Case Report

Indian Journal of Pathology: Research and Practice
Volume 4 Number 2, July - December 2015
DOI : https://dx.doi.org/10.21088/ijprp.2278.148X.4215.11

Sarcomatoid Renal Cell Carcinoma

Sreelekha B. V.*, M. Shanthi, C. Bharath****

*Assistant Professor, ** Professor, Department of Pathology,Vijayanagara Institute of Medical Sciences, Ballari,Karnataka

Abstract

Sarcomatoid renal cell carcinoma although a rare tumour of the kidney in adults [1%] is an aggressive neoplasm with poor prognosis [1]. On histology an epithelial type of renal cell carcinoma is combined with the sarcomatoid component as anaplastic spindle shaped cells without organisation. Sincesarcomatoid renal cell carcinoma accounts for 10-20% of the patients presenting with advanced disease and mortality there is need for improved and early diagnostic and therapeutic modalities in this regard.

Keywords: Sarcomatoid; Renal Cell Carcinoma; Kidney Cancer.

Case Details

A 50 year old female presented with right flankmass since four months. Clinical imaging revealed an enlarged right kidney. She underwent right sided radical nephrectomy the gross and histology of which revealed an aggressive neoplasm.The detailed microscopic study of multiple sections from this tumor was concluded asa sarcomatoid renal cell carcinoma. After the cytoreductive surgery, chemotherapy was included to complete the management. By now the patient had stage III [T3aNoMo]disease. Postsurgery the patient survived for fifteen days after which she died of extensive lung metastases.

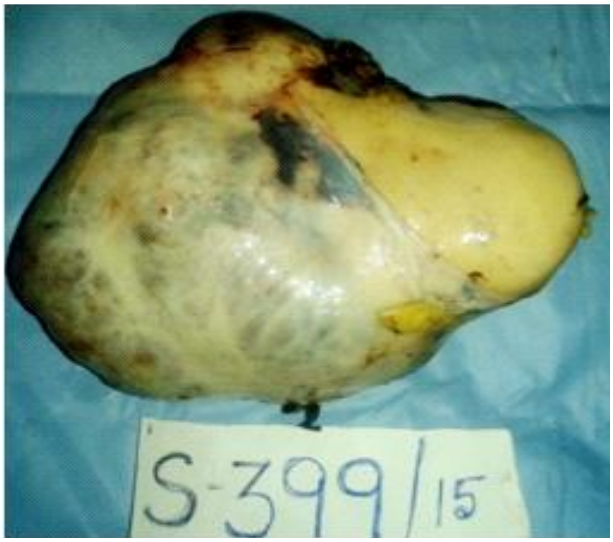

Fig. 1: External surface of radical nephrectomy specimen

Corresponding Author: Sreelekha B. V., Assistant Professor, Department of Pathology, Vijayanagara Institute of Medical Sciences, Ballari, Karnataka 583104.

