Renal Tumors

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Classification

1. Primary Kidney Tumors

A. Benign Kidney Tumors

- Renal Adenoma
- Oncocytoma
- Angiomyolipoma
- Other benign tumors.

B. Malignant Kidney Tumors

- Renal Cell Carcinoma (RCC) - 90%
- Transitional Cell Carcinoma (TCC)
- Wilms Tumor
- Renal Sarcoma

2. Secondary Kidney Tumors

- The kidney is a frequent site of Mets for both solid and hematogenous tumors.

- The most frequent primary site of cancer was lung (20%), followed by breast (12%), stomach (11%), and renal (9%).

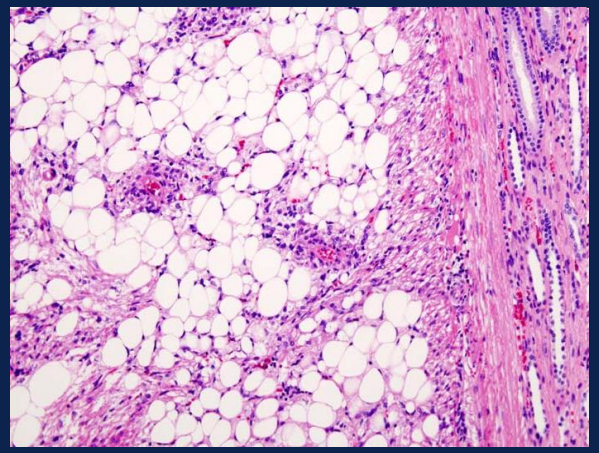
Benign
Renal Tumors

Renal Adenoma

- Small, well differentiated glandular tumor of the renal cortex.
- Site : proximal convoluted tubules – like Renal cell carcinoma.

Angiomyolipoma – renal hamartoma

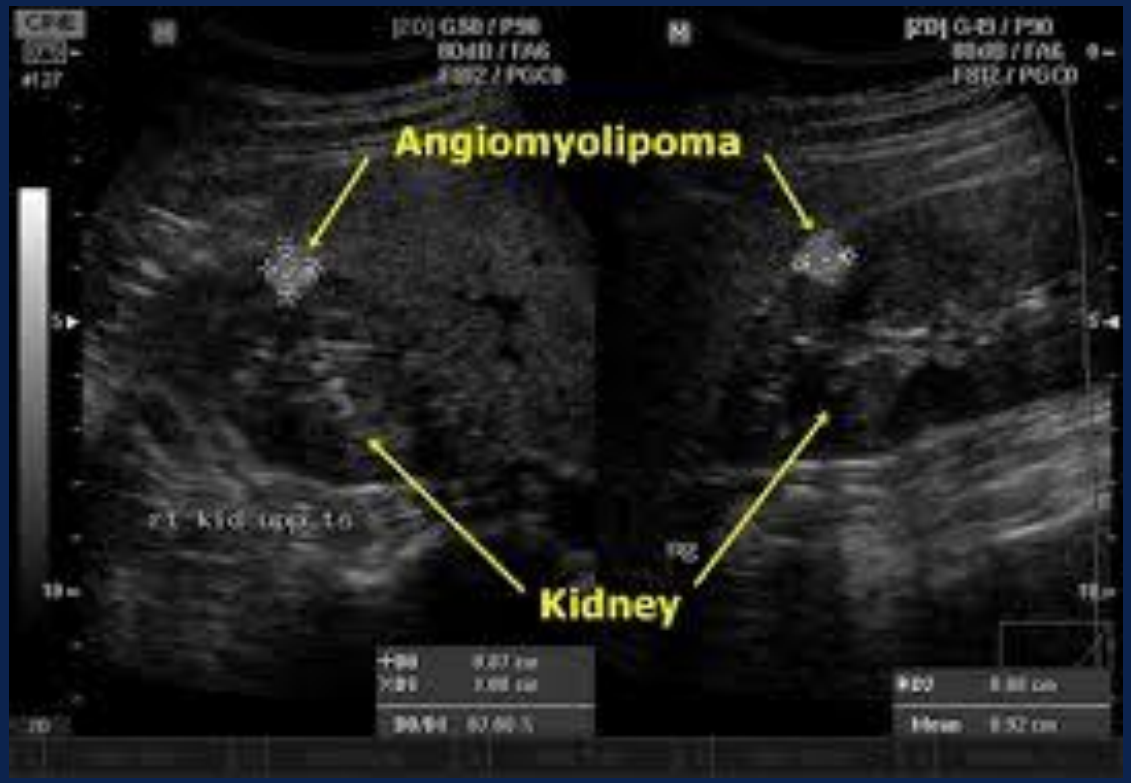
- Rare benign tumor
- Can occur with or without tuberous sclerosis (approximately 45–80% with TS).
- Those with tuberous sclerosis, the lesion tend to be smaller, bilateral & asymptomatic.
- **Tuberous sclerosis**: AD syndrome characterized by mental retardation, epilepsy, adenoma sebaceum and other hamartomas .
- In patients without TS, renal angiomyolipomas can be unilateral and tend to be larger.
- **Grossly**: uncapsulated, yellow to gray lesions, typically round to oval, that elevate the renal capsule, producing a bulging smooth or irregular mass.
- **Microscopically**: composed from blood vessels, mature fat cells & smooth muscle cells.



