# FINAL PATHOLOGIC DIAGNOSIS:

Right kidney, adrenal, and caval thrombus, radical nephrectomy;

- Malignant high-grade sarcoma, most consistent with leiomyosarcoma (see comment)
- Tumor involves kidney and is 13 cm in greatest dimension.
- Tumor invades renal capsule into perirenal adipose tissue and renal sinus
- Tumor within caval thrombus and within lumen of renal vein.
- Renal vein margin positive for tumor,
- Tumor is within 0.5 mm of the perinephric adipose tissue margin.
- Ureteral margin negative for tumor.
- Adrenal gland, no tumor present.
- No tumor present in one lymph node within renal sinus.
- Non-neoplastic renal parenchyma and tumor with extensive infarction, consistent with therapeutic embolism.

## COMMENT:

The histologic differential diagnosis was that of a high grade malignant sarcomatoid neoplasm and included malignant fibrous histiocytoma, leiomyosarcoma, malignant melanoma, and unclassified renal cell carcinoma with sarcomatoid features. No definitive evidence of epithelial differentiation was identified either by histology or immunohistochemical studies. The combined histologic and immunohistochemical phenotype (see Microscopic Description) support a diagnosis of leiomyosarcoma. Although the tumor grossly appears to be arising from within the kidney, the very remote possibility of a metastatic tumor from another source such as uterus, can not be completely excluded so that clinical/radiologic correlation is recommended.

### CLINICAL HISTORY:

34 year old female with a renal cell carcinoma and caval thrombus

Clinical diagnosis: Right renal tumor with caval thrombus.

Specimen site: Right kidney with caval thrombus

### SPECIMEN(S) RECEIVED

A: Right kidney with caval thrombus"

### **GROSS DESCRIPTION:**

A. The specimen is labeled with the patient's name and number and designated as "Right kidney with caval thrombus": Received fixed in formulin is a 1,230 g. right nephrectomy specimen (23x14x10 cm.) with right adrenal gland  $(4, Ox1 \cdot 5x1.5 \text{ cm.})$  and surrounding perirenal fat measuring in thickness from 0 to 1.0 cm. Extending from the renal pelvis is a ureter that is 20 cm, in length and 1 cm. in greatest diameter. Replacing the upper pole of the kidney is a circumscribed, multilobulated, golden brown tumor mass that is 13x13x12 cm. The tumor has multiple areas of hemorrhage and necrosis. The tumor protrudes into the renal vein for a distance of 3 cm., and is present at the inked margin. There is a nodule of tumor that appears to penetrate the capsule and invade into the perirenal fat. Tumor invades into the renal sinus. The remaining of the renal cortex is tanbrown and without a well defined corticomedullary junction. There are small areas of hemorrhage inside the non-neoplastic renal cortex that measure between 0.3 and 0.5 cm. The adrenal gland consists of normal medulla and cortex and there appears to be no invasion by the previously described tumor. The adipose tissue is serially sectioned and no lymph nodes are identified. Pictures are taken. Representative sections are submitted.

Three additional fragments of apparent thrombus are identified inside the same container. The largest one is 9x5x3 cm. And the other two are 12x6x5x2 and 9x2x1 cm. No vessel wall is identified grossly, The largest thrombus appears to have a 3x1.5x1 cm. nodule of golden brown tissue suspicious for tumor.

### MICROSCOPIC DESCRIPTION:

The tumor is diffusely involving superior half of the kidney and extending in a circumscribed pushing fashion into renal sinus and perinephric adipose tissue. Tumor is within the lumen of the renal vein and is within caval thrombus. The majority of the tumor and non-neoplastic kidney is necrotic and there is foreign material and organizing thrombi within renal arteries, consistent with pervious therapeutic embolism. The tumor is composed of fascicles and sheets of pleomorphic spindled to round cells with numerous multinucleated bizarre giant cells. There are regions of extensive hyalinization. The mitotic activity is brisk (18 per 10 HPFs). Immunohistochemical stains demonstrate that the tumor cells are diffusely positive for smooth muscle actin, caldesmon, and vimentin while negative for EMA, cytokeratin AE1/3, CAN 5.2, Sl00, CD117 (CKIT), and CD34. The tumor involves renal vein and separately submitted caval thrombus. The tumor extends to the renal vein resection margin and is less than 0.5 mm from the perinephric adipose tissue margin of excision.

Subject ID: LM00020 Pathology Report Date: 1/14/04 Pathology Report Summary

#### **DIAGNOSIS:**

Thirty-seven outside slides, S-03-32847 A1-A26 (11-29-03), right nephrectomy specimen:

HIGH-GRADE MALIGNANT SPINDLE CELL NEOPLASM INVOLVING KIDNEY WITH EXTENSION INTO PERINEPHRIC CONNECTIVE TISSUE, RENAL SINUS, AND RENAL VEIN; arterial, ureteral, and soft tissue resection margins appear free of tumor. (SEE COMMENT)

Changes consistent with preoperative embolization,

Ureter segment, no tumor present.

Renal artery segment, no tumor present,

Adrenal gland, no tumor present.

### COMMENT

The contributor indicates that tumor formed a 13.0 x 13.0 x 12.0 cm mass situated within the upper pole of the kidney. The tumor extends into perinephric fat and renal sinus. The cytological/histological features of the tumor are those of a form of a high-grade malignant spindle cell neoplasia. The differential diagnosis includes: sarcoma, sarcomatoid carcinoma, and spindle cell malignant melanoma. The contributor has constructed a partial immunohistochemical profile for this tumor and it is available for review:

| negative  | CAM 5.2:  | negative   |
|---|---|--|
| negative  | S-100:  | negative   |
| negative  |   |  |
| negative in neoplastic  | cells   |  |
|   |   |  |
| 4 + immunoreactivit   | y in $>98\%$ of neo   | plastic cells, cytoplasrnic  |
| (finely granular and  | coarsely granula  | r) pattern   |
| 4 + immunoreactivity in >80% of neoplastic cells, cytoplasmic |   |  |
| (finely granular) pat   | tern  |  |
| 4+ immunoreactivity   | y in >60% of neop   | plastic cells, cytoplasmic   |
| (finely granular) pat   | tern  |  |
| negative  |   |  |
|   | negative<br>negative<br>negative<br>negative in neoplastic<br>4 + immunoreactivit<br>(finely granular and<br>4 + immunoreactivit<br>(finely granular) pat<br>4+ immunoreactivity<br>(finely granular) pat<br>negative | negativeCAM 5.2:negativeS-100:negativenegative in neoplastic cells4 + immunoreactivity in >98% of neo(finely granular and coarsely granula4 + immunoreactivity in >80% of neo(finely granular) pattern4+ immunoreactivity in >60% of neop(finely granular) patternnegative |

The results of this imunohistochemical profile strongly suggests that this lesion is of smooth muscle origin; i.e., leiomyosarcoma.