

# Sarcomatoid Variant of Renal Cell Carcinoma

Pages with reference to book, From 125 To 126

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## Introduction

Sarcomatoid renal cell carcinoma is a rare tumour of the kidney which accounts for 1% of renal neoplasms in adults and has been reported in its 6.5% of renal cortical carcinomas in different studies. The diagnostic morphological feature of this cancer is the intermingling of typical renal cell carcinoma with a component of sarcomatoid features, comprising of spindle cells without organisation or resembling malignant fibrous histiocytoma or fibrosarcoma<sup>4</sup>. The prognosis of this highly malignant carcinoma is poor and nephrectomy is ineffective in its management because extra-renal invasion is usually present at the time of diagnosis<sup>5</sup>. Three cases diagnosed histologically as sarcomatoid renal cell carcinoma are presented.

## Case Report

### Case 1

A 50 years old male presented with the complaints of right sided flank pain of a few month's duration. Examination revealed a firm mass occupying the right side of the lumbar region. Urine examination showed microscopic haematuria. Ultrasonography confirmed the mass in the right kidney. At exploratory laparotomy a huge right renal tumour was found which was adherent to peri renal fascia. The tumour mass, along with right kidney and right suprarenal gland was excised in toto.



Figure 1. Sarcomatoid variant of renal cell carcinoma showing spindle shaped component, giving sarcoma - like appearance (H&Ex200).

On gross examination an oval mass of 18x12x11 cm was found at the upper pole of the kidney, involving the right suprarenal gland and perirenal fat. Histological examination revealed a tumour comprising of pleomorphic epithelial cells, forming tubular papillary and sarcomatoid patterns. Granular and oncocytic differentiation was also seen and tumour cells had invaded the right suprarenal gland, renal capsule, perinephric fat and three lumbar lymph nodes.