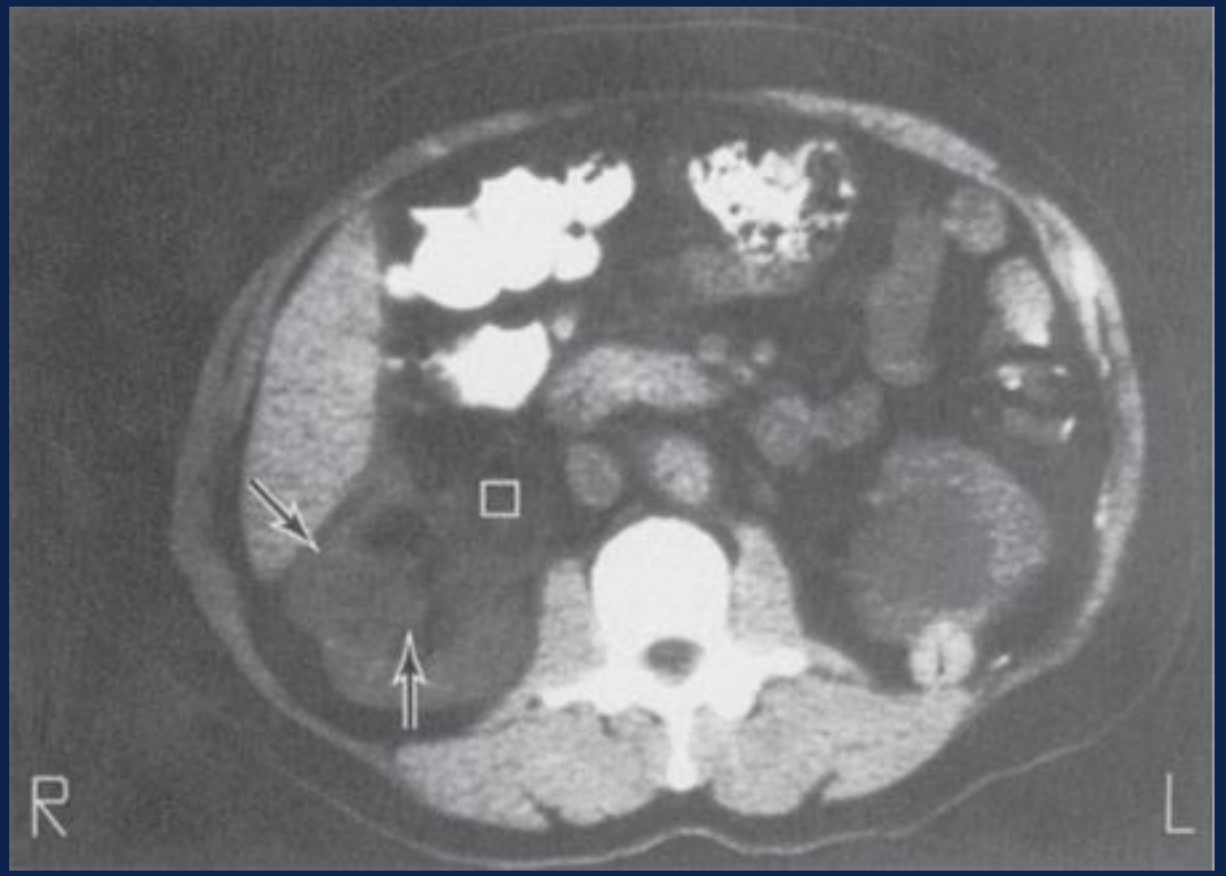
Angiomyolipoma – renal hamartoma

- **Diagnosis**:
  - **US: No acoustic shadow, fat visualized on US appears as very high intensity echoes.**
  - **CT scan: the only tumor that can be differentiated on CT because it contains fat.**
    Fat imaged by CT has a negative density, −20 to −80 Hounsfield units, which is **pathognomonic** for angiomyolipomas when observed in the kidney.
  - **MRI: is particularly useful in distinguishing lipid poor angiomyolipoma which have much lower fat content compared to routine angiomyolipomas, from other solid renal lesions.**

Angiomyolipoma – renal hamartoma



Ultra sound of an angiomyolipoma



Computed tomogram of an angiomyolipoma

Angiomyolipoma – renal hamartoma

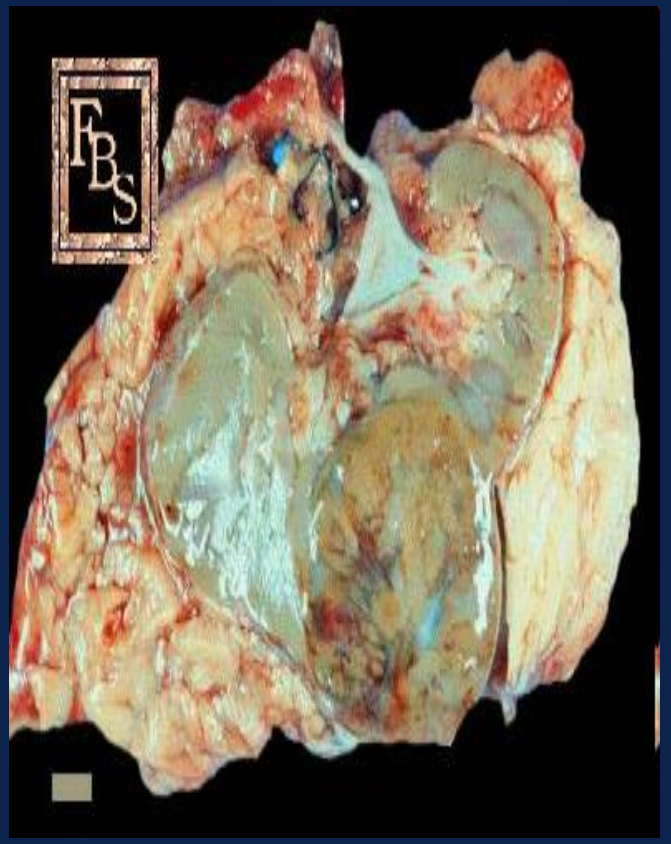
- **Management**:

  - The management of angiomyolipomas historically has been correlated with symptoms:

  - Patients with isolated lesions **<4 cm** be followed up with yearly CT or US.
  - Patients with asymptomatic or mildly symptomatic lesions **>4 cm** should be followed up with semiannual US.
  - Patients with lesions **>4 cm** with moderate or severe symptoms (bleeding or pain) should undergo renal-sparing surgery or renal arterial embolization.
  - In pts with TS , in whom multiple bilateral lesions are present, conservative treatment should be attempted .

Oncocytoma

- **Benign tumor**, **30 % ass. with RCC.**
- Uncommon , accounting for 3 – 5 % of renal tumors .
- ♂ **>** ♀
- Can affect many organs like the adrenal, parathyroid, thyroid, salivary & kidneys.
- Composed of large epithelial cells with finely granular eosinophilic cytoplasm (oncocytes).
- Renal oncocytomas generally occur & are contained within a well-defined fibrous capsule.
- Usually solitary and unilateral, although several bilateral cases & multiple oncocytomas occurring at the same time.



Oncocytoma

- **Origin** : The cellular origin of renal oncocytes has not been fully elucidated, although some early evidence suggested that **oncocytes resemble proximal convoluted tubular cells**, other findings suggest that their origin may be a **precursor stem cel**l or the **intercalated cells of the collecting ducts.**

- High-grade oncocytomas may be intermixed with elements of RCC and can be found as coexisting lesions within the same or opposite kidney.

