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CASE REPORT

Primary Fibrosarcoma Of Kidney- A Rare Case

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ABSTRACT

Primary Fibrosarcoma of the kidney is a very rare tumour.^[1,2] We report here, a case of a 55 year old man, presenting with a gradually progressive left loin swelling and weakness.

Key Words: Fibrosarcoma, kidney, tumour.

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Introduction

A majority of the renal malignancies are either renal cell carcinomas or transitional cell carcinomas. Primary Fibrosarcoma of the kidney is a very rare tumour, which may be confused with leiomyosarcoma or sarcomatoid renal cell carcinoma. Immunohistochemistry for cytokeratin, vimentin and desmin helps to categorise the lesions. The rarity of the lesion and the aggressive nature of Primary fibrosarcoma of the kidney prompted us to write this case report.

Case Report

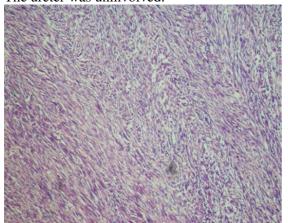
A 55-year-old male, non smoker and nonalcoholic presented to the OPD with complaints of anorexia, weight loss, a left loin mass and pain in the lumbar region. The lump gradually increased in size. On examination, the patient had marked pallor. There was no other significant finding on general examination. Per abdomen examination revealed a mass in the left lumbar region, which was firm to hard in consistency. The liver and spleen were not palpable. The kidney function tests revealed microscopic haematuria. Ultrasonography and contrast enhanced computer tomography of the abdomen showed a lobulated, heterogeneously enhancing soft tissue mass, measuring 22 X 12 cm in the left kidney, extending upto the splenic hilum, infiltrating the psoas muscle,

encasing the left renal artery and invading the left renal vein. A clinical diagnosis of renal cell carcinoma was thus made. The patient underwent left radical nephrectomy and splenectomy.



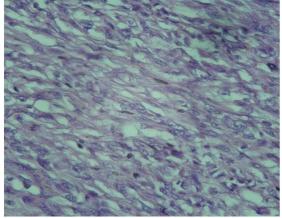
[Table/Fig 1]: Gross: Specimen of the left kidney, enlarged measuring $22 \times 12 \times 10$ cm. with bosselated external surface.

We received the specimen of the left kidney which was enlarged and measured $22 \times 12 \times 10$ cm [Table/Fig 1]. The external surface was bosselated and partially encapsulated. The cut surface showed a lobulated firm grayish-white tumour, involving almost the whole kidney and reaching up to the capsule. The ureter was uninvolved.



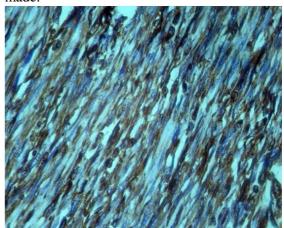
[Table/Fig 2]: Photomicrograph showing spindle shaped cells arranged in Herring bone pattern (H & E, x10).

Microscopic examination showed a partially encapsulated tumour tissue comprising of spindle cells in the fascicular arrangement [Table/Fig 2]. The cells were spindle to round shaped, having ill-defined, scant to moderate eosinophilic cytoplasm. The nuclei were spindle to oval shaped, with moderate anisonucleosis, having irregular nuclear and irregularly membranes distributed chromatin [Table/Fig 3].



[Table/Fig 3]: Photomicrograph showing spindle to round cells having ill-defined, scant to moderate eosinophilic cytoplasm with nuclei showing moderate anisonucleosis having irregular nuclear membrane and irregularly distributed chromatin (H & E, x40).

Based on the morphological findings, a differential diagnosis of sarcomatoid Renal and carcinoma (RCC) fibrosarcoma of the kidney were considered. Following this, immunohistochemistry was done with a panel of markers including Pan Cytokeratin, Desmin and Vimentin. The tumour cells were negative for most of the markers, except for Vimentin [Table/Fig 4], which was diffusely positive in most of the tumour cells. Based on the morphological and the immunohistochemical findings, a final diagnosis of Fibrosarcoma of the kidney was made.



[Table/Fig 4]: Immunohistochemistry photomicrograph showing strong cytoplasmic positivity for vimentin in the spindle shaped cells.

The patient made an uneventful recovery from the surgery and received postoperative radiotherapy. Thereafter, the patient was lost to follow up.

Discussion

Primary sarcoma of the kidney is very rare and it constitutes about 1-3% of all renal tumours.[1],[2] Histopathologically, sarcomas are malignant mesenchymal tumours of the kidney, of which leiomyosarcomas are more common and primary fibrosarcomas of the kidney are very rare. Some reported cases actually represented sarcomatoid RCC. with Presently, modern improved histochemical techniques, pathologists are able to separate the actual cases of primary fibrosarcoma of the kidney from sarcomatoid renal cell carcinomas. Cavaliere et al. found a single case in a 10-year survey of priming renal sarcomas and Grignon et al found a single case in a study of 17 cases of primary sarcomas of the kidney.[3],[4] They can be

differentiated only on the basis of immunohistochemistry, as fibrosarcomas are diffusely positive for vimentin and negative for CK and desmin, whereas sarcomatoid RCCs and leiomyosarcomas are positive for cytokeratin and desmin, respectively.

Fibrosarcoma is a tumour of the 40-60 year age group; affecting both sexes equally. Its origin is postulated to be from the capsule of the kidney, which contains much of the fibrous and the connective tissues. The tumour is large, solid and fleshy, with infiltrative margins. About 40% of the cases show the evidence of renal vein invasion peroperatively.

It is due to the lack of specific symptoms that a majority of the renal fibrosarcomas are difficult to diagnose early. The usual clinical presentation including abdominal haematuria and flank pain are late symptoms. diagnosed patients are when gastrointestinal or metastatic symptoms develop. [5]

Five-year survival rates are generally poor and have been reported to be as low as 10%. [6] Pettirssen in his review, found only two long-term survivors among 21 cases. [7]

Conclusion

Primary Fibrosarcoma of the kidney is a rare but an aggressive malignant renal tumour, with a very poor prognosis and a low five year survival. Hence, it must be differentiated from its morphological look-alikes, leiomyosarcomas and sarcomatoid renal cell carcinomas by immunohistochemistry.

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