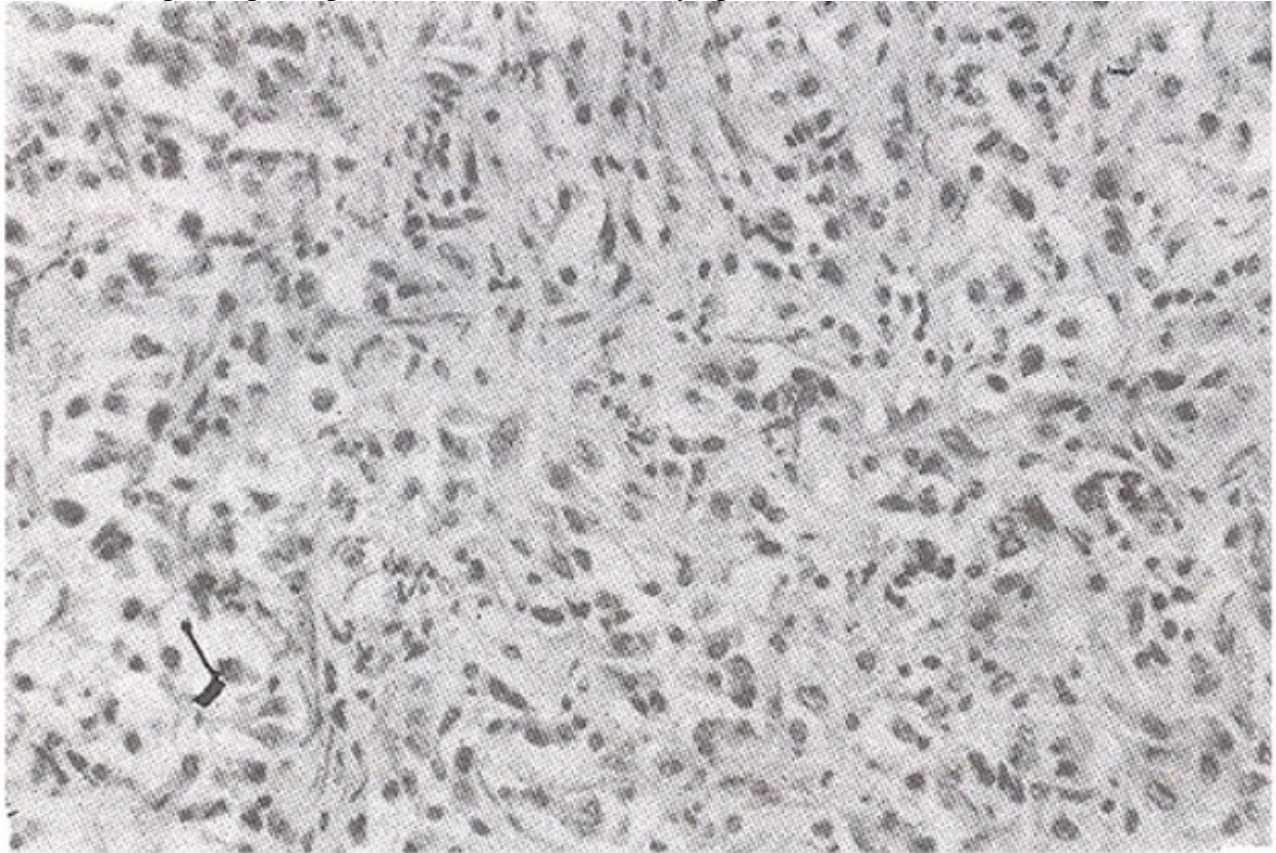


Figure 2. Area of characteristic clear cells of renal cell carcinoma admixed with spindle shaped cell (H&Ex400).

### Case 2

A 59 years old male presented with complaints of haematuria and pain in the left lumbar region. On examination a renal mass was found in the left side of the abdomen. Ultrasonography and intravenous urography showed a left renal mass. Surgical exploration revealed a tumour in the upper pole of the left kidney, both of which were excised. On gross examination of the specimen a cystic, rounded mass measuring 16x14x10 cm surrounding the upper pole of the kidney was seen. Histological examination revealed a tumour comprising of sheets and acini of round to poly-hedral cells with granular cytoplasm. Papillary areas and spindle cell components were also seen at places. The tumour cells were invading the renal capsule.

### Case 3

A 72 years female presented with mass in the right Iwnbar region and hypertension. Ultrasonography and urography showed a renal mass in the right side. On laparotomy atumourmass was found in the right kidney which was excised. Gross examination of the specimen showed a distorted kidney measuring 12x7.5x6 cm. On cut section, there was complete loss of renal architecture and was replaced by a yellow white tumour with a whorled, fish-flesh appearance. Histological examination revealed a highly aggressive tumour comprising of whorling fascicles of spindle shaped tumour cells with bizarre



nuclei and high mitotic rates. Rhabdomyoblastic differentiation and areas of typical clear cell carcinoma were also seen.

## Discussion

Renal cell carcinoma is the commonest malignant tumour of the kidney in adults. Its incidence increases with age. The overall 5 years survival rate of this tumour is about 70%. The prognosis is related to several factors, including the involvement of renal vein, renal capsule, distant spread, tumour size, microscopic grade and histological types of renal cell carcinoma. The sarcomatoid renal cell carcinoma has the poorest prognosis, because majority of the patients have disseminated tumour (Stage IV) at the initial presentation and the median survival of all the patients is 6 months<sup>6</sup>. This may also be related to tumour grade since sarcomatoid renal cell carcinoma by definition belongs to grade IV category<sup>7</sup>. The proportion of sarcomatoid component in the tumour does not determine the prognosis and no difference in the prognosis has been found whether the sarcomatoid component is higher (more than 50%) or low (less than 50%)<sup>2</sup>. In one of our cases, the sarcomatoid component was nearly 70%, in the remaining two, the sarcomatoid component was less than 50% (about 30% and 10% respectively). The stromal component comprised mostly of fibrohistiocytic tissue in two cases (Figure 1). Rhabdoid element was also seen in one case. These sarcomatoid components had high degree of anaplasia. The epithelial differentiation was towards classical clear cell carcinoma in one case (Figure 2) and granular cell differentiation with papillary areas in two cases. In two cases the tumour involved the upper poles of the kidneys, in the third entire kidney was involved by the tumour. The tumour diameter ranged from 12-18 cm. A solid, multinodular appearance with widespread cystic areas was observed. On cut section, the sarcomatoid components showed white, firm fish-flesh appearance. In all three cases renal capsule and perinephric fat were invaded by the tumour, which is consistent with other studies describing this disease<sup>4</sup>. Three lumbar lymph nodes and ipsilateral adrenal glands also showed tumour invasion in one case.