- **Diagnosis**:
  - Is predominantly **pathologic** because there are no reliable distinguishing clinical characteristics.
  - Gross hematuria and flank pain occur in <20% of patients.
  - No characteristic features of the tumors appear on CT, US, IVU or MRI.
  - Angiographic features of oncocytomas include the "**spoke wheel**" appearance of tumor arterioles, the "**lucent rim sign**" of the capsule, and a homogeneous capillary nephrogram phase.
    →Unfortunately, these findings are not invariable, and similar findings have been reported in pts with RCC.
  - The role of FNA in the preoperative diagnosis of oncocytomas remains controversial and limited due to a lack of characteristic features that distinguish oncocytoma from RCC.

- **Treatment**: Radical or partial nephrectomy is indicated, as for renal carcinoma.

Other Rare Benign Renal Tumors

- **Leiomyomas:**
  - rare small tumors typically found in **smooth-muscle-containing areas of the kidney including the renal capsule and renal pelvis**.
  - Two groups of renal leiomyomas have been described.
  - The more common group comprises cortical tumors that are <2 cm and may be multiple.
  - These tumors are typically found at autopsy and are **not clinically significant.**
  - A larger, commonly solitary leiomyoma has been described, which may cause symptoms and is confirmed pathologically after nephrectomy.

- **Hemangiomas:**
  - **small vascular tumors** occurring in the kidney with a frequency second only to that in the liver among visceral organs.
  - Multiple lesions in one kidney occur in approximately 12% of cases; however, they are rarely bilateral.
  - They can occasionally be the elusive source of hematuria in an otherwise well-evaluated patient.
  - The diagnosis may be determined by CT angiography, MR angiography, or by direct visualization by endoscopy

Other Rare Benign Renal Tumors

- **Renal lipomas:**
  - are very uncommon deposits of mature adipose cells without evident mitosis that arise from the renal capsule or perirenal tissue.
  - They are seen primarily in middle-aged females and, owing to the characteristic CT differentiation of fat, are best detected radiographically on CT scanning.

- **The juxtaglomerular cell tumor:**

- is the most clinically significant member of this subgroup of rare benign tumors because it causes significant hypertension that can be cured by surgical treatment.
- It is a very rare lesion, with <100 reported cases and may have characteristic chromosomal alterations
- Occur more commonly in young adults, more often females in their 20s and 30s and are rarely malignant.
- Originate from the pericytes of afferent arterioles in the juxtaglomerular apparatus and can be shown to contain renin secretory granules.
- They are typically encapsulated and located in the cortical area.
- Symptoms of the “typical” tumors include HTN, hypokalemia, hyperaldosteronism, and high renin.
- Some atypical cases may demonstrate just HTN with normal K+ levels or may even be non functional.
- The diagnosis is confirmed by selected renal vein sampling for renin.
- Although complete nephrectomy was advocated in the past, several recent reports indicate that partial nephrectomy can be equally effective.

Renal cell carcinoma

Renal cell carcinoma - Epidemiology

- Also known as **hypernephroma**.
- Majority of RCC occurs sporadically, Rarely hereditary.
- 2.8% of the adult tumors & constitutes approximately **85%** of all primary malignant tumor of the kidney.
- **5th- 6th decade**.
- male: female 2:1
- More common in **black** men.
- Highest incidence in Scandinavia and North America, lowest in Africa.

Renal cell carcinoma – Risk Factors

Environmental

- **Modifiable Risk Factors**
  - Smoking (↑ 2X)
  - Obesity
  - Hypertension (1.4-2 fold risk)
- **Others**
  - Renal failure and dialysis (>30X)
  - Exposure to asbestos, solvents & cadmium.
  - The analgesic phenacitin.
  - low socio-economic status

