

Sarcomatoid renal cell carcinoma: A case series

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Abstract

Introduction: Renal cell carcinoma is the most common form of Renal malignancy. Sarcomatoid renal cell carcinoma represents a high grade dedifferentiation of Renal cell carcinoma. This phenotype can occur in all subtypes of renal cell carcinomas, including clear cell, papillary, chromophobe, and collecting duct carcinoma. Although prognosis in sarcomatoid renal cell carcinoma is known to be extremely poor, little is known about the clinicopathologic information about them. **Methods and Methodology:** The population of this retrospective study consisted of all patients who underwent surgery for Renal cell carcinoma between January 2010 and February 2013 in the Department of Pathology, Sri Ramachandra university and Research Institute. A total of 64 renal cell carcinoma cases were diagnosed among which 9 had sarcomatoid changes. All 9 cases were reassessed and the diagnosis was confirmed on the basis of morphologic features. Complete baseline and follow-up data were available for analysis for all 9 patients. **Conclusion:** Present study showed that sarcomatoid changes were seen more commonly in younger age group with male preponderance and necrosis was a consistent feature. Sarcomatoid areas constitute 1-25% in majority of cases which makes extensive sampling an important measure. Majority of renal cell carcinomas with sarcomatoid changes have a high grade and high stage. Identification of sarcomatoid areas in renal cell carcinoma is of prime significance as it has prognostic implications.

Keywords: sarcomatoid, renal cell carcinoma.

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INTRODUCTION

Renal cell carcinoma is the most common form of renal malignancy¹. Sarcomatoid renal cell carcinoma represents a high grade dedifferentiation of Renal cell carcinoma. This phenotype can occur in all subtypes of renal cell carcinomas, including clear cell, papillary, chromophobe, and collecting duct carcinoma²⁻³. Although prognosis in sarcomatoid renal cell carcinoma is known to be extremely poor⁴, little is known about the clinicopathologic information about them.

METHODS AND METHODOLOGY

The population of this retrospective study consisted of all patients who underwent surgery for renal cell carcinoma

between January 2010 and January 2014 at the Department of Pathology, Sri Ramachandra university and Research Institute. A total of 64 renal cell carcinoma cases were diagnosed among which 9 had sarcomatoid changes. All 9 cases were reassessed and the diagnosis was confirmed on the basis of morphologic features. Complete baseline and follow-up data were available for analysis for all 9 patients. Staging was reassessed according to the 2010 American Joint Committee on Cancer/International Union Against Cancer classification^{4,5}. Nuclear grading was done according to Fuhrman grading system⁶. The diagnostic criteria for SRCC was considered as atypical spindle cells resembling any sarcoma, present at least in 1% of the total tumor or present in 1 microscopic low-power field. The pattern of sarcomatoid change was categorised as fibrosarcoma like, malignant fibrous histiocytoma like, leiomyosarcoma like, Unclassified and heterogenous components if present as Chondrosarcoma like, Osteosarcoma like and Rhabdomyosarcoma like⁷. Tumor necrosis was considered as the presence of any microscopic coagulative necrosis within the tumor⁸.

RESULTS

Out of the 64 renal cell carcinoma cases evaluated 9 cases (14.2%) were found to have sarcomatoid differentiation

(Fig 1). Demographic details: There were 7 males and 2 females with a male-to-female ratio of 3.5:1. The age ranges of these patients were between 32 – 76 years with a median age of 51.5 years. In total, 6 patients had disease on the right side and 3 had disease on the left side. No bilateral tumor cases were reported. Radical nephrectomy was done for all the 9 patients. The mean tumor size was 10.9 cm (range: 5-18cm) (Table 1). Carcinomatous component: The carcinomatous component of sarcomatoid renal cell carcinoma was identified to be clear cell type in three cases (33.3%), collecting duct type in two cases (22.2%), papillary type in one case (11.1%), chromophobe type in one case (11.1%), unclassified in one case (11.1%) and mixed type in one case (11.1%) (Fig 2). The pT stage was high (stage 3 and 4) in majority of the cases (55.5%) and low (stage 1 and 2) in 45.5 % of cases. Out of the 9 SRCC 6 patients (66.6%) showed Fuhrman’s nuclear grade 4, two cases of grade 3 (22.2%) and one case showed nuclear grade 2 (11.1%). Tumor necrosis was a consistent feature in all the SRCC cases with majority of cases showing 1-25% necrosis. Lymphovascular invasion was present in 2 cases (22.2%)

and hilar invasion was identified in 3 cases (33%). (Table 2).

Sarcomatoid component

The sarcomatoid component was morphologically assessed to be Fibrosarcoma like in 4 cases (44.4%), Malignant Fibrous Histiocytoma like in 3 cases (33.3%), Leiomyosarcoma like in 2 cases (22.2%). No heterologous sarcomatoid changes were found in the cases studied. Percentage of sarcomatoid component was 1-25 % in 4 cases (44.4%), 26-50% in 2 cases (22.2%), 51-75% in 2 cases (22.2%) and 75-100% in one case (11.1%). (Fig 3 and 4)

Metastasis

Lymph node metastasis was seen in one patient (11.1%). Lymphovascular invasion was present 2 cases and hilar invasion was present in 3 cases. Distant metastasis was observed in 2 cases (22.2%) (Table 3). One case to bone and the other to lungs.