E-mail: ramspsm@gmail.com

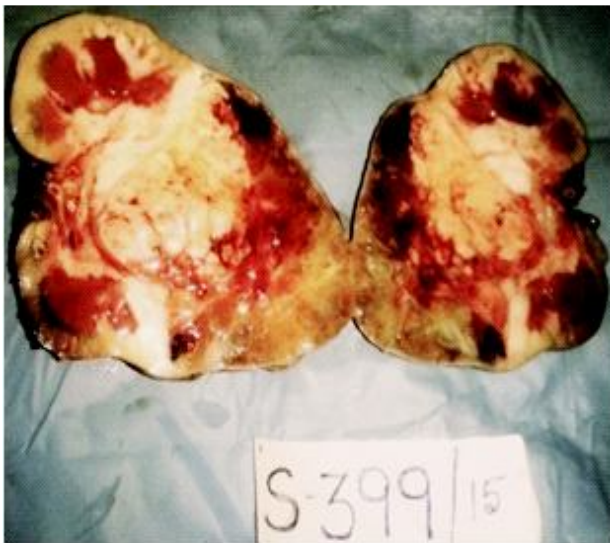

Fig. 2: Cut section showing grey white areas

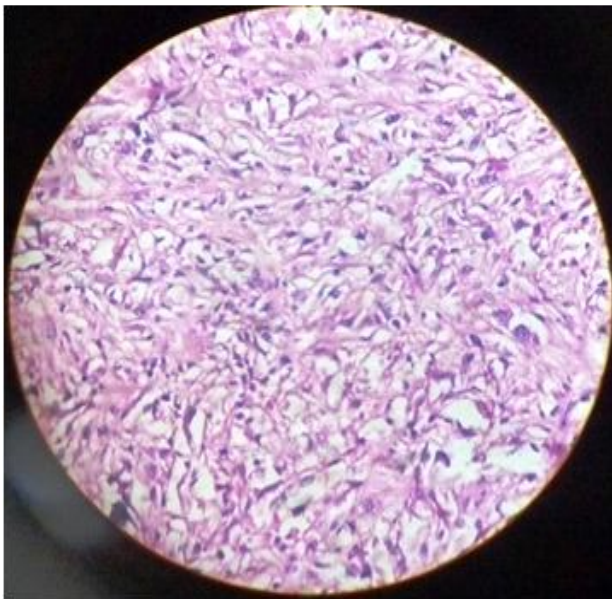

Fig. 3: ShowingSarcomatoidcomponent

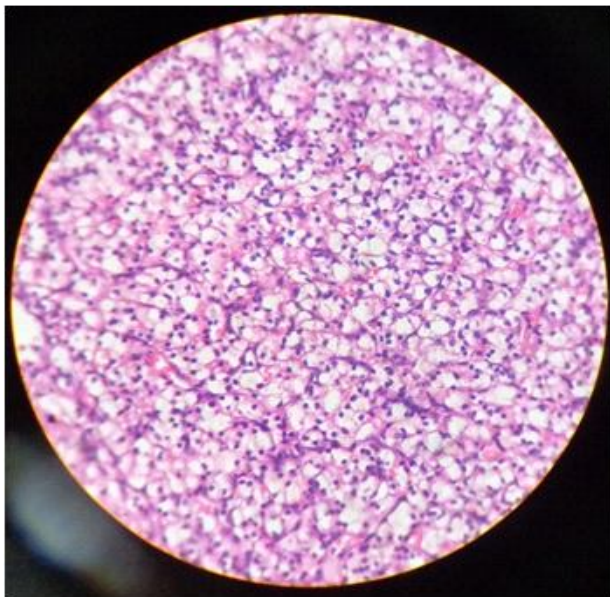

Fig. 4: Showing Clearcell component

Gross of the specimen measured 15x 12x8cm with nodular external surface. Cut section-showed a large tumor occupying the whole of the cut section with parts of poles being spared. The tumor showed a grey white fish flesh like appearance.

Microscopic study of multiple sections from the tumor showed predominantly anaplastic spindle shaped cells with moderate to scant cytoplasm having pleomorphic nucleus with a high nuclear grade. Mitotic rate was 2-4 / hpf. Many areas of necrosis were seen to be surrounded by these tumor cells. Multiple foci showed lobules of cell with clear cytoplasm and uniform round nucleus confirming the classical clear cell component. As the sarcomatoid component was more than 50% of the tumor a final diagnosis of Sarcomatoid renal cell carcinoma was made. Extrarenal extension and capular invasion were also noted on microscopy.

Discussion

The term sarcomatoid renal cell carcinoma was established and considered a separate histologic type because of its highly aggressive nature [**3**]. Updated classification schemes in the late 1990s disbanded this category and considered this entity to be a feature related to extensive chromosomal rearrangements [4, 5]. It was believed that these rearrangements led to identical spindle-cell morphology regardless of the primary epithelial histology. Speculations had also risen that the sarcomatoid pattern may not represent a continuum of dedifferentiation from classic RCC, but rather result from activation of a separate sarcomatoid stem cell within the tumor. Delahunt et al [6] later termed sarcomatoid characteristics the "final common dedifferentiation pathway" for renal tumors. Although now reclassified by the American Joint Committee on Cancer and Heidelberg pathology schemes, many urologic and medical oncologists consider sarcomatoid RCC to be a clinically relevant grouping because of the cohort's poor prognosis and its relative resistance to multiple forms of systemic therapy [7, 8]. Sarcomatoid renal cell carcinoma is currently defined in the 2004 WHO classification of renal tumors as any histologic type of renal cell carcinoma containing foci of high grade malignant spindle cells.

Renal cell carcinoma is the commonest malignant tumour of the kidney in adults with prognosis depending upon histologic type, microscopic grade, tumour size, involvement of the renal vein, capsule and distant spread.

In our case patient had a stage III disease [T3aNOMO] which progressed to a stage IV disease within a span of fifteen days culminating in lung metastases. Overall 5-year survival rate is seventy percent for renal cell carcinomasbut the sarcomatoid variant has the poorest prognosis with median survival of six months [2] and in our case survival was just four and a half months.