Genetic

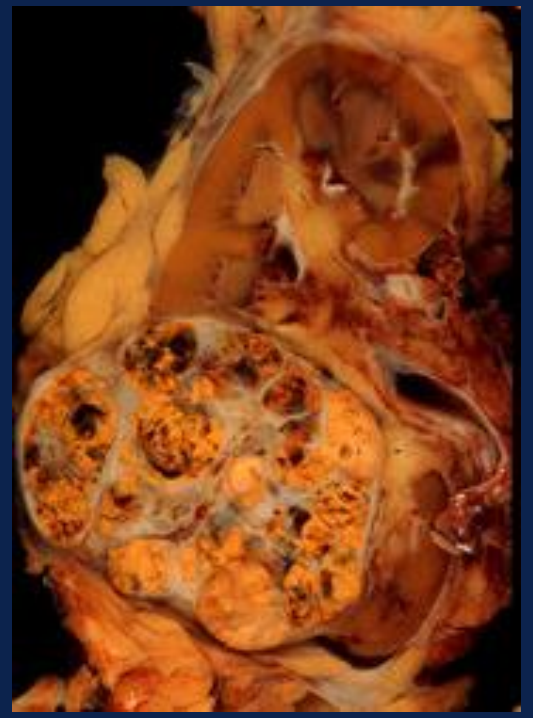
- **Von Hippel–Lindau syndrome**

  Bilateral RCC + Cerebellar hemangioblastoma + Retinal angioma.
- **Hereditary papillary renal carcinoma**

  Multiple bilateral renal tumors with a papillary histologic appearance.

Renal cell carcinoma - Pathology

- RCC originate from proximal renal tubular epithelium in the cortex.
- These tumors occur with equal frequency in either kidney and are randomly distributed in the upper and lower poles.
- Originate in the cortex to grow out into perinephric tissue, causing the characteristic bulge or mass effect that aids in their detection by diagnostic imaging studies.
- **Grossly**: they are yellow-orange “because of the abundance of lipids”.
- RCCs don’t have true capsule but may have a **pseudocapsule** of compressed renal parenchyma, fibrous tissue & inflammatory cells.



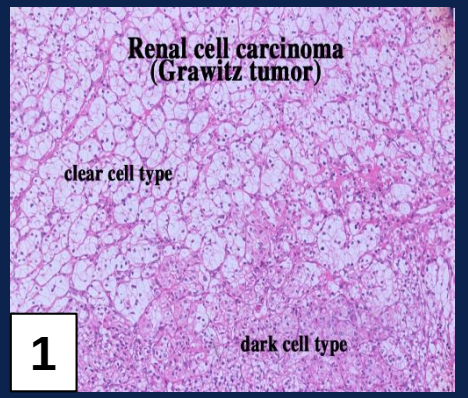
Renal cell carcinoma – Histological Types

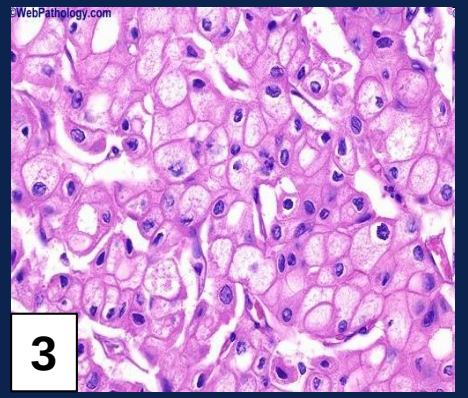
1. **Clear Cell RCC:**
   - The most common type (80%).
   - Under a microscope: appear very pale or clear.

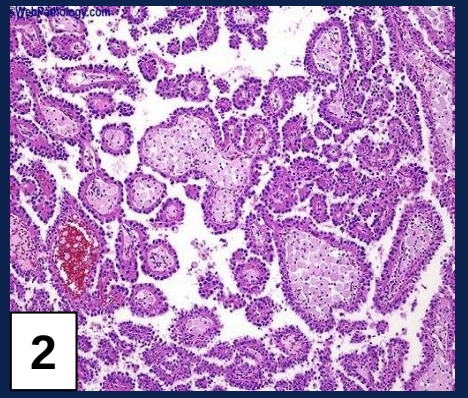
2. **Papillary RCC " chromophilic":**
   - The second most common type (10% to 15%).
   - little finger-like projections (called papillae).

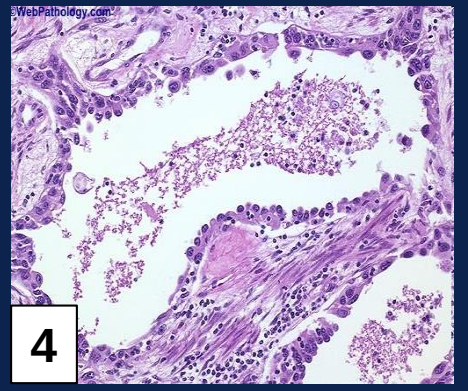
3. **Chromophobe RCC:**
   - The third most common type (5%).
   - Also pale, but are much larger than clear cell RCC.

4. **Collecting Duct RCC:**
   - The rarest type of RCC.
   - The major characteristic is forming irregular tubes.









