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SARCOMATOID RENAL CELL CARCINOMA WITH HEMOPTYSIS A RARE CASE REPORT

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Abstract :
Renal cell carcinomas are rare tumours and sarcomatoid renal cell carcinomas are even more uncommon. Clearcell and chromophil components are the most common carcinomatous constituents in sarcomatoid renal cell carcinomas. These tumours usually present with a triad of haematuria, abdominal pain and an abdominal mass with more than 80 percent of patients presenting with metastasis. We describe a 62 -year old man with metastatic sarcomatoid renal cell carcinoma at presentation who underwent radical nephrectomy and splenectomy but succumbed prior to initiation of chemotherapy.

Keyword :
Renal cell carcinoma Sarcomatoid metastasis hemoptysis

ntroduction Sarcomatoid renal cell carcinomas are rare tumours with grave prognosis. Sarcomatoid renal cell carcinomas (RCC) were first described in 1968 by Farrow *et al.*[1]

We describe a case report of a 62 year old man with sarcomatoid RCC. Case Our patient presented to casualty with four episodes of hemoptysis, hematuria and left loin pain for the last 15 days. Examination revealed distended abdomen and a palpable mass in the left hypochondrium and lumbar region. Ultrasonogram showed a left-sided irregular heteroechoic lesion measuring 12x9.1cm and extending up to the diaphragmatic surface and compressing the spleen. On computed tomography, a heterogeneous contrast-enhancing solid and exophytic mass measuring 15x4cm arising from the posterior part of the left kidney and occupying the upper pole was observed [Figure1A]. A left-sided pleural effusion and a contrast -enhancing focal mass in the right lower lobe of lung were also noted[Figure 1B]. A complete blood count, liver and renal function tests were normal. Urine revealed gross haematuria.The patient underwent a left-sided radical nephrectomy with splenectomy.

Macroscopy:

The nephrectomy specimen measured 14x13x9cm and weighed 190 grams. The external surface was irregular with a capsular breach. The cut surface showed an ill-circumscribed greyish white solid mass of size 11x8x7cm [Figure2A].The tumour grossly infiltrated into the pelvis and perinephric fat. The spleen specimen measured 11x9x7cm and the cut surface appeared normal[Figure 2B].

Microscopy: Sections studied showed a tumour composed of small nests and clusters of round to polygonal cells with variable amount of clear to granular cytoplasm. The nuclei was oval with moderate to severe anisokaryosis . There were also areas showing bands of plump spindle shaped cells with eosinophilic cytoplasm and oval to bipolar nuclei. Few scattered mitotic figures with occasional atypical mitotic figures were seen [Fig 3A,3B,3C]. Immunohistochemical staining showed strong vimentin positivity[Fig 4A] and positive for cytokeratin.[Fig 4B]

Discussion:

Renal cell carcinomas comprise 85% of renal neoplasms in adults. [2]These tumors usually present with a triad of haematuria, abdominal pain and an abdominal mass as our patient had presented. Clinical features of metastasis at presentation are common with sarcomatoid RCC and more than 80% patients demonstrate metastasis at the time of presentation. Lung and bone are the commonest sites of metastasis in sarcomatoid RCC[3]. Our patient had lung metastasis. Lung metastasis is associated with the worst prognosis.[3] Other prognostic factors in sarcomatoid RCC are angioinvasion,[4] female sex, increasing age,[5] with pathologic grading[3]. Fuhrman system of nuclear grading based on nuclear size, contour and conspicuousness of nucleoli is the most widely accepted grading system. More than 2/3rds of sarcomatoid RCC have high Fuhrman nuclear grade at presentation[6]. The Fuhrman nuclear grading in our patient was also 4. Median survival with metastatic sarcomatoid RCC is about 4 to 18 months.[7] Sarcomatoid RCC is a distinct pathologic variant of RCC, defined histologically by presence of highly pleomorphic spindle cells resembling sarcoma along with varying degrees of clear to **granular epithelial cells of RCC. Microscopically, our case had sarcomatous pleomorphic spindle cells admixed with epithelial components along with atypical mitotic figures. Renal cell carcinomas (RCC) are classified based on their gross, histological, ultrastructural and immunohistochemical appearance by the WHO into clear cell, chromophobe, chromophil, renal medullary, collecting duct and unclassified types.**[2] , [3] Sarcomatoid RCC is not included under any subtype since they can dedifferentiate from any of the above types. Sarcomatoid component can be homologous or heterologous. The homologous components resemble fibrosarcoma, malignant fibrous histiocytoma while the heterologous elements includes controsarcoma and astiosarcoma[3,8]. A neuroendocrine and liposarcomatoid component has also been described in sarcomatoid RCC.[9, 10] A sarcomatoid component of the tumour without a definite epithelial component leads to classification under the unclassified type[3]. The incidence of sarcomatoid transformation in RCC is about 5%,[8] and it varies from 2.9% to 32% in various studies. [2] ,[11] Sarcomatoid component expresses cytokeratin and vimentin.