The clinical, sonographic and morphological features of our cases are consistent with other studies. The differential diagnosis however, includes anaplastic clear cell and granular cell carcinomas, high grade transitional carcinoma of the renal pelvis, malignant rhabdoid tumour of the kidney, Wilms' tumour of the kidney in adults and primary renal sarcomas e.g., malignant fibrous histiocytoma and fibrosarcoma. Since the classical areas of clear cell carcinoma and granular cell carcinoma were found in all cases and immature embryonal structures were absent, the diagnosis was simple and straight forward. Fine needle aspiration biopsy is a useful technique for the diagnosis of renal cell carcinoma and other renal tumours<sup>8</sup>. On cytology sarcomatoid renal cell carcinoma appears as clusters of cells with the appearance of renal cell carcinoma along with cells cytologically consistent with a sarcoma<sup>9</sup>. Prior to surgery fine needle aspiration biopsy was not performed in the present study. On immunocytochemistry in most cases the sarcomatoid cells were keratin, EMA and vimentin positive<sup>3</sup>. Cytogenetic studies are also helpful in establishing the diagnosis. We could not perform these tests due to lack of facilities. It is suggested that sarcomatoid carcinoma although a very rare variant of renal cell carcinoma, should be included in the differential diagnosis especially when stromal elements are found in adenocarcinoma of the kidney. This is a distinct entity because it always has a highly malignant behaviour and the worst prognosis among the renal parenchymal carcinomas. In addition to surgery, adjuvant radiotherapy and chemotherapy is required in preventing its dismal prognosis.

## References

1. Tomera, K.M., Farrow. G.M. and Lirber. M.M. Sarcomatoid renal carcinoma. J. Urol.,

1983;130:657-59.

2. Bertoni, F., Fern. C., Benati, P. et al. Sarcomatoid carcinoma of the kidney. *J. Urol.*, 1987;3825-28.
3. Deitchman, B. arid Sidher, G.S. Ultrastructural study of a sarcomatoid variant of renal cell carcinoma. *Cancer*, 1 1980;46:1152-57.
4. Peterson, R.O, Urologic pathology. 2nd Ed.. Philadelphia, Lipponcot, 1992, p. 95.
5. Porter, K.A. Tumours of the kidney. In Syniiners, W. Ste Ed. Systemic pathology Vol. 8, The kidneys. London, Churchill Livingstone, 1992, P. 581.
6. Ro, J.Y., Ayala, AG. and Sella, A. Sarcomatoid renal cell carcinoma: Clinicopathological: A study of 42 cases. *Cancer*, 1987;59:516-20.
7. Lanigan, D., Conroy. R., Barry- Walsh, C. et al. A comparative analysis of grading system in renal adenocarcinoma. *Histopathology*, 1 994;24:473.6.
8. Pilloti, S., Rilke, F, Alasio, L. et al The role of fine needle aspiration in the assessment of renal masses. *Act" cytol.*, 1988;32:1-9.
9. Cristallini, E.G., Paganelli, C and Bolis, G.B. Role of fine needle aspiration biopsy in the assessment of renal masses. *Diagn. Cytopathol.*, 1991 ;7:32-35.