



Case Report

Primary Fibrosarcoma of kidney: A Case Report

Subrata Pal*¹, Biplab Kr Biswas², Sritanu Jana³, Arindam Rakshit⁴, Rajani Sinha⁵

^{1&5}Post graduate trainees, ²Associate professor, ³Assistant professor, ⁴Demonstrator, Department of Pathology, Bankura Sammilani Medical College. Add: Gobindanagar, Kenduadihi, Bankura, West Bengal, India.

ABSTRACT

Primary renal fibrosarcoma is a rare entity. Renal sarcomas accounts only 1-3% of primary renal tumours, among which fibrosarcoma is rarer aggressive malignant mesenchymal lesion. Diagnosis is difficult because early lesion remains asymptomatic and tumour lacks specific symptoms. It should be differentiated from sarcomatoid renal cell carcinoma, secondary involvement by retroperitoneal fibrosarcoma or from other sites and other primary spindle cell sarcomas of kidney.

KEYWORDS: Primary fibrosarcoma, kidney

INTRODUCTION

Most of the malignant tumours of kidney arise from renal parenchyma and renal pelvis. Renal cell carcinoma (RCC) and transitional cell carcinoma (TCC) comprise most of the malignant renal lesions in adult age[1],[2]. Primary renal fibrosarcoma is a very rare entity. It is difficult to differentiate from sarcomatoid RCC and leiomyosarcoma[2],[3]. Recent advancement in immunohistochemical application of cytokeratin, vimentin, desmin and S-100 helps to differentiate rare mesenchymal entities[3]. We want to share such a rare case of primary renal fibrosarcoma diagnosed by us.

CASE REPORT

A 62 year male patient was admitted in our emergency with complaints of recurrent right loin pain, anorexia and weight loss for last 4months. Clinical examination revealed a firm to hard lump at right lumbar region. He had no hepatosplenomegally and ascites. On abdominal ultrasound he had a large solid heterogeneous mass of 13x10 cm in dimension, involving lower pole and middle portion of right kidney. Radiological assay did not reveal any invasion into surrounding tissue, renal vein and inferior vena cava (IVC). He was clinically diagnosed as renal cell carcinoma. Radical

nephrectomy was done and specimen was sent for histopathological examination.

On gross examination (Figure1) the kidney contained a mass involving the lower lobe and mid-portion. The mass was circumscribed and partially encapsulated, measuring 13x10x8 cm with bosselated outer surface. On cut section, it was gray- white lobular mass compressing the normal renal tissue at upper pole. Renal pelvis was encroached by the tumour tissue but ureter was not involved. No area of haemorrhage or necrosis was seen in gross examination. Microscopy showed encapsulated tumour tissue composed of elongated spindle cells arranged in fascicles giving rise to herring bone pattern (Figure 2, 3).

The spindle cells have delicate eosinophilic cytoplasm, spindle to oval nuclei with irregular nuclear membrane, irregular chromatin condensation and moderate anisonucleosis. Based on the light microscopical findings, we concluded the diagnosis of primary renal fibrosarcoma. Immunohistochemical staining was done with cytokeratin cocktail, desmin, vimentin and S-100 protein. The tumour cells showed diffuse vimentin positivity. With the assistance of IHC staining pattern and histopathological examination, final diagnosis of primary renal fibrosarcoma was given. After 6 month follow up patient was uneventful after nephrectomy.

Figure 1: Gross picture of the cut open kidney with solid white fleshy tumour



Figure 2: Microscopy of renal fibrosarcoma- fascicular arrangement of spindle cells in herring bone pattern (H & E stain, low power view)