Cytokeratin 8, 18 and 19 and EMA are common to RCC and SRCC[3]. **Our case showed strong vimentin positivity and** cytokeratin positivity. Chromosomal changes in sarcomatoid RCC are nonspecific. One study reported p53 gene alterations in sarcomatoid transformation.[3] Advances in molecular genetics and immunohistochemistry have enabled diagnosis of more variants of RCC.

Treatment includes surgery (debulking), hormonal (medroxypregesterone and androgen) therapy, chemotherapy (either doxorubicin or non-doxorubicin based) and immunotherapy(IFN alfa, IL-2). Interleukin 2 based therapies have shown the best results for survival[3]. Our patient was planned for chemotherapy but succumbed prior to treatment on 10th post operative day.

In conclusion we report a rare case of clear cell RCC with sarcomatoid differentiation and lung metastasis. This malignancy is being described for its rarity and poor prognosis.

References:

[1] Farrow GM, Harrison EG, Jr., Utz DC: Sarcomas and sarcomatoid and mixed malignant tumors of the kidney in adults. 3. Cancer 1968, 22:556-63.

[2] Pradhan D, Kakkar N, Bal A, Singh SK, Joshi K. Sub-typing of renal cell tumours; contribution of ancillary techniques. Diagn Pathol 2009;4:21.

[3] Chao D, Zisman A, Freedland SJ, Pantuck AJ, Said JW, Belldegrun AS. Sarcomatoid renal cell carcinoma: basic biology, clinical behavior and response to therapy. Urol Oncol 2001;6:231-8.

[4] Viswanathan S, Desai SB, Prabhu SR, Amin MB: Squamous differentiation in a sarcomatoid chromophobe renal cell carcinoma: an unusual case report with review of the literature. Archives of pathology & laboratory medicine 2008, 132:1672-4.

[5] Cangiano T, Liao J, Naitoh J, Dorey F, Figlin R, Belldegrun A. Sarcomatoid renal cell carcinoma: biologic behavior, prognosis, and response to combined surgical resection and immunotherapy. J Clin Oncol 1999;17:523-8.

[6] Dall'Oglio MF, Lieberknecht M, Gouveia V, Sant'Anna AC, Leite KR, Srougi M. Sarcomatoid differentiation in renal cell carcinoma: prognostic implications. Int Braz J Urol 2005;31:10-6.

[7] Pal SK, Jones JO, Carmichael C, et al. Clinical outcome in patients receiving systemic therapyfor metastatic sarcomatoid renal cell carcinoma: A retrospective analysis. Urol Oncol 2012.

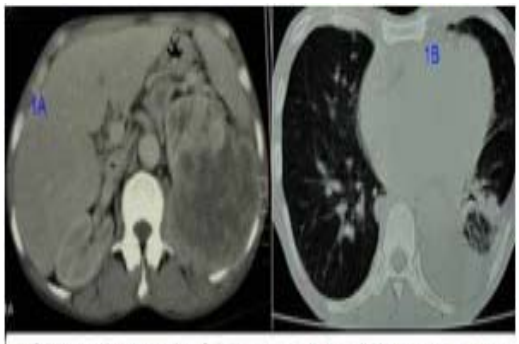
[8] Quiroga-Garza G, Khurana H, Shen S, Ayala AG, Ro JY. Sarcomatoid chromophobe renal cell carcinoma with heterologous sarcomatoid elements. A case report and review of the literature. Arch Pathol Lab Med 2009;133:1857-60.

[9] Kuroda N, Tamura M, Hes O, Michal M, Gatalica Z. Chromophobe renal cell carcinoma with neuroendocrine differentiation and sarcomatoid change. Pathol Int 2011;61:552-4.