5. **About 5% of renal cancers are unclassified** because their appearance does not fit into any of the other categories.

Renal cell carcinoma - Pathogenesis

- These tumors are **highly vascular** due to the production of **VEGF.**
- They spread by direct invasion through the renal capsule into perinephric fat and adjacent visceral structures or by direct extension into the renal vein
- Approximately 25–30% of patients have evidence of metastatic disease at presentation.
- The most common site of distant metastasis is the lungs 75%.
- But also, liver, bone “osteolytic lesions”, LN, Adrenal, opposite kidney, brain…

Renal cell carcinoma – Staging

- **T1: limited to the kidney, 7 cm or less in greatest dimension**
  - T1a: limited to kidney <4 cm
  - T1b: limited to kidney 4-7cm
- **T2: limited to kidney >7 cm**
  - T2a: limited to kidney, >7 cm but not more than 10 cm
  - T2b: limited to kidney, >10 cm
- **T3: tumor/tumor thrombus extension into major veins or perinephric tissues, but not into ipsilateral adrenal gland or beyond Gerota's fascia**
  - T3a: spread to renal vein or it's segmental branches or perirenal fat or renal sinus fat.
  - T3b: spread to infra diaphragmatic IVC
  - T3c: spread to supra diaphragmatic IVC or invades the wall of the IVC
- **T4: involves ipsilateral adrenal gland or invades beyond Gerota's fascia**

N

- N0: no nodal involvement
- N1: metastatic involvement of regional lymph node/s

M

- M0: no distant metastases
- M1: distant metastases

Renal cell carcinoma – Grading

- **Fuhrman Nuclear Grade – depends on:**
  - ➢ Size and shape of the nucleus.
  - ➢ Number and size of nucleoli
  - ➢ Chromatin clumping

Grade	Nuclei		Nucleoli
	Size (μm)	Shape	
1	10	Round, unifrom	Inconspicuous or absent
2	15	Slightly irregular	Evident at high power (×400 magnification)
3	20	Obviously irregular	Prominent, large at low power (×100 magnification)
4	>20	Bizarre, often multilobed	Heavy chromatin clump

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❖ **grade I carries the best prognosis and grade IV the worst.**

Renal cell carcinoma - Clinical Presentation

- **Classic triad (only in 5%-10% of patients)**
  - **Hematuria** (alone in 50% of patients).
  - **Flank pain** (alone in 40% of patients).
  - **Palpable abdominal mass** (alone in 30% of patients).

- **Systemic symptoms (in 25% of patients)**

  Anemia, Fatigue, Cachexia, Wt. Loss, Hypercalcemia, Hepatic Dysfunction

- **Paraneoplastic Syndrome (in 10–40% of patients)**

  - RCC is associated with a wide spectrum of paraneoplastic syndromes including **erythrocytosis, hypercalcemia, hypertension, and nonmetastatic hepatic dysfunction "Stauffer's Syndrome" .**

Renal cell carcinoma - Diagnosis

General: History, Physical examination.

Laboratory studies: CBC, LFT's, alkaline phosphatase, BUN, creatinine, urinalysis.

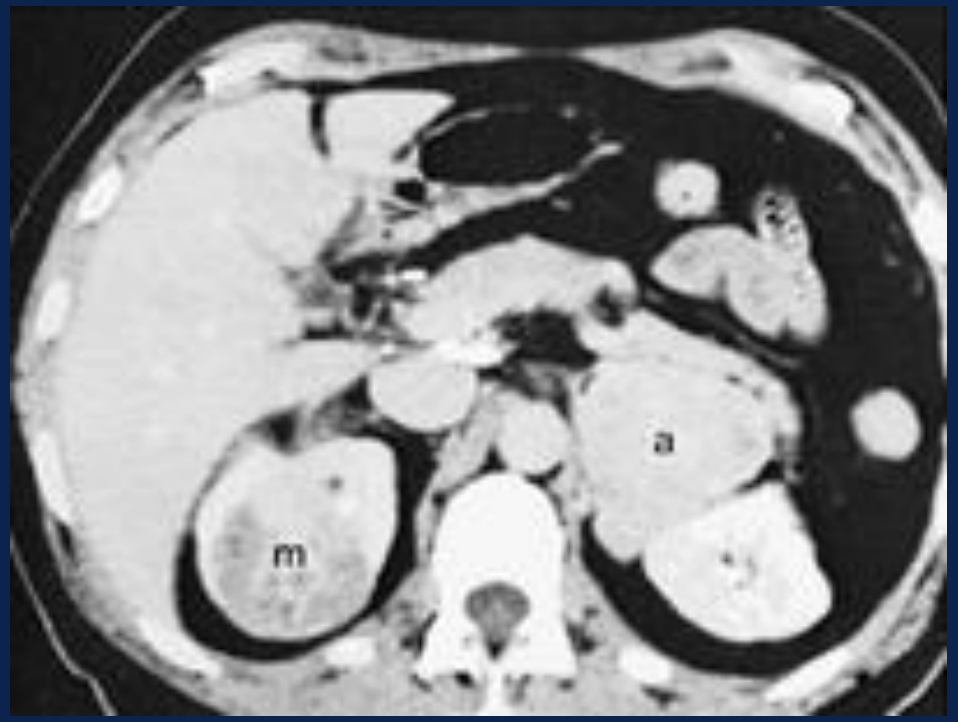
Radiographic studies:

- **X-Ray** for **calcification**: central calcification 80% malignant, peripheral calcification is 80% benign.
- **U/S:** Excellent in distinguishing **cystic from solid** masses (98% accurate!)
- **IV Urography:** Starting point for hematuria evaluations and function of contralateral kidney
- **CT with contrast:** **The diagnostic method** + useful **for staging** the tumor by demonstrating the invasiveness of the tumor.
- **MRI:** excellent demonstration of solid renal masses and is image test of choice to demonstrate **extent of vena caval involvement** with tumor. Useful in patients with renal insufficiency

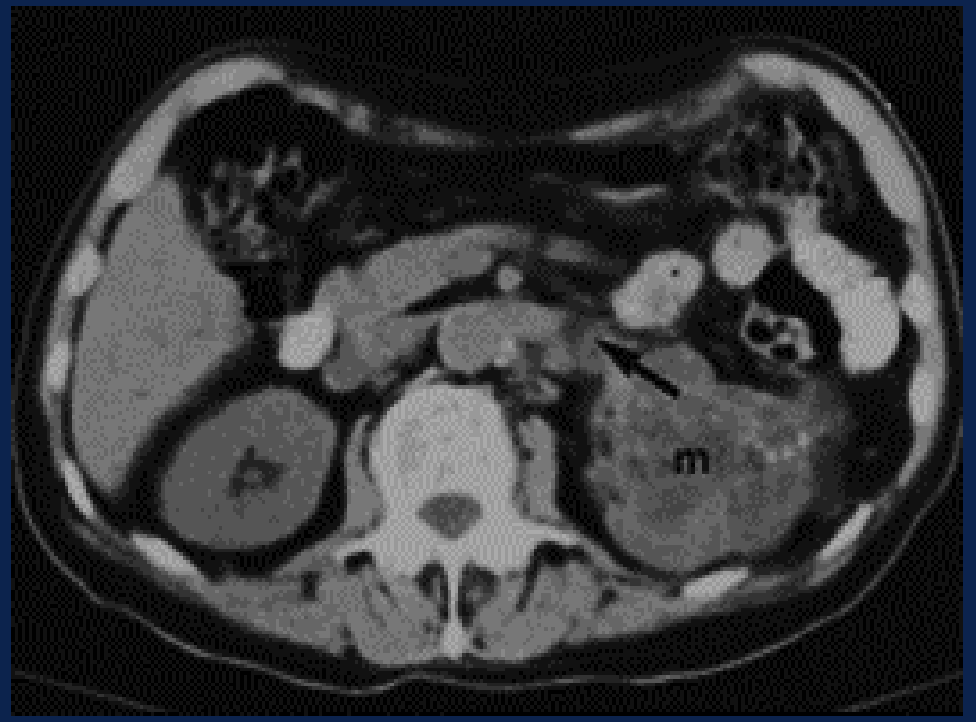
Renal cell carcinoma – Metastatic Work-Up

- Chest X-ray or Chest CT
- CT/MRI scan of abdomen or pelvis
- Radionuclide bone scan with plan films (for elevated alkaline phosphatase or bone pain).

Renal cell carcinoma



CT scan demonstrates a right renal carcinoma (m) with a large contralateral adrenal metastasis (a).

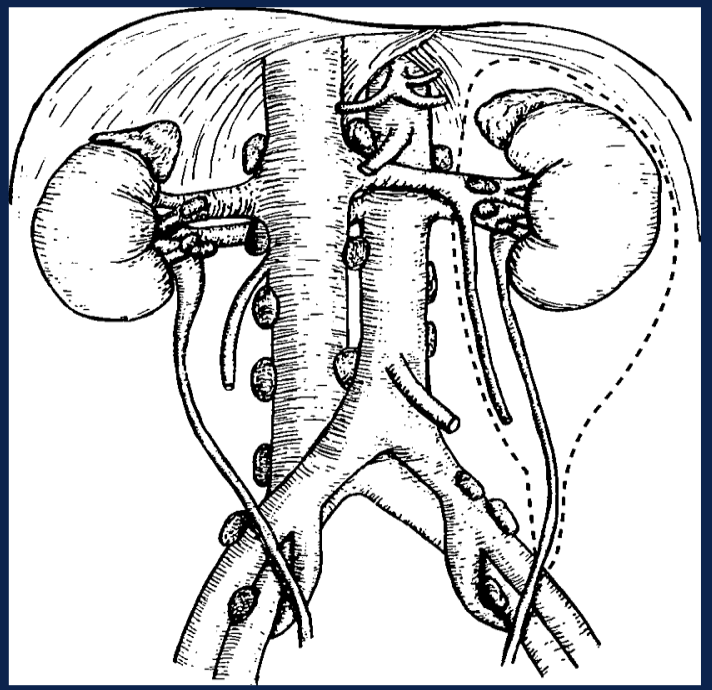


CT scan shows large left renal mass with calcification (m) invading the left renal vein (arrow).

Renal cell carcinoma – Management

❖ Appropriate therapy depends almost entirely on the stage of tumor at presentation and therefore requires a thorough staging evaluation.

- **Localized tumor T1-T3a :**
  - **Radical or partial nephrectomies** are the primary treatments for localized RCC.
  - Radical nephrectomy “removal of the kidney + proximal half of the ureter + gerota’s fascia including the ipsilateral adrenal gland + hilar LN (up to the area of transection of the renal vessels)”.



Boundaries of a left radical nephrectomy. Dotted line represents both the surgical margin and Gerota’s fascia.

Renal cell carcinoma – Management

- Para aortic LN dissection has not been beneficial and is not recommended.
- Radical nephrectomy can be done by open surgery or laparoscopic.
- The likelihood of local recurrence after radical nephrectomy is 2–3%

Renal cell carcinoma – Management

- **Indication of partial Nephrectomy :**
  - 1) small lesion <4 cm.
  - 2) Single kidney.
  - 3) bilateral involvement of the kidneys.
  - 4) DM, or renal insufficiency.

  - In general, RCC is resistant for the radiotherapy & chemotherapy.

Renal cell carcinoma – Management

- **For more advanced disease:**
  - The treatment of choice is by Immunotherapy.
  - This is done by the administration of IFN-alpha SC 3-5 day/week
  - Combination of IL-2+INF-alpha , 30% response rate , this is the best convenential method available.

Renal cell carcinoma – Follow up

- Patients with stage T1 disease need less follow-up, with yearly chest x-rays , liver and renal function tests.

- Those with stage T2 or T3 disease require more frequent follow-up of at least 3-month or 6-month intervals in the early postoperative period.

- Repeat CT scans of the abdomen should also be obtained, especially in those who have undergone partial nephrectomy, to rule out local recurrence.

- Patients with metastatic disease who are not undergoing therapy need continued follow-up to provide appropriate supportive care.

Renal cell carcinoma – Prognosis

- **depends on stage: 5 – years survival :**
  - T1 : 84 %
  - T2 : 74 %
  - T3 : 53 %
  - T4 : 8 %

Transitional cell carcinoma

Transitional cell carcinoma

- About **5% to 10%** of all kidney tumors.
- Also known as **urothelial carcinomas**.
- Under the microscope, looks like bladder cancer cells and act very much like bladder cancer.
- Linked to cigarette smoking and occupational exposures to certain cancer-causing chemicals.

- **The signs and symptoms** of transitional cell carcinoma are typically the same as with the signs and symptoms of kidney cancer - blood in the urine and, sometimes, back pain.

- **Treated by** surgically removing the entire kidney and the ureter, as well as the section of the bladder where the ureter is attached. Chemotherapy and radiotherapy are often used in addition to surgery, depending on how much cancer is found.

Thank You ☺