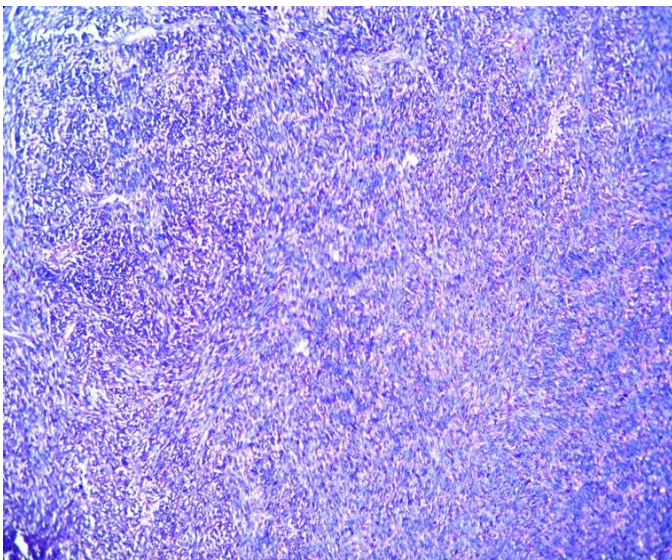
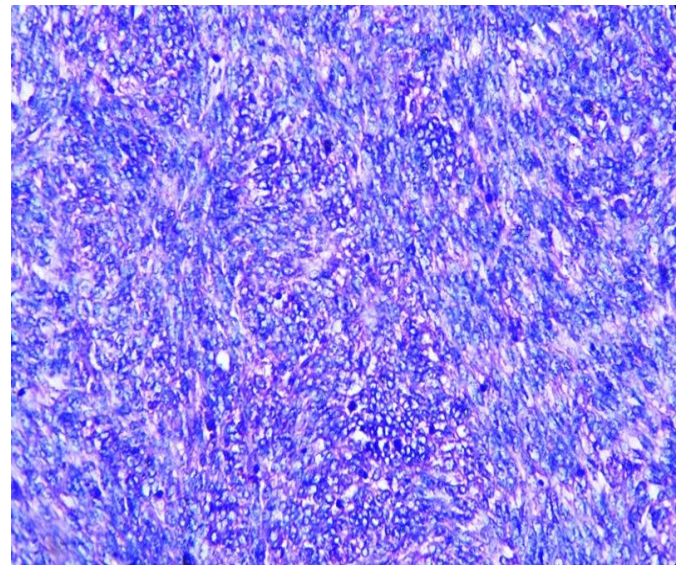


Figure 3: Microscopy shows spindle cells with delicate polar cytoplasm, moderate anisonucleosis (H & E stain, high power view).



DISCUSSION

Primary renal sarcomas are uncommon tumours accounting only 1-3% of all renal neoplasms[4],[5]. Among these renal malignant mesenchymal neoplasms leiomyosarcoma is most frequent histologic type. Other rare sarcomas of kidney include fibrosarcoma, rhabdomyosarcoma, osteogenic sarcoma and liposarcoma[4]. Diagnosis of primary renal

fibrosarcoma should meet following criteria: (1) Exclusion of metastatic sarcomatous lesion from elsewhere in the body. (2) Gross features should be suggestive of its renal origin rather of secondary involvement by retroperitoneal sarcoma. (3) Exclusion of sarcomatoid RCC[1],[6].

In our case, the encapsulated tumour mass had only renal involvement without any other site of involvement. No retroperitoneal soft tissue invasion found. Multiple serial sections did not reveal any epithelial component and IHC showed negative staining for epithelial markers (CKs) excluding the close differential diagnosis, sarcomatoid RCC.

Fibrosarcoma of kidney is a rare tumour of elderly age (40-60year) with equal sex distribution[1],[3]. It is supposed to be originated from mesenchymal tissue and contains fibrous and connective tissue. The tumour is usually large, solid and encapsulated with a fleshy cut surface and infiltrating margin. The tumour is rapidly growing and may show renal vein invasion in about 40% of cases[1],[2],[3]. Early lesions are usually asymptomatic. Due to lack of specific symptoms like haemorrhage, abdominal mass, flank pain, renal fibrosarcoma often diagnosed at advanced stage. Computerised tomogram or excretory urography is able to show large mass lesion and lesion becomes hypo-vascular in renal arteriography[1],[2].

Microscopy of renal fibrosarcoma comprises of fascicular arrangement of spindle cells with focal 'herring bone' pattern. The cells have scanty eosinophilic cytoplasm and oval to spindle shaped nuclei with minimal pleomorphism. Mitosis is frequent and focal necrosis may be evident. Immunohistochemistry show diffuse positive staining with vimentin in the tumour cells and negative staining for actin and S-100 protein[1],[4]. Radical nephrectomy is the definitive management for primary renal fibrosarcoma and it has been proved resistant to radiotherapy and chemotherapy[1],[2],[4]. Long term follow up show 5 year survival rate is only 10% in different literature[1],[2],[4].

CONCLUSION

Primary renal fibrosarcoma is a very rare, potentially aggressive malignant mesenchymal tumour. It must be distinguished from secondary involvement from retroperitoneal fibrosarcoma, sarcomatoid RCC and other primary spindle cell sarcomas of kidney. Immunohistochemistry and other ancillary imaging modalities have an important role adjacent to histopathology in correct diagnosis.

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*Corresponding author: Dr. Subrata Pal
E-Mail: subratapal1985@gmail.com