Tumors of kidney

Methodic materials for international students (IV-VI year)

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I. Primary tumors of kidney

1. Renal cell carcinoma

Prevalence: Renal cell carcinoma accounts for 2.3% of all adult cancers. In the United States in 1996, approximately 30,600 cases of renal cell carcinoma will be diagnosed and 12,000 deaths will result. Renal cell carcinoma has a peak incidence in the sixth decade of life and a male-to-female ratio of 2:1.

Aethiology

The cause is unknown. Cigarette smoking is the only significant environmental risk factor that has been identified. Familial settings for renal cell carcinoma have been identified (von Hippel-Lindau syndrome) as well as an association with dialysis-related acquired cystic disease, but sporadic tumors are far more common.

Morphology

Renal cell carcinoma originates from the proximal tubule cells. Various cell types (clear, granular, spindle) and histologic patterns (acinar, papillary, solid) are observed. However, cell type and histologic pattern do not affect treatment.

Classification

The TNM classification (TNM staging system) of the American Joint Cancer Committee for kidney cancer:

T: Primary tumor

Tx	Cannot be assessed
T0	No evidence of primary tumor
T1	Tumor £ 2.5 cm limited to kidney
T2	Tumor > 2.5 cm limited to kidney
T3a	Tumor invades adrenal gland or perinephric tissue

T3b Tumor extends into renal vein or vena cava
T4 Tumor invades outside of Gerota's fascia

N: Regional lymph nodes

Nx Cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single lymph node £ 2 cm
N2 Metastasis in a single lymph node > 2 cm and < 5 cm or multiple nodes none > 5 cm
N3 Metastasis in lymph node > 5 cm

M: Distant metastasis

Mx Cannot be assessed
M0 No distant metastasis
M1 Distant metastasis present

Clinical manifestations:

1. Gross or microscopic hematuria – 60% of patients
2. Flank pain
3. Abdominal mass } approx. 30% of cases.

The triad of flank pain, hematuria, and mass: only 10–15% of patients and is often a sign of advanced disease.

Sometimes abdominal palpation reveals one-sided nephroptosis (due to increase of the kidney weight). So, in cases if nephroptosis is revealed in patients, who are not predisposed constitutionally (thin young persons, more often females, with other signs of visceroptosis) the tumor should be suspected.

4. Symptoms of renal colic may be present in case of gross hematuria (due to renal obstruction): acute pain at the side of the affected kidney, irradiating to leg; the pain may be relieved by spasmolytics

5. Symptoms of metastatic disease (lungs, bones: cough, bone pain) - in 20–30% of patients at presentation.

6. Systemic symptoms such as fever, weight loss may be prominent.

7. Paraneoplastic syndromes:

8. Because of the more widespread use of ultrasound and CT scanning for diverse indications, renal tumors are being detected incidentally in patients with no urologic symptoms.

Laboratory Findings:

1. Hematuria is present in 60% of patients.

2. Paraneoplastic syndromes are not uncommon in renal cell carcinoma.

- Erythrocytosis from increased erythropoietin production occurs in 5%, though anemia is far more common

- Hypercalcemia may be present in up to 10% of patients.

- Stauffer's syndrome is a reversible syndrome of hepatic dysfunction in the absence of metastatic disease.

3. Intravenous urography:

- renal masses may be detected

4. Ultrasonic: renal masses; it’s possible to determine whether it is solid or cystic.
5. CT scanning is the most valuable imaging test for renal cell carcinoma. It confirms the character of the mass and further stages the lesion with respect to regional lymph nodes, renal vein, or hepatic involvement. It also gives valuable information on the contralateral kidney (function, bilaterality of neoplasm).
6. Chest radiographs exclude pulmonary metastases,
7. bone scans should be performed for large tumors and in patients with bone pain or elevated alkaline phosphatase levels.
8. MRI and duplex Doppler ultrasonography are excellent methods of assessing for the presence and extent of tumor thrombus within the renal vein or vena cava in selected patients.

Differential Diagnosis

I. If solid masses are already revealed:

Solid lesions of the kidney are renal cell carcinoma until proved otherwise.

Other solid masses include:

- angiomyolipomas (fat density usually visible by CT)

- transitional cell cancers of the renal pelvis (more centrally located, involvement of the collecting system, positive urinary cytology reports)
- adrenal tumors (supero-anterior to the kidney)
- oncocytomas (indistinguishable from renal cell carcinoma preoperatively)
- renal abscesses.

II. If solid masses are not yet revealed or their presence is doubtnful (suspected renal cancer)

Treatment & Prognosis

Radical nephrectomy is the primary treatment for localized renal cell carcinoma. Tumors confined to the renal capsule (T1–T2) demonstrate 5-year disease-free survivals of 90–100%. Tumors extending beyond the renal capsule (T3 or T4) and node-positive tumors have 50–60% and 0–15% 5-year disease-free survivals, respectively.

No effective chemotherapy is available for metastatic renal cell carcinoma.

- Vinblastine is the single most effective agent, with short-term partial response rates of 15%.
- Biologic response modifiers have received much attention, including alpha interferon and interleukin-2. Partial response rates of 15–20% and 15–35%, respectively, have been reported. Responders tend to have lower tumor burdens, metastatic disease confined to the lung, and a high performance status.
- Because of these low response rates, new investigations are ongoing with tumor vaccines and gene therapy.

One subgroup of metastatic patients has demonstrated long-term survival, namely, those with solitary resectable metastases. In this setting, radical nephrectomy with resection of the metastasis has resulted in 5-year disease-free survival rates of 15–30%.

2. OTHER PRIMARY TUMORS OF THE KIDNEY

Oncocytomas account for 3–5% of renal tumors and are indistinguishable from renal cell carcinoma by all imaging modalities. The biologic potential of these lesions is not well defined. These tumors are seen in other organs, including the adrenals, the salivary glands, and the thyroid and parathyroid glands.
Angiomyolipomas are **rare benign tumors** composed of fat, smooth muscle, and blood vessels. They are most commonly seen in patients with tuberous sclerosis (often multiple and bilateral) or in young to middle-aged women. CT scanning may identify the fat component, which is diagnostic for angiomyolipoma. Asymptomatic lesions less than 5 cm in diameter usually do not require intervention.

II. SECONDARY TUMORS OF THE KIDNEY (metastases)

The kidney is not an infrequent site for metastatic disease. Of the solid tumors, the lung is the most common (20%), followed by breast (10%), stomach (10%), and the contralateral kidney (10%). Lymphoma, both Hodgkin's and non-Hodgkin's, may also involve the kidney, though it tends to be a diffusely infiltrative process resulting in renal enlargement rather than a discrete mass.