[10] Anila KR, Mathew AP, Somanathan T, Mathews A, Jayasree K. Chromophobe renal cell carcinoma with heterologous (liposarcomatous) differentiation: a case report. Int J Surg Pathol 2012;20:416-9

[11] Kanamaru H, Sasaki M, Miwa Y, Akino H, Okada K. Prognostic value of sarcomatoid histology and volume-weighted mean nuclear volume in renal cell carcinoma. BJU 1999;83:222-6

CT Images



1A] Computed tomography of abdomen revealing a solid heterogenous mass in the left kidney compressing the spleen.

1B] Computed tomography of chest shows reduced left-lung volume, left sided pleural effusion and infiltrates

Gross Images H & E Images

2A] Nephrectomy specimen with ill-circumscribed, greyish-white solid mass in the upper pole with foci of brownish areas. Tumor infiltrates into pelvis and peri-nephric fat

2B]Splenectomy specimen with normal cut-surface

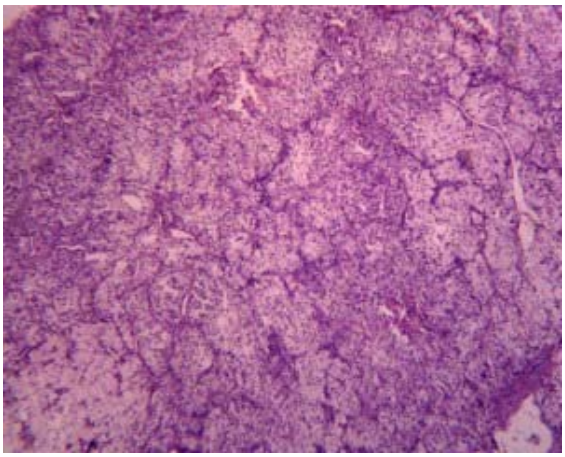
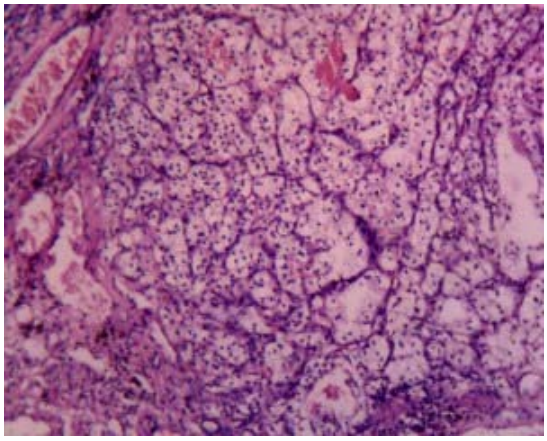


Fig 3A: 10 X Tumour shows nest of Clear Cells with fibro vascular septa Fig 3B: 45 X Tumour shows nest of Clear Cells with intervening congestion



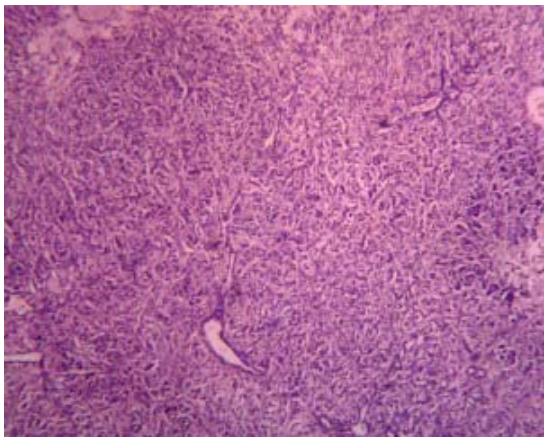


Fig 3C : 10 X Tumour shows areas of sarcomatoid spindle Cells with atypical mytotic figures

Immunohistochemistry Images (Vimentin & Cytokeratin)

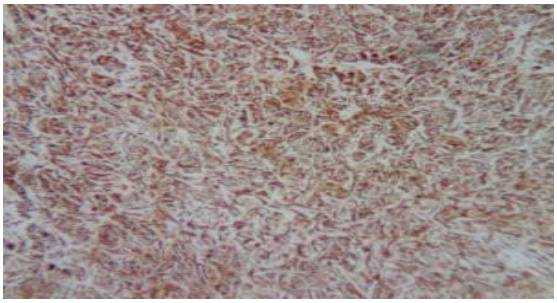


Fig 4A: Sarcomatoid spindle Cells shows positive for vimentin Fig 4B : Tumour Cell Shows positive for Cytokeratin