# RENAL TUMORS

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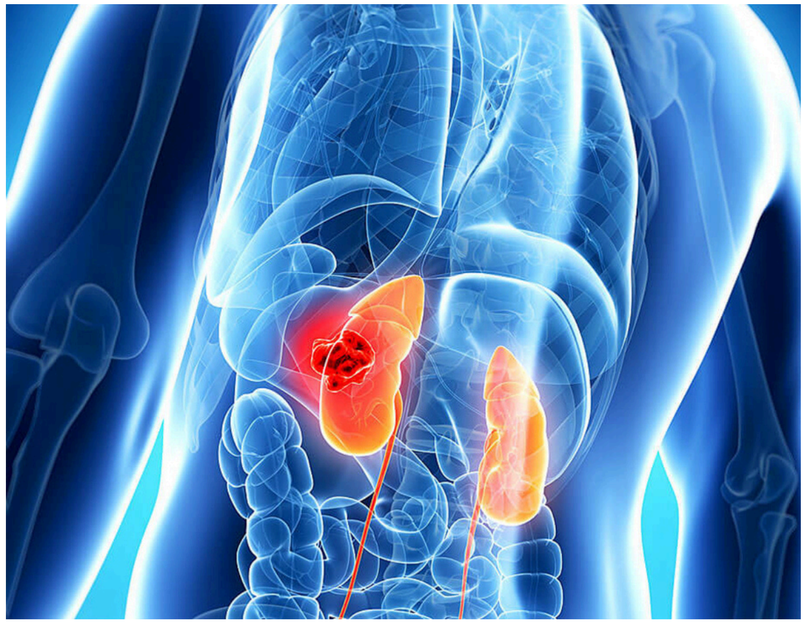
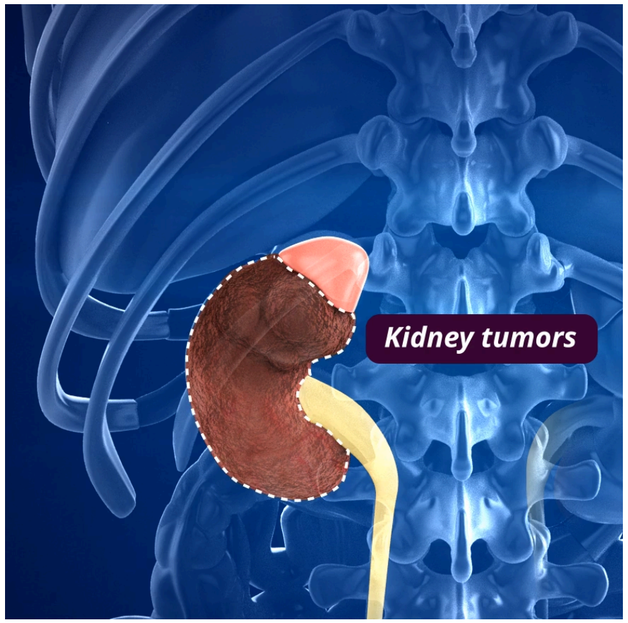
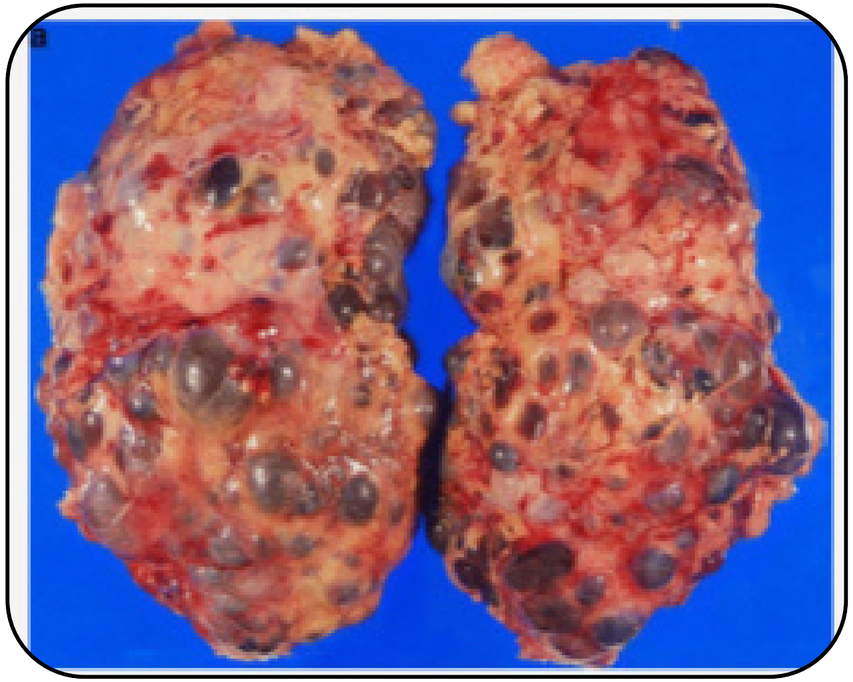
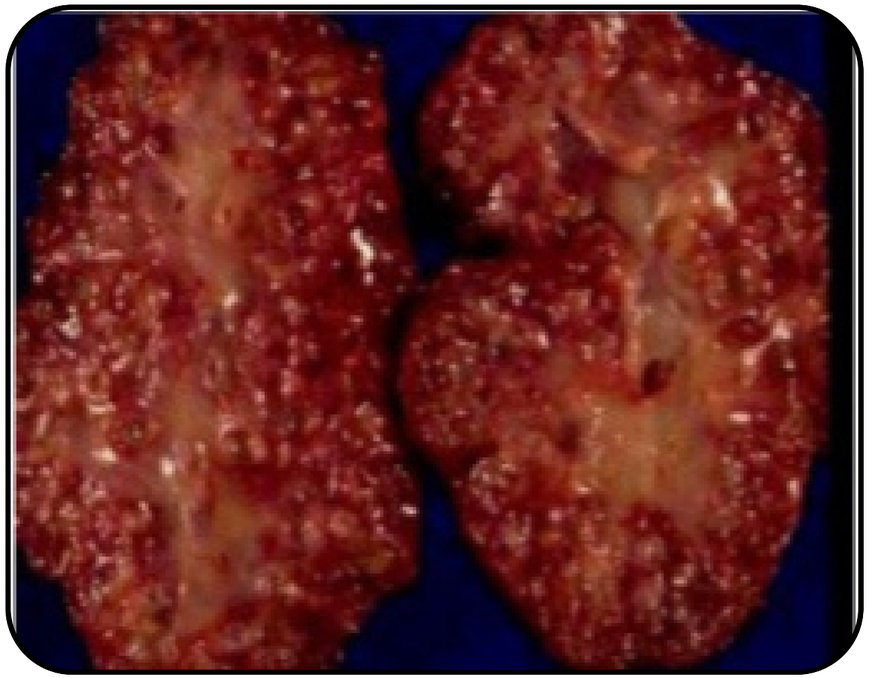