Grossly the tumor caused marked enlargement of the right kidney which correlates with the large size of the sarcomatoid RCC seen in other studies. Cut section showed a greywhite fish flesh appearance that closely relates to the sarcomatoid areas. On histology, thesarcomatoid component was more than 50% [80%] comprising of neoplastc spindled cells while the epithelial component was the classical clear cell type

present only focally. Because of the high incidence of clear cell RCC, this histology gets associated with >80% of thesarcomatoidRCCs [3, 5, 7].

Although sarcomatoid RCCs may resemble classic sarcomas, important differences are recognizable. First, primary renal sarcomas are extremely rare in adults, accounting for <1% of renal malignancies. When they do occur, almost half are leiomyosarcomas, which contain smooth muscle components that are rarely observed in sRCCs [6]. Additionally, primary renal sarcomas should not contain any classic areas of RCC [7]. Other tumors that may mimic sRCCs are sarcomatous urothelial tumors. These may be distinguished by the presence of flat in situ regions and/or squamous differentiation.The differential diagnosis also includes high grade transitional carcinoma of renal pelvis, malignant rhabdoidtumour of kidney, wilmstumour of kidney in adults and primary renal sarcomas like malignant fibrous histiocytoma, fibrosarcoma[9].

A review of IHC staining of sRCCs by DeLong and colleagues demonstrated that the sarcomatoid areas still express cytokeratin AE1/AE3 and vimentin in 97% and 56% of cases, respectively [10]. Classic markers observed in mesenchymal tissue and sarcomas, such as desmin and actin, are infrequently expressed in sRCCs.

Conclusion

Sarcomatoid renal cell carcinoma almost alwayspresents with advanced stage of the disease and accounts for 10-20% of the mortality from renal cancer. This entity should be treated as a separate clinical cohort since diagnostic cytoreductive surgery alone is inefficient [11]. It has to be invariably combined with appropriate targeted chemotherapy and adjuvant radiotherapy [12].

References

1. Nelon G. Ordonez, JuanRoai. Urinarytract. Ackermann's surgical pathology,9th edition volume1, page no 1191-1192.
2. Ro, J.Y., Ayala, AG.and Sella, A. Sarcomatoid renal cell carcinoma: Clinicopathological: A study of 42 cases. Cancer, 1987; 59: 516-20.
3. Farrow GM, Harrison EG, Jr, Utz DC, et al. Sarcomas andsarcomatoid and mixed malignant tumors ofthe kidney in adults. Cancer. 1968;22:545–563.
4. Kovacs G, Akhtar M, Beckwith BJ, et al. The Heidelberg classification of renal cell tumours.J Pathol. 1997; 183: 131–133.
5. Störkel S, Eble JN, Adlakha K, et al. Classification of renal cell carcinoma: Workgroup No. 1. Union InternationaleContre le Cancer (UICC) and the American Joint Committee on Cancer (AJCC) Cancer. 1997; 80: 987–989.
6. Delahunt B, Eble JN, McCredie MR, et al. Morphologic typing of papillary renal cell carcinoma: Comparison of growth kinetics and patient survival in 66 cases. Hum Pathol. 2001; 32: 590–595.
7. de Peralta-Venturina M, Moch H, Amin M, et al. Sarcomatoid differentiation in renal cell carcinoma: A study of 101 cases. Am JSurgPathol. 2001;25:275–284.
8. Cheville JC, Lohse CM, Zincke H, et al. Sarcomatoid renal cell carcinoma: An examination of underlying histologic subtype and an analysis of associations with patient outcome. Am J SurgPathol. 2004; 28: 435–441.
9. Peterson. R. O, Urologic pathology. 2nd Ed. Philadelphia, Lippincot,1992; p.95
10. Ro JY, Ayala AG, Sella A, et al. Sarcomatoid renal cell carcinoma: Clinicopathologic. A study of 42 cases.Cancer. 1987;59:516–526.
11. Porter, K. A. Tumour of the kidney. In Syniiners, W. SteEd. Sytemic pathology Vol.8, The kidneys. London,Churchill Livingstone, 1992; P.581.
12. David M Nanus, Alexandria Garino, Matthew, Maureen, Janice. Active Chemotherapy for Sarcomatoid and Rapidly Progressing Renal Cell Carcinoma.Cancer, 2004; 101: 1545-51.