Prognosis

4 out of 9 cases succumbed to death within 6 months of diagnosis despite radical nephrectomy. 3 patients are alive till date with response to surgery and chemotherapy. The follow up details of 2 patients were not available.

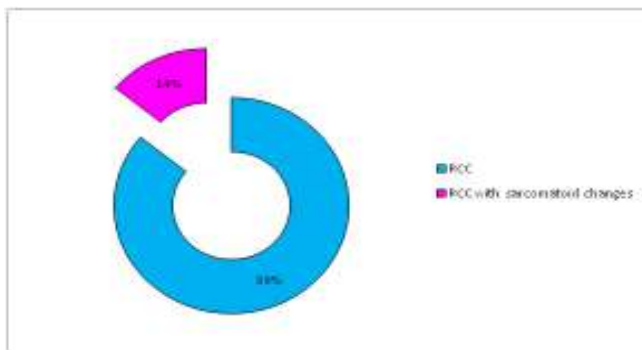


Figure 1

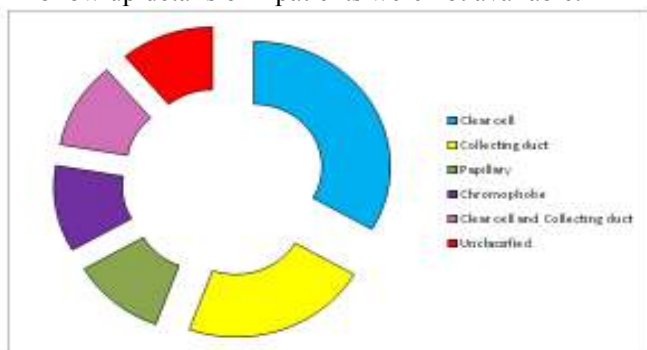


Figure 2

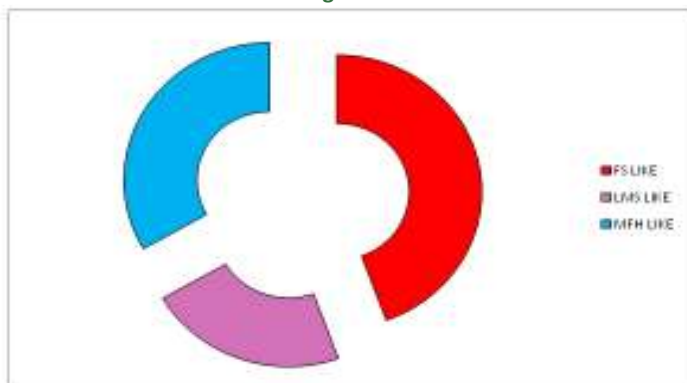


Figure 3

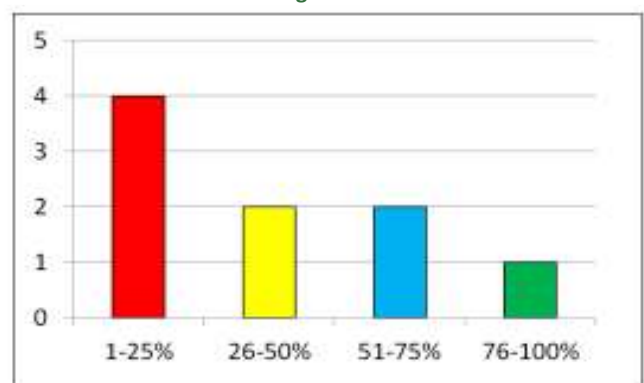


Figure 4

Legend

- Figure 1: Incidence of sarcomatoid changes in renal cell carcinoma
- Figure 2: Primary epithelial component in sarcomatoid renal cell carcinoma
- Figure 3: Pattern of sarcomatoid changes
- Figure 4: Percentage of sarcomatoid component

Table 1: Demographic data

Characteristic	Value
Number of patients	9
Age	
Range	32 – 76 years
Median	51.5 years
Sex	
Males	7
Females	2
Tumor laterality	
Right	6
Left	3
Tumor size	
Range	5-18cm
Median	10.9 cm

Table 2: Carcinomatous component

Primary RCC	
Clear cell RCC	3
Collecting duct RCC	2
Papillary RCC	1
Chromopobe RCC	1
Clear cell and collecting duct RCC	1
Unclassified RCC	1
pT stage	
1a	0
1b	1
2a	1
2b	2
3a	4

3b	0
3C	0
4	1
Fuhrman grade	
1	0
2	1
3	2
4	6
Necrosis	
No of cases examined	9
No of cases with necrosis	9
Percentage of necrosis	
0-25%	5
36-50%	3
51-100%	1
Lymphovascular invasion	
No of cases with LVI	2
Percentage of cases with LVI	22.2%
Hilar invasion	
No of cases with Hilar invasion	3
Percentage of cases with Hilar invasion	33.3%

Table 3

Lymph node metastasis	
No of cases with LN metastasis	1
Percentage of cases with LN metastasis	11.1%
Distant metastasis	
No of cases with distant metastasis	2
Percentage of cases with distant metastasis	22.2%

Table 4

Features	Current study	Cheville JC <i>et al</i> (2004)	De Peralta Venturina <i>et al</i> (2001)	Mian BM <i>et al</i> (2002)
Number of SRCC cases	9/64	120/2381	101/952	108
Incidence	14.2%	5%	10.6%	NA
Range	32 – 76 yrs	36–87	33-80 yrs	35-76
Median	51.5 yrs	61	60 yrs	56
Sex				
Males	7	64.2%	62%	69%
Females	2	35.8%	38%	31%
Laterality				
Right	67%			50%
Left	33%			50%
Tumor size				
Range	5-18cm	2.5–23 cm	3-25 cm	3-25
Median	10.9 cm	9.4 cm	9.2 cm	11

DISCUSSION

Sarcomatoid renal cell carcinoma was recognized and was first named by Farrow *et al.* in 1968⁹. Sarcomatoid renal cell carcinoma represents a high grade dedifferentiation of epithelial neoplasm of the kidney and is not a specific morphogenetic subtype of renal cell carcinoma.^{2,10} Pure sarcomatoid carcinoma or sarcomatoid carcinoma associated with epithelial elements that do not conform to usual renal carcinoma cell types are considered as unclassified renal cell carcinoma. Sarcomatoid renal cell carcinoma was seen in

14 % of all renal cell carcinoma cases which is higher than the reports in the literature^{3,8}. In our study a male preponderance was seen with a male to female ratio of 3.5:1 which is similar to the previously reported case series of sarcomatoid renal cell carcinoma.^{3,8,11} The age incidence was 32-76 with a median age of 51 years which is a little less than than the other studys^{3,8,11}. Right sided tumors were more common in our study which was in discordance with the study by Mian BM *et al* in which sarcomatoid changes was seen equally on both sides. In the current study tumor size varied from 5 to 18 cms with

a median tumor size of 10.9 cm which was similar to the other studies. (Table 4) The epithelial component of sarcomatoid renal cell carcinoma was more commonly clear cell type which is in concurrence with the study by Cheville JC *et al* and Mian BM *et al* but in discordance with the study by De Peralta Venturina *et al* according to which chromophobe renal cell carcinoma was more commonly associated with sarcomatoid changes. The most commonly reported Furhman nuclear grade in our series of sarcomatoid renal cell carcinoma was grade 4 with Furhman nuclear grade 3 and 4 accounting for 89% which is similar to the study by De Peralta Venturina *et al* accounting for 95%. The most common primary tumor stage of sarcomatoid renal cell carcinoma was pT stage 3 with stage 3 and 4 accounting for 56% of all stages which was similar to the trends seen in the study by Cheville JC *et al*(58%) but lesser than the study by De Peralta Venturina *et al* (69%).Necrosis was a consistent feature in all the 9 sarcomatoid renal cell carcinoma cases in our series but was seen only in 91% in the study by Cheville JC *et al* and 75% in the case series of De Peralta Venturina *et al*. The extent of tumor necrosis was <50% in 89% of the patients in our series and was 93% in the study by Cheville JC *et al*. The most common sarcomatoid pattern was Fibrosarcoma like pattern in concurrence with the study by De Peralta Venturina *et al*.44% of cases showed sarcomatoid component in less than 25% of the tumor. No heterologous component was identified in our case series unlike the study by De Peralta Venturina *et al* where 2% cases and the study by Cheville JC *et al* 1% cases showed heterologous components. Nodal metastasis was present in only 1 case in our series which is in discordance with the other studies which show increased tendency for metastasis^{3,8,11}. Distant metastasis was present in 2 cases which is lower than the other studys. Little information is available on genetic alterations in sarcomatoid renal cell carcinoma. Mutations of the p53 tumor suppressor gene are reported to be more prevalent in sarcomatoid components (79%) compared with clear cell components (14%) of SRCC arising from clear cell renal cell carcinoma¹².Ultrastructural findings show frequent desmosomal junctions, confirming the epithelial nature of the neoplasm.¹³

Limitations

In the study only limited no of patients were included and complete prognostic evaluation could not be done.

CONCLUSION

Present study showed that sarcomatoid changes were seen more commonly in younger age group with male preponderance and necrosis was a consistent feature. Sarcomatoid areas constitute 1-25% in majority of cases

which makes extensive sampling an important measure. Majority of renal cell carcinomas with sarcomatoid changes have a high grade and high stage. Identification of sarcomatoid areas in renal cell carcinoma is of prime significance as it has prognostic implications.

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