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SOLITARY FIBROUS TUMOR OF KIDNEY: A CASE REPORT

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Abstract

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Key words: Solitary fibrous tumour of kidney, hemangiopericytoma.



Solitary fibrous tumor of kidney is a rare mesenchymal cell tumor. It is an unusual spindle cell neoplasm identical to hemangiopericytoma. About 50 cases have been reported in the available literature so far. We present a case report of a 60 year old female who presented with urinary incontinence and swelling of lower limbs since 2 months. The CT scan showed a well defined, large 22x16x14.5cm sized, lobulated mass arising from upper pole of left kidney. She was operated upon and the mass excised. Histology and immunohistochemistry helped in arriving at a definite diagnosis of a solitary fibrous tumour. It is being reported due to its rarity of occurrence in the kidney.

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INTRODUCTION

Solitary fibrous tumor of the kidney is a rare mesenchymal neoplasm. It is an unusual spindle cell tumor identical to hemangiopericytoma. It was first defined in the kidney by Gelb et al. in 1996 [1].

Though solitary fibrous tumors have been commonly seen as arising from the pleura, an origin from lung, orbit, salivary gland, breast, meninges, liver, kidney, prostate and urinary bladder is also known [2]. Origin from kidney is unusual,about50 cases have been reported in the available literature [3]. When occurring in the kidney, it presents as a mass lesion which can be mistaken clinically for a renal cell carcinoma and diagnosis is made only by histological examination and IHC techniques [4].

The present case is reported for the rarity of it's occurrence in the kidney, large size and having been mistaken for a renal cell carcinoma clinically.

CASE REPORT

A 60 year old female presented with complaints of urinary incontinence and swelling of lower limbs since 2 months. Hemogram, renal function tests and liver function tests were within normal limits. Chest x-ray showed borderline cardiomegaly. Per abdomen examination showed a periumbilical mass measuring 18x15cm. A contrast enhanced CT scan showed a well defined, large 22x16x14.5cm sized, lobulated mass arising from upper pole of left kidney, with a few calcific foci.Considering patient's history and clinical observation, a malignant tumor of kidney was suspected, possibly a renal cell carcinoma. She was operated upon and the mass excised. Grossly histopathology examinationrevealed а 23x19x12cm bosselated and nodular left sided nephrectomy specimen weighing 2600 gm with congested blood vessels on the surface andattached stump of ureter

measuring 1 cm in length; and perinephric fat sent separately measured 20x8x1cm. [Fig. 1] Cut section showed a grey white, nodular/lobulated mass occupying the upper part of the kidney with only 3x5cm of normal appearing kidney in the lower pole. The entire mass measured 20x16cm. Nodules were separated by fibrous septa. The largest nodule measured 15x11cm and smallest one measured 4x3cm. [Fig. 2]Pelvis and calyces were compressed and attenuated.

On microscopy, well circumscribed, а multilobulated neoplasm with hypercellular and hypocellular areas was noted. Hypercellular areas were composed of spindle cells arranged in sheets and storiform pattern with collagenisation. Fine collagen bands separated tumour cells. Hypocellular areas showed myxoid degeneration. [Fig. 3] Occasional mitosis was noted about 1/10HPF. Large areas of necrosis were seen. Within the tumour many blood vessels were identified, giving an appearance of hemangiopericytoma.Adjacent compressed parenchyma showed features of chronic renal pyelonephritis. Ureter (surgical margin) and Gerota's fascia were free of tumour. A tentative diagnosis of solitary fibrous tumour of the kidney was made; to be confirmed by IHC in order to exclude a monophasic synovial sarcoma.

On immunohistochemistry,tumor showed strong positivity with CD 34 and Bcl 2. [Fig. 4] About 30-40 % of tumor cells showed nuclear positivity for Ki 67. The diagnosis of solitary fibrous tumor was confirmed. Patient was advised follow up.

DISCUSSION

Solitary fibrous tumor is a rare mesenchymal tumor. It is mostly of pleural origin. Nonetheless, extrapleural cases have also been reported [2].Among the tumours of renal origin, in the literature surveyed, 15%



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were located in renal capsule, 6% in peripelvis, and 3% in renal pelvis, while 76% had an unknown site of origin [5]. The present case arose within the kidney and occupied almost the entire renal parenchyma.

Macroscopically, SFTs are well-demarcated, solid lesions with a gray-whitish colour on cross-section, measuring from 1 to 22cm [4]. Similar findings were noted in this case where the neoplasm was lobulated, grey white and measured 20 cm in greatest dimension.

Microscopically, the present case showed long fascicles of spindle cell proliferation with hypo- and hypercellular areas. Cells were separated by hyalinised collagenous tissue. A diagnosis of SFT/monophasic synovial sarcoma was considered. Since IHC showed strong positive reaction with CD34 and Bcl 2, a final diagnosis of SFT was made. Synovial sarcomas are usually CD34 negative [4].

About 10-15% of all SFTs are malignant [3]. Malignancy is based on hypercellularity, mitoses (>4/10HPF), necrosis and an infiltrative edge [4]. The present case was well circumscribed, showed hypercellularity and occasional mitoses. The only feature favouring malignancy was necrosis. Well circumscribed tumors with necrosis behave indolently. However, the clinical behaviour cannot be predicted on histopathological basis with benign-appearing tumors also exhibiting aggressive behaviour and vice versa. Therefore, a long term follow up is mandatory.

CONCLUSION

Solitary fibrous tumor of the kidney is a rare mesenchymal cell tumor. Its definite diagnosis is made by histology and immunohistochemistry. Though the prognosis is good, follow-up is mandatory.



Fig. 1: Gross nephrectomy specimen with fibrofatty tissue



Fig. 2: Gross, cut section showing lobules



Fig. 3: Microscopy: Hypercellular and hypocellular areas with spindle cells in sheets and storiform pattern, 45x



Fig 4: CD 34 showed strong positivity **REFERENCES**

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