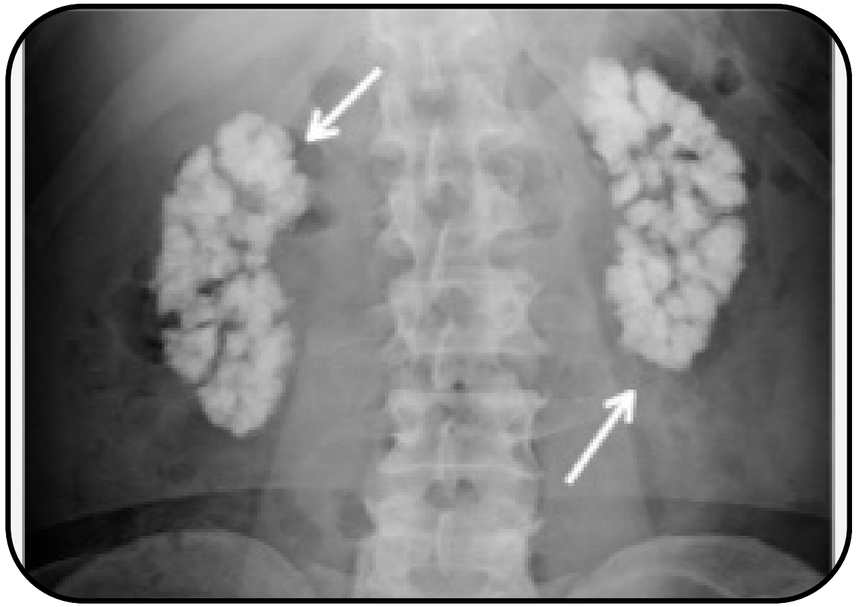
# RENAL CYSTIC DISEASES



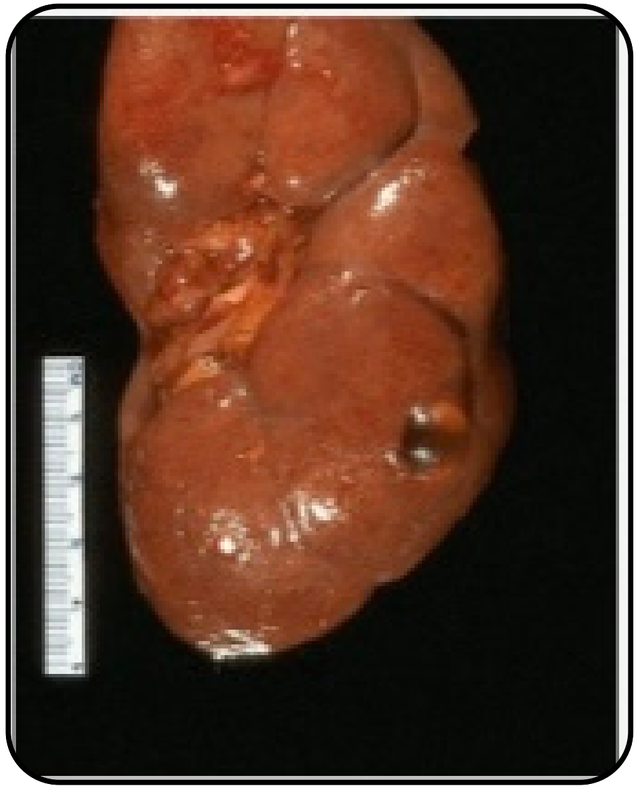
1- Autosomal dominant polycystic kidney disease (ADPKD)
progressive bilateral disease leading to hypertension and renal failure associated with hepatic cysts and cerebral aneurysms



2- Autosomal Recessive Polycystic Kidney Disease (ARPKD)
massive kidneys and early renal failure in children ,associated with hepatic disease and abnormalities in the blood vessels of the brain and heart.



**3- medullary sponge kidney**
**cystic dilation of the collecting ducts,may present with hematuria, UTIs or nephrolithiasis , and can be asymptomatic .**
**thought to be associated with hyperparathyroidism and parathyroid adenoma**



**4- simple Renal cysts**

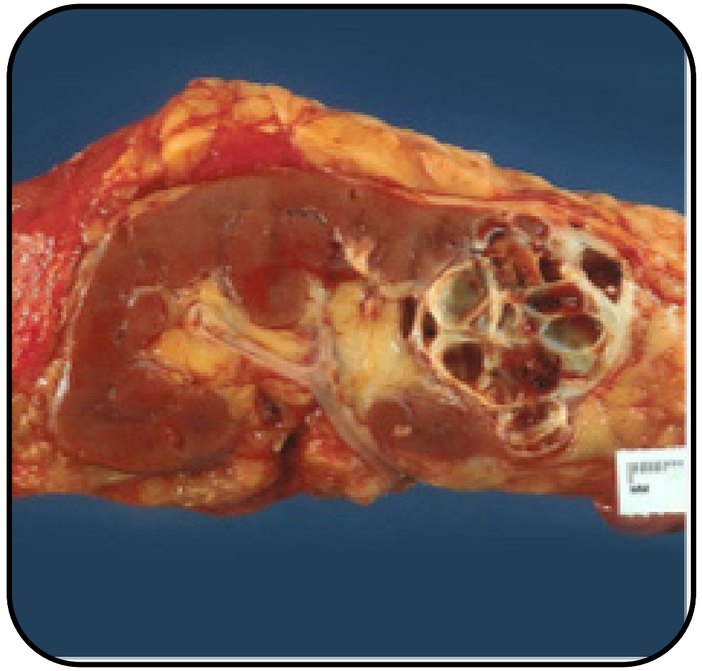
# Cysts associated with systemic diseases

1.Von Hippel-Lindau Syndrome (VHLS)

2.Tuberous Sclerosis (TS)

3.Acquired Simple cysts, acquired cystic renal disease

4.Malignancy Cystic renal cell carcinoma (RCC)



# Simple Renal Cyst

**are fluid-filled sacs that develop within the kidneys.**

- **Simple Cysts are The most common (70%)**
- **>50% in patients aged >50y**
- **It is defined as round pouch of smooth thin-walled**

**tissue or closed pocket that is usually filled with fluid.**

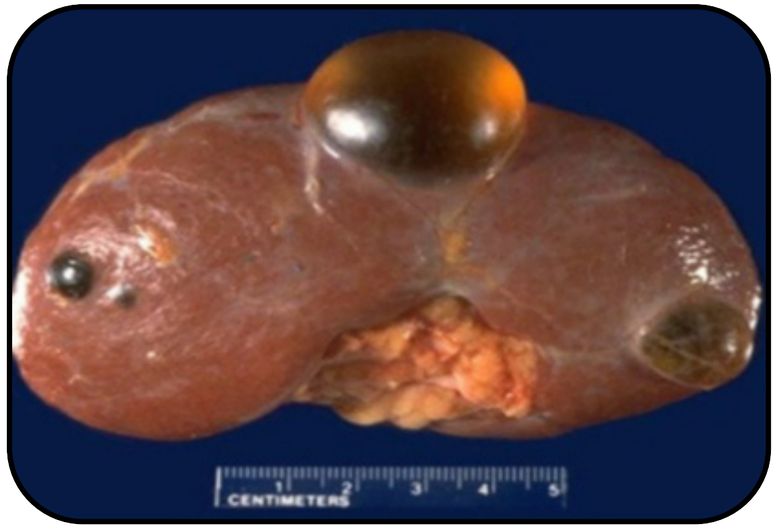
- **usually incidental finding on abdominal imaging**

**The cyst:**

**Contains Clear Fluid & may contain altered blood**

**Lined by Flat epithelium.**

**Surrounded by Fibrous tissue**



# SYMPTOMS OF SIMPLE SYST

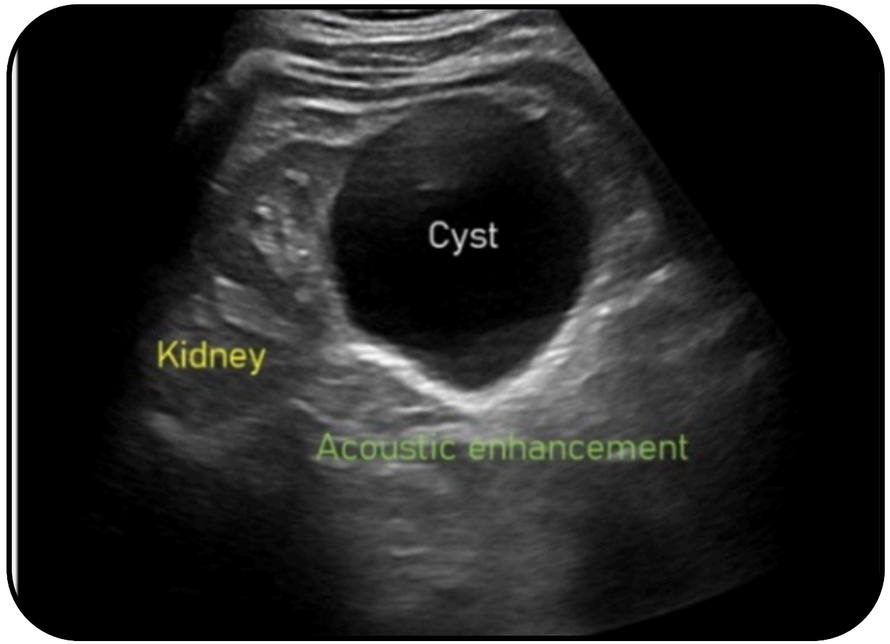
- Usually ASYMPTOMATIC.
- Dull aching PAIN in the loin due to stretch of renal capsule.
- A SWELLING may be felt in the loin.
- Clinical picture of complications (e.g. Hematuria ,pyuria...etc.)

# COMPLICTION

- hemorrhae
- rupture
- pressure on ureter >> hydronephrosis
- infection
- calcification

# Diagnosis

**1- ultrasound**
**very helful**
**Features of simple cyst are:**
**1- Anechoic ( echo lucent ;absence of echoes )**
**2- Posterior acoustic enhancement**
**3- round/oval shape**
**4- no septation, no calcification**
**5- Sharply marginated thin smooth wall**



**2- IVU**

- **simple cyst signs:**

**1- smooth amputated calyx**

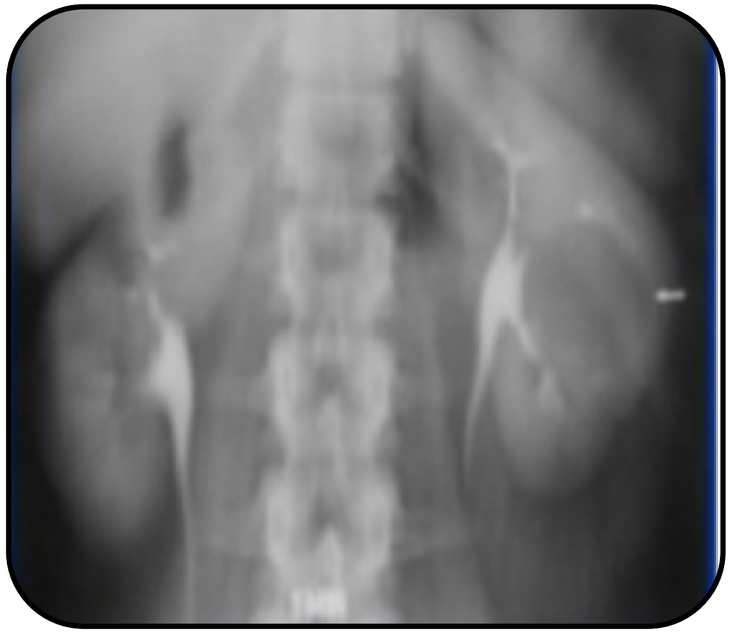
**2-”beak” “claw” sign resulted from stretching of calyces by cyst**

**3- Renal angiography (not used)**

**to differentiate between cyst & tumor :**

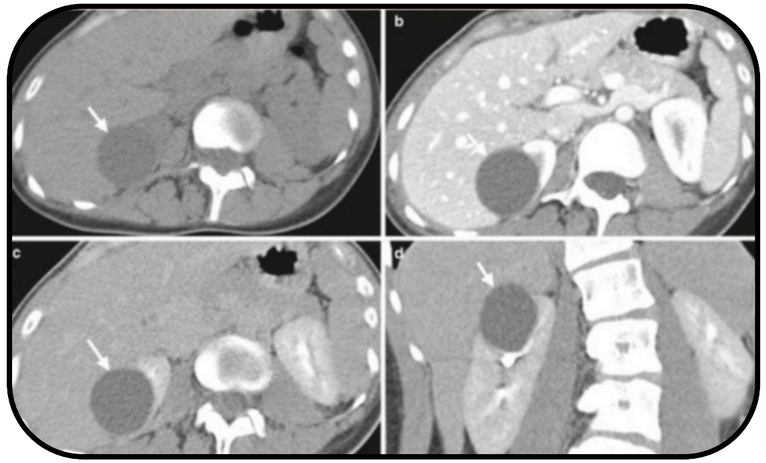
**cyst >> avascular**

**tumor >> hypervascular**



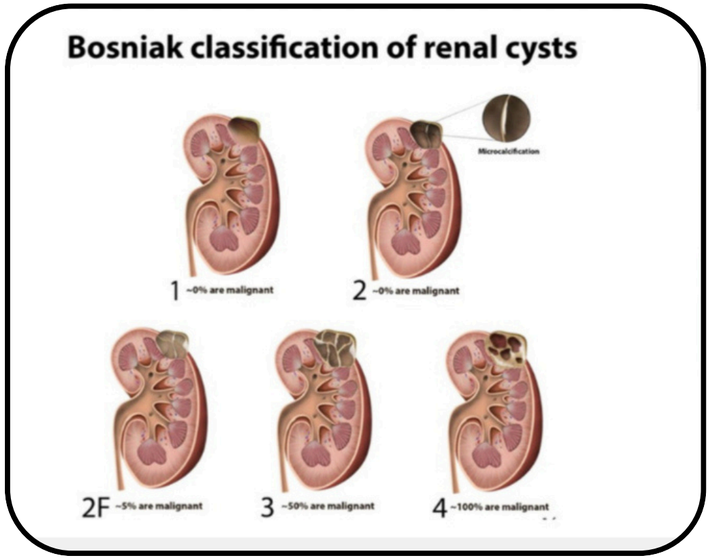
# 4- CT

## Distinguishes fluid – only filled cysts from solid masses



## Bosniak classification of renal cyst


| *Bosniak stage* | Risk of malignancy (%) | Features |
|---|---|---|
| I | 0 | Thin wall without septa or solid components.<br>No internal enhancement. |
| II | 0 | Few (≤3) thin septa. It may show minimal enhancement of the septa.<br>Hyperintense on T1 without enhancement. |
| IIF | 5 | Multiple (>3) thin septa.<br>Smooth mild thickening (3 mm) of the wall or septa.<br>It may contain minimal enhancement of the septa. |
| III | 50 | Thickened (>3 mm) wall or septa with enhancement.<br>Irregular wall or septa with enhancement. |
| IV | 90 | Soft tissue enhancing mass independent from the wall. |

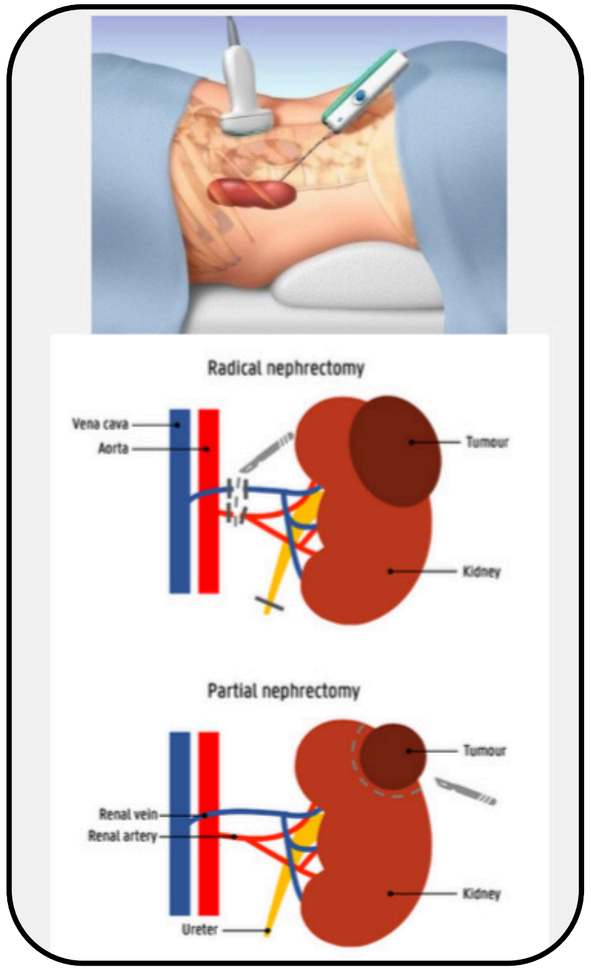


# Treatment

**Usually no need for treatment but follow up is required if too much pressure or the cyst affects kidney function; (hydronephrosis):**

- **Aspiration and sclerotherapy by 95% alcohol**
- **Excision**

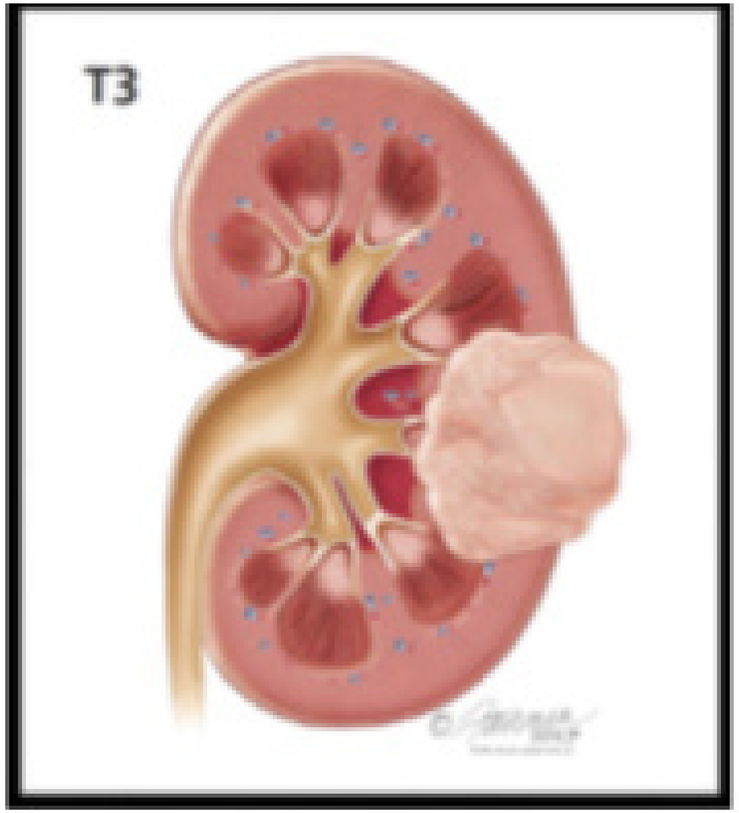
**Atypical cyst (hemorrhagic , thick wall or cloudy fluid) : PNA of content for analysis (Blood , high fat content or +ve cytology gives high suspicion of malignancy)**
**Excise the extrarenal portion of cyst .**
**Partial nephrectomy may be considered**



# Renal Tumors

**BENIGN RENAL TUMORS**

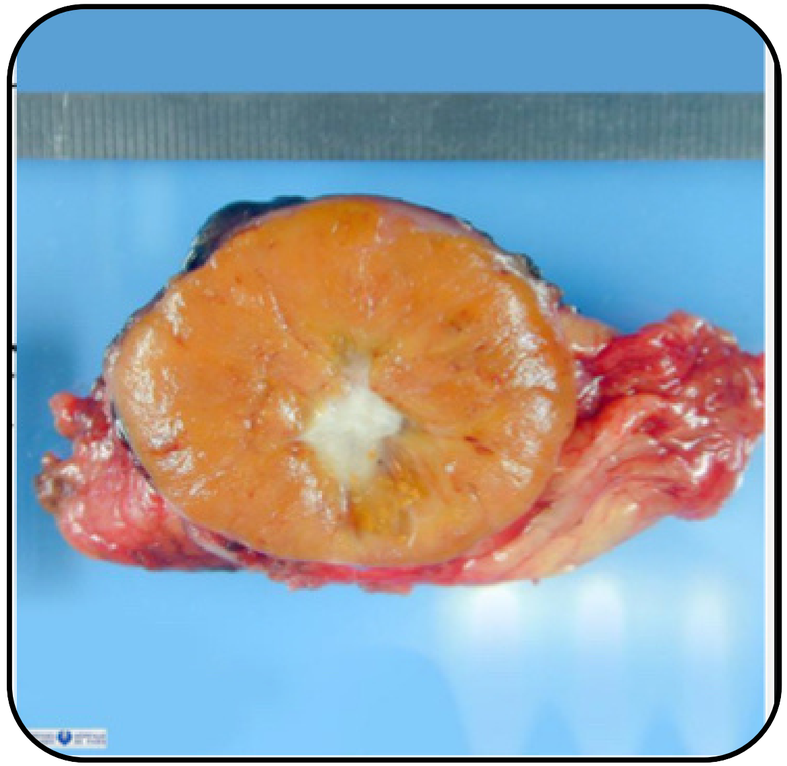
- **1- Oncocytoma**
- **2- Angiomyolipoma**
- **3- Papillary adenom**



# Oncocytoma

**BENIGN RENAL EPITHELIAL NEOPLASM that arise from the intercalated cells of collecting ducts that comprises approximately 5-9% of renal tubular epithelial tumors**

- **more common in males .**
- **They occur with RCC in 7-32% of the cases.**
- **Presentation :**
- **83% present as an incidental finding or with flank pain or hematuria**
- **Histological:**
- **organized eosinophilic cells originating from intercalated cells of collecting duct**



## 1-Gross Appearance:

**Mainly unilateral, can be multiple and bilateral Well defined fibrous capsule with 10-20% extending into the perinephric fat**

## 2 Size:

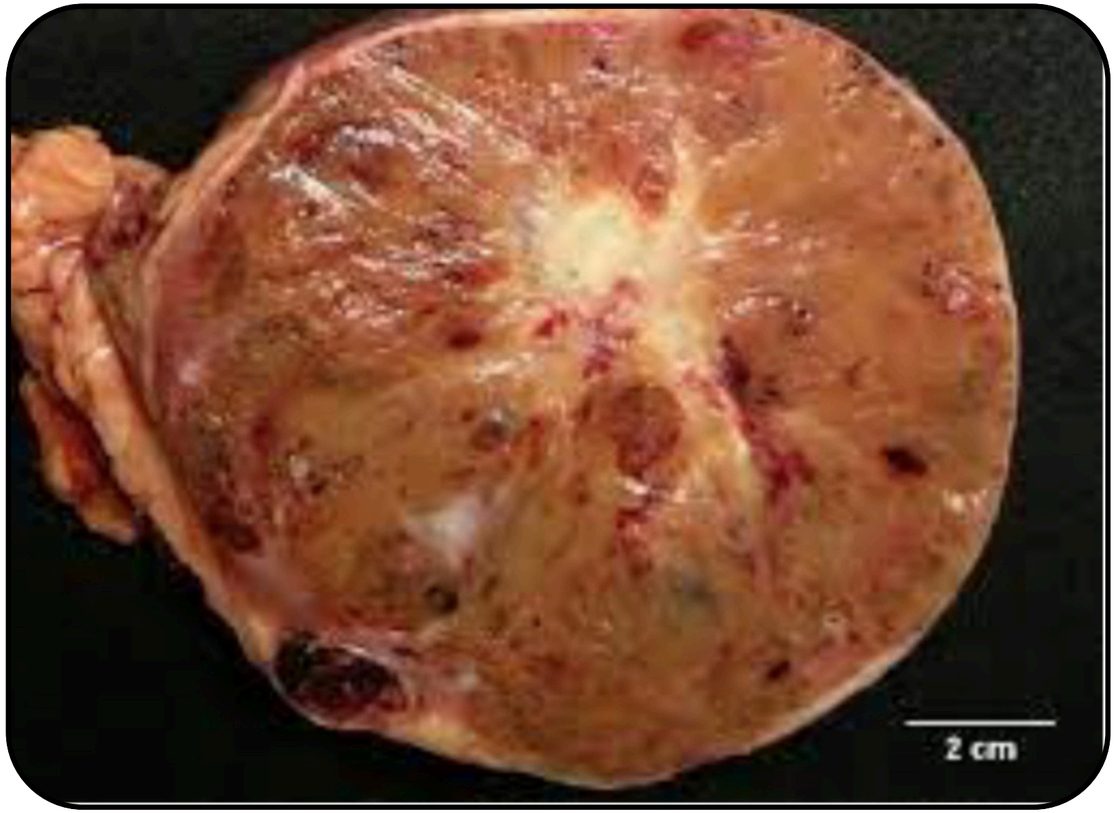
**Mean size 4-6cm**

## 3 Color and Texture:

**Brown with a central scar but no necrosis, well defined defined fibrous capsule**

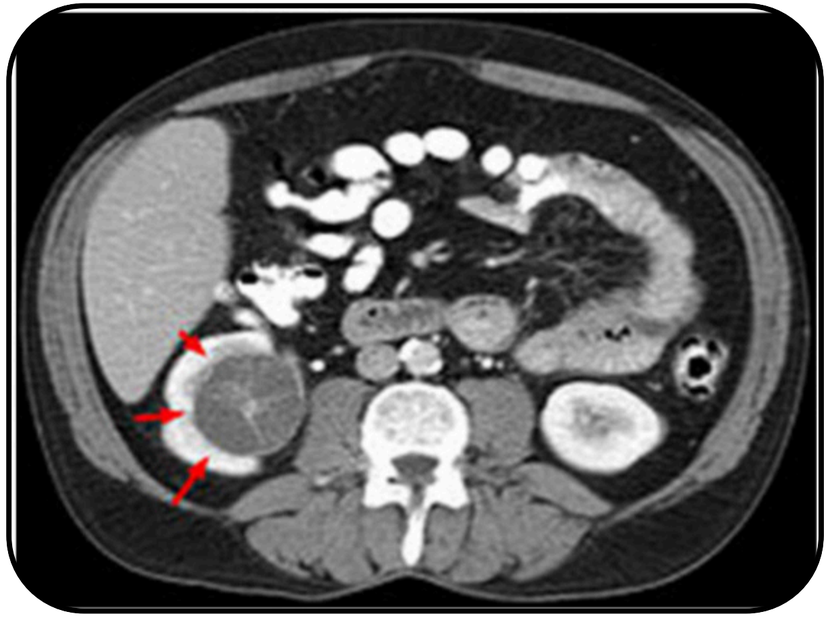
## 4 Shape:

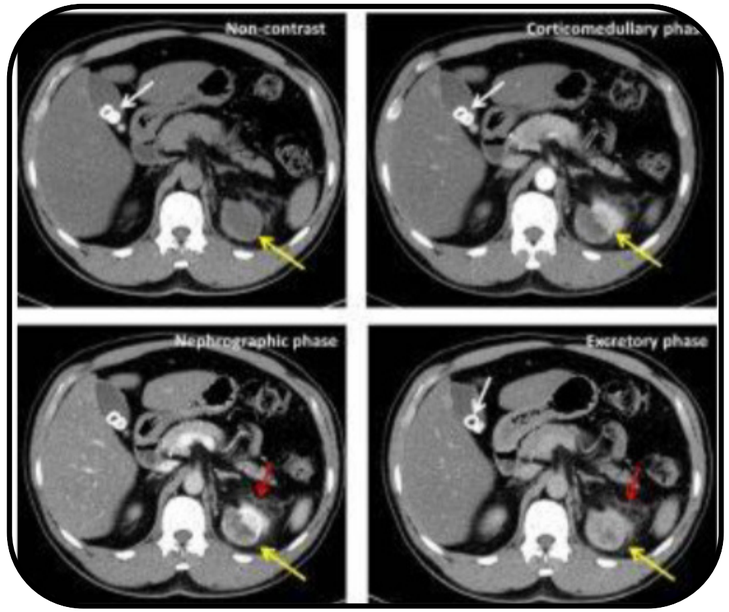
**Spherical, solitary lesion**



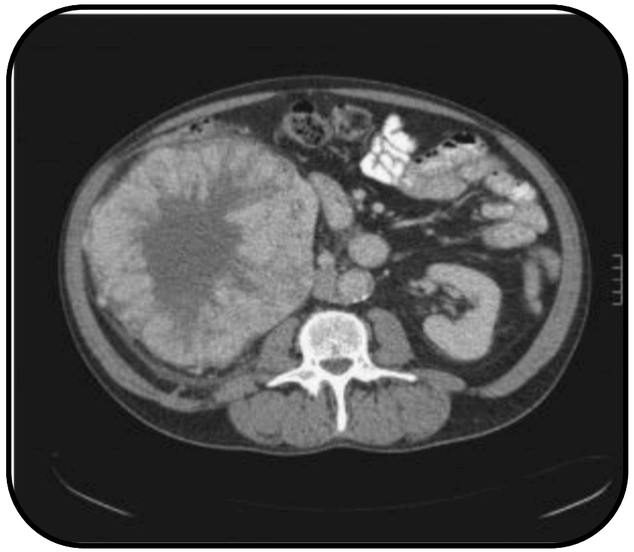
# Diagnosis

- oncocytoma cannot often be distinguished radiologically from RCC , it may coexist with RCC.
- rarely they exhibit a SPOKE WHEEL PATTERN on CT scanning






# Management



## 1- Huge Oncocytomas:

- **Partial or radical nephrectomy**

## 2- Small Oncocytomas:

- **Minimally invasive techniques :**
- **1- Radiofrequency ablation (RFA). (less than 3-4cm )**
- **2- High intensity focused ultrasound (HIFU) Non invasive therapeutic technique that uses non ionizing ultrasonic wave**

# Angiomyolipoma

- Rare benign clonal neoplasms , less than 1%

- F>M

- 20% associated with tuberous sclerosis ,TS syndrome (autosomal dominant)characterized by mental retardation ,epilepsy , adenoma sebaceum , and other hamartomas (mean age 30) .

- frequently , AMLs are multifocal and bilateral.

- Solitary AMLs are more frequently found in the right kidney.

## Presentation:

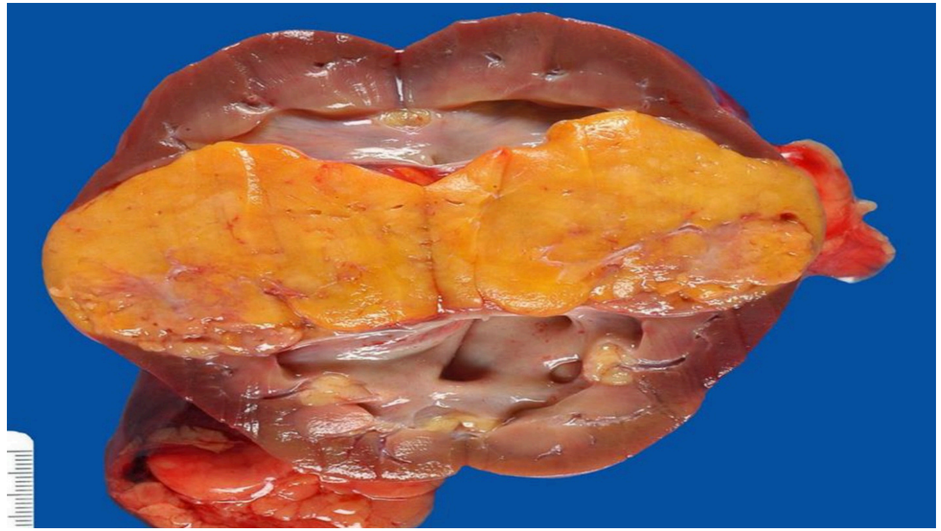
• incidental finding >50% on US and CT, they may present with (Lenk’s triad) :
- flank pain

-palpable mass or painless hematuria.

-Massive and life-threatening retroperitoneal bleeding occurs inup to 10% of cases

• (Wunderlich's syndrome) ; spontaneous, nontraumatic renal hemorrhage confined to the subcapsular and perirenal space. It may be the first manifestation of a renal angiomyolipoma (AML), or rupture of renal artery or intraparenchymal aneurysm.

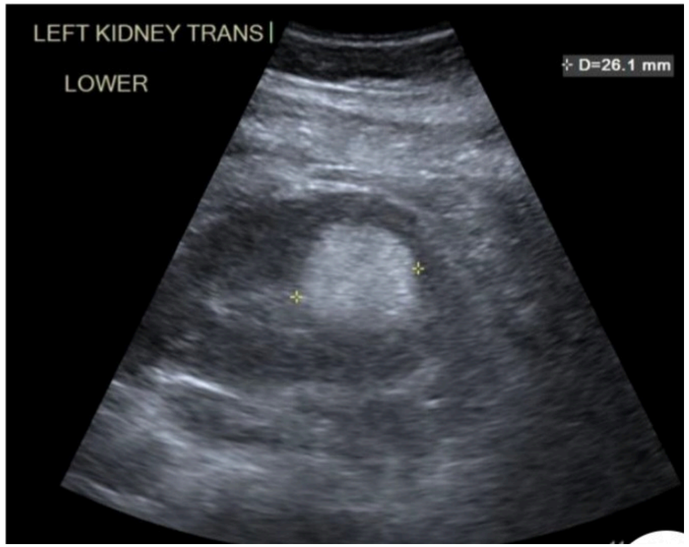
AML is composed of PERIVASCULAR epithelioid cells (PEC)



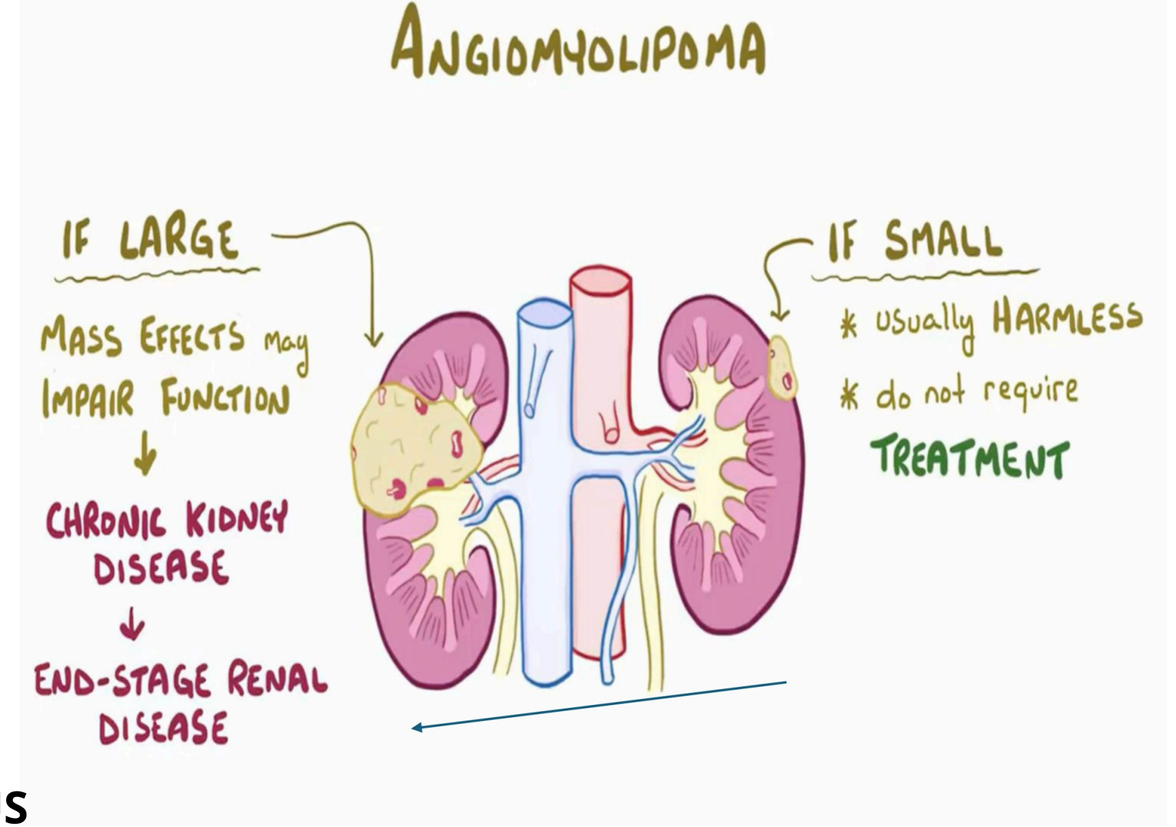
• ➢ Gross appearance:
o non-capsulated
o oval, yellow to gray tumor
o a well circumscribed lump of fat

## Investigation

Ultrasound
* Homogenous
* Well- defined
o characteristic hyperechoic ;
brightechopattern (fat)




This does not cast an ‘acoustic shadow’ beyond,helping to distinguish an AML from a calculus.




## Management

**asymptomatic AMLs**
**if size <4 cm**
**Follow up with serial US**

**asymptomatic AMLs or >4 cm**

- **nephrectomy or selective renal artery embolization**

- **in patient with TS in whom multiple bilateral lesions are present , conservation treatment should be attempted**

# Renal adenoma

**Renal adenomas are the most common form of benign renal tumors**
**Their cause is unknown.**
**Incidence increase with age**

**It is traditionally classified into three distinct types :**
**1. renal papillary adenoma (the most common renal epithelial neoplasms)**

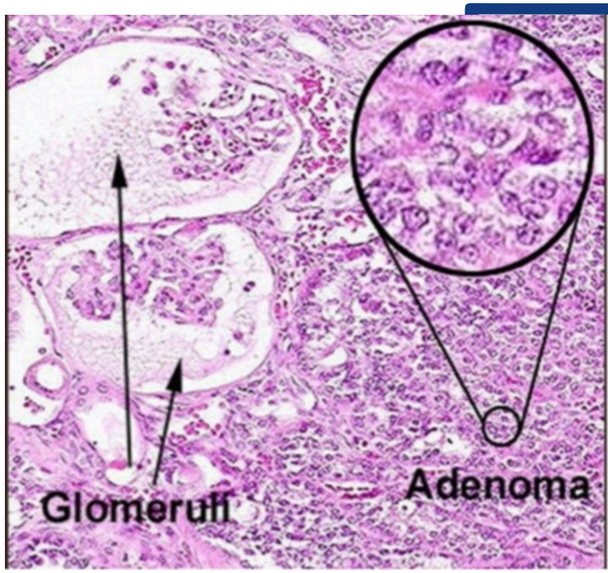
**2. renal tubular adenoma**

**3. alveolar renal adenoma**

7 - 23 % at autopsyo
M:F 3:1

- Cytogenetics : which may exhibit trisomy of chromosome 7 & 17 Rarely symptomatic

Grossly. : Solitary cortical solid ,non-capsulated pale_yellow-gray discrete nodules < 2 cm
(Majority_are solitary_ but may be multifocal)




Adenoma cells look much like low-grade RCC cells under a microscope. In fact, while they are considered benign, there is presently no known cellular classification to differentiate them from RCCs. Many researchers and physicians regard them as early stage precancers, to be treated accordingly. Controversy aS to whether this represent benign or pre-malignant neoplasm.Incidental finding on CT

Histologically :
organized cells with no or little atypia

## Malignant renal tumors

1. Renal cell carcinoma

2. Nephroblastoma (wilm's tumor)

3. Neuroblastoma

### RCC (hypernephroma/ Grawitz's tumort)

-The most common malignant renal tumor (85% of renal malignancies).
-Constituting 2-3% of all cancer deaths.
-The most lethal of all urological tumors, approximately 40% of patients dying of the condition.

# Risk factors

-Smoking cigarettes doubles the risk of developing kidney cancer

-Gender ; Men are 2 to 3 times more likely to develop kidney cancer than women.

-Race : Black people have higher rates of kidney cancer

-Age ' Kidney cancer is typically found in adults and is usually diagnosed between the ages of 50 and 70.

-Chronic kidney disease.

-Nutrition and weight Research has often shown a link between kidney cancer and obesity vitamins A,C,_E, and fruit and vegetable consumption are protective

- High blood pressure- Overuse of certain medications esp. Painkillers containing phenacetin Diuretics and analgesic pain pills, such as aspirin, acetaminophen, and ibuprofen, have also been linked to kidney cancer

-Occupational with asbestos and cadmium exposure

-Long-term dialysis

- Family history of kidney cancer People who have first-degree relatives with kidney cancer have an increased risk

-Anatomical risk factors include polycystic and horseshoe kidneys.

**Origin: RCC is adenocarcinoma of the renal cortex,**
**arise from Epithelium of proximal convoluted tubules.**

**Site**
**Usually unilateral mainly from upper pole**

**Bilateral tumor (1-2%)**
**Von-Hippel Lindau disease**
**hereditary papillary renal carcinoma**

**Histological classification:**
**Clear cell renal cell carcinoma (Conventional) (70-80%): arises from the proximal tubule: highly vascular , associated with deletion of chromosome 3p and/or mutations of the VHL gene.**

**Von Hippel-Lindau_(VHL)_syndrome: Half of individuals with this autosomal dominant syndrome, characterized by pheochromocytoma, renal and pancreatic cysts, and cerebellar hemangioblastoma develop RCC, often bilateral and multifocal.**
**Patients typically_present in third,_fourth,_or fifth decades**

Chromophobe RCC (5%): rare tumor arises from the cortical portion of the collecting duct , associated with Birt-Hogg-Dubé syndrome (BHD) , associated with skin tumor and lung cyst (is a result of loss of chromosome 17 )

-papillary RCC (10-15%): also has an autosomal dominant familial component characterized by trisomy 7 and 17 , (papillary, tubular, and solid variants; 40% multifocal )

-Collecting_duct_(Bellini): rare:; young patients: poor prognosis

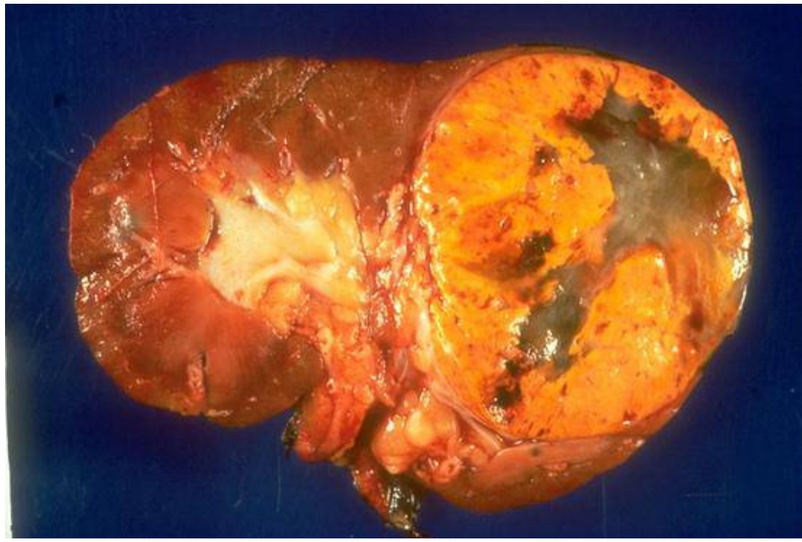
-Medullary cell: rare: arises from calyceal epithelium: young, Black, sickle-cell sufferers; poor prognosis

-Tuberous sclerosis complex_(TSC)_is a genetic condition associated with changes in the skin, brain, kidney, and heart. People with TSC also have an increased risk of developing angiomyolipomas of the kidney and kidney cancer

sarcomatoid : infiltrative, poorly differentiatedvariant of any type, it is carcinoma

# Macroscopic picture:

Mass (mainly from upper pole of kidney) infiltrating ledge area of hemorrhage ,necrosis and scarring



-golden_yellow color due to high_ lipid_content
They are usually circumscribed by pseudocapsule of compressed tissue

-7-20% are multifocal,10-20% contain calcification and 10-25% contain cysts or are predominantly cystic.

-usually invades the pelvis early. capsule late

# Spread

-By Direct extension to adrenal gland (7.5% in tumors >5 cm), through the renal capsule, into the renal vein , inferior vena cava (IVC), right atrium

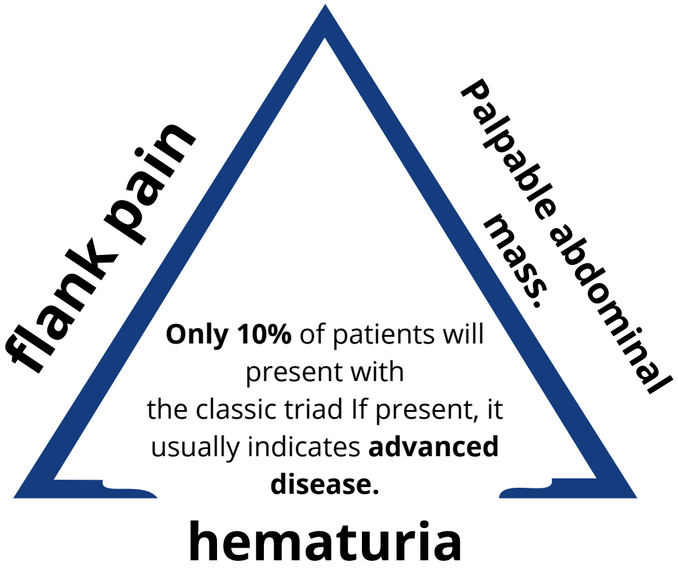
-By Lymphatics to hilar and para-aortic lymph nodes.

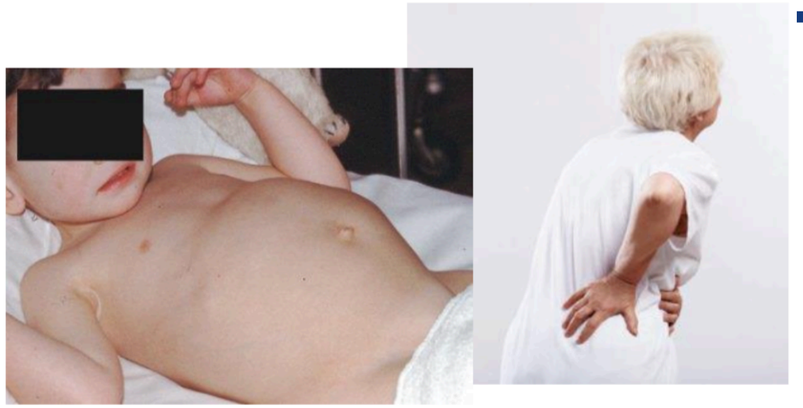