sarcomatoid renal cell carcinoma with distant metastasis in a 75 yrs old unknown male patient. Immunohistochemistry confirmed the Sarcomatoid variant of Renal Cell Carcinoma.

Keywords: Sarcomatoid Renal Cell Carcinoma

I. Introduction

Sarcomatoid Renal Cell Carcinoma is defined in the 2004 World Health Organization (WHO) classification of renal tumors as any histologic type of renal cell carcinoma containing foci of high grade malignant spindle cell.^[1] Sarcomatoid tumors are characterised by a relatively high incidence of metastasis to the lung at presentation.^[2] Sarcomatoid carcinomas signify a poor prognosis.^[3] The prognostic implication of the proportion of sarcomatoid component within an renal cell carcinoma is an area of controversy. A higher proportion of sarcomatoid differentiation has been associated with worse survival in some series.^[4]

II. Case Report

A 75 yrs old unknown male brought unconscious to hospital and died within one hour. The body was sent for postmortem examination and organs were sent for Histopathological examination.

III. Gross Examination

We received left kidney with attached tumor mass of total size 20x12x7 cms and weight 500 gms . Poorly defined , variegated tumor mass of size 11x9x7cms seen originating from lower pole of kidney.



Fig 1-shows Lt.Kidney with attached tumor mass



Fig 2-Lung pieces showing nodular metastatic deposits



Fig 3-Omentum with tumor nodules on surface

Cut section of kidney :

Shows grayish white tumor mass of size 6x5x4 cms along with cystic area of size 2x2 cms filled with mucinous material. At places areas of hemorrhages and necrosis seen.



Fig 4- C/S of kidney showing mucin filled cystic area and grayish white tumor mass

IV. Microscopy

- Section shows normal tubules and glomeruli along with spindle shaped tumor cells with pleomorphic nuclei and prominent nucleoli arranged in sheets, bundles, fascicles and at places herring bone pattern seen.
- At places conventional Renal cell carcinoma pattern with round to polygonal cells having vacuolated cytoplasm and central nuclei are seen.
- Section of lung shows alveoli, alveolar septa with spindle shaped tumor cells arranged in bundles and fascicles.
- Section of omentum shows clusters of mature adipocytes along with spindle shaped tumor cells arranged in bundles and fascicles.

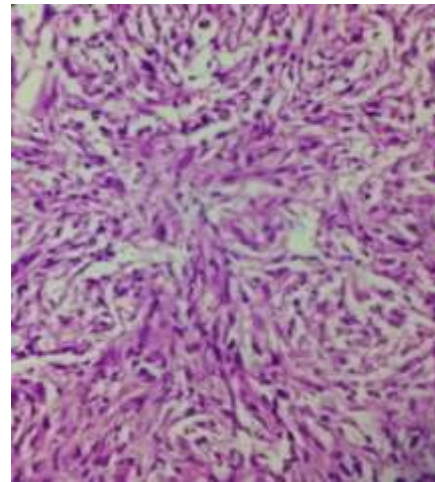
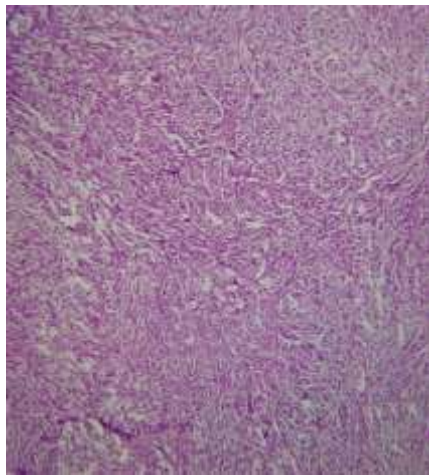


Fig 5(10X) H&E&Fig 6(40X) H&E :Section shows spindle shaped tumor cells with pleomorphic nuclei and prominent nucleoli arranged in sheets,bundles

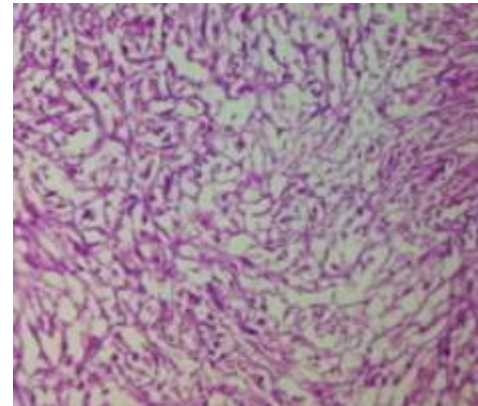
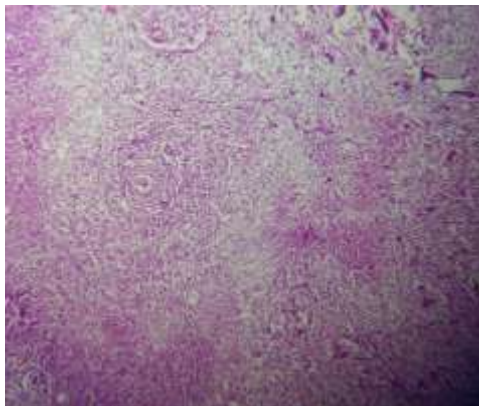


Fig 7(10X)H&E, Fig8 (40X)H&E-section shows conventional renal cell carcinoma

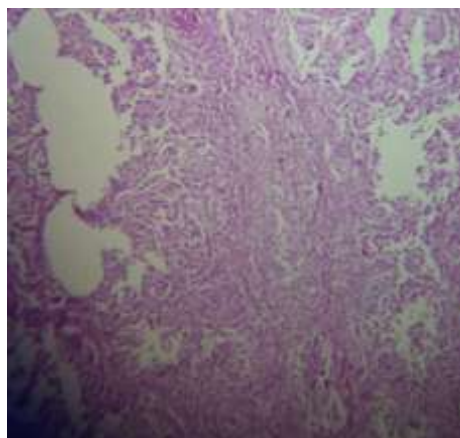


Fig 9- Lung section showing foci of spindle Shaped tumor cells (10X) H & E

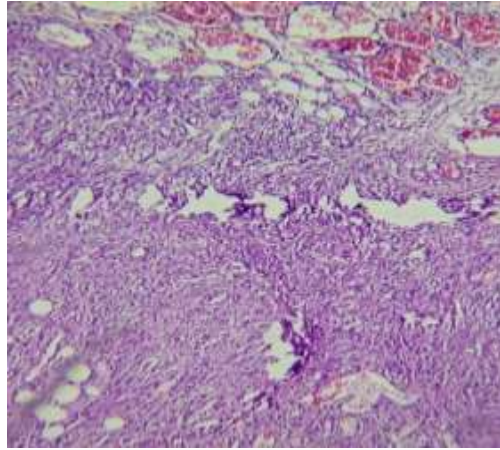


Fig 10-omental section showing foci of spindle shaped tumor cells indicates metastatic deposits(10X) H &E

V. Immunohistochemistry

The tumor cells are immunoreactive for EMA, Vimentin, CD10. And immunonegative for pancytokeratin AE1/AE3, PAX8, GATA3.

- EMA immunoreactivity indicates epithelial nature of tumor cells.
- Vimentin immunoreactivity indicates mesenchymal origin.
- CD10 is strongly and diffusely expressed by Renal cell carcinoma.



Fig 11- Stain for vimentin shows strong immunoreactivity

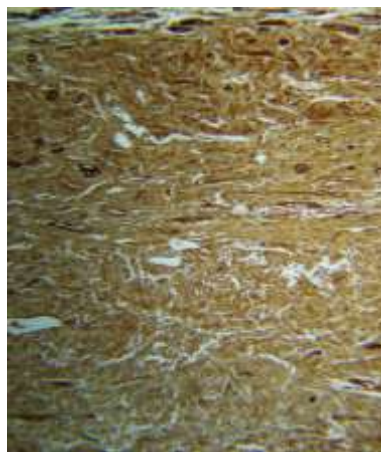


Fig 12- Stain for EMA shows strong immunoreactivity



Fig 13- Stain for CD10 shows strong immunoreactivity



Fig 14- Stain for Pancytokeratin AE1/AE3, GATA3, PAX8 show immunonegativity

VI. DISCUSSION

Renal cell carcinoma (RCC) is the most common malignancy of the kidney and consists of multiple subtypes.^[5] It accounts for 3% of adult malignancies and more than 90% of renal cancers.^[6] A history of cigarette smoking and obesity are risk factors that have the strongest correlation with RCC. According to the

2004 histologic classification of RCC by the WHO, the clear cell histologic subtype is by far the most common, comprising 70% of all cases. Other less common histologic subtypes include papillary, chromophobe and collecting duct which represent 10%, 5% and less than 1% of all renal cell carcinomas, respectively.^[1]

Histologic differentiation of these subtypes of RCC is clinically important as both the papillary and chromophobe subtypes have a less aggressive clinical course and better prognosis than the clear cell variety. While initially felt to represent a primary renal cell sarcoma, sarcomatoid RCC is now considered a form of dedifferentiated carcinoma and is therefore not a distinct histologic entity. The diagnostic feature of this cancer is the intermingling of typical renal cell carcinoma with a component of sarcomatoid features comprising of spindle cells without organization or resembling malignant fibrous histiocytoma or fibrosarcoma.^[7]

It is defined as any subtype containing foci of is seen in 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. Heterogeneous sarcomatoid transformation has been reported in other histologic subtypes of sarcomatoid RCC and conventional RCC is the main type of tumor to undergo heterologous transformation^[8]. Sarcomatoid differentiation is indicative of an aggressive tumor as demonstrated by their rapid growth and poor prognosis. They have a high incidence of metastasis to lungs and bone at presentation^[9]. Several small series have shown a median survival of less than one year in patients with sarcomatoid differentiation^[10].

On microscopic examination, two main histologic subtypes of sarcomatoid component have been described. A fibrosarcoma like appearance and malignant fibrous histiocytoma characterized by a greater degree of nuclear pleomorphism and occasional multinucleate osteoclast like giant cells. Rare cases show hemangioendothelioma like pattern. Some tumors 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. On immunohistochemistry they are positive for AE1/AE3, vimentin, EMA, CD-10, MIB-1 which supports epithelial origin^[11].

Investigations 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 differentiation could be suggested based on imaging characteristics, potentially with MRI. On MRI, sarcomatoid RCC have an irregular or infiltrative morphology and demonstrates heterogeneous T₂ signal intensity and enhancement. Internal necrosis and evidence of aggressive local or distant behaviour was frequently observed^[12].

Surgical management has been the mainstay of treatment for RCC but does not improve the prognosis in those with sarcomatoid differentiation. This has prompted the use of chemotherapy in combination with nephrectomy in these patients and seen promise in prolonging survival^[13]. Little information is available on genetic alterations in sarcomatoid RCC. Mutations of the P53 tumor suppressor gene are reported to be more prevalent in sarcomatoid cell component (79%) compared with clear cell component (14%) of sarcomatoid renal cell carcinoma arising from clear cell renal cell carcinoma^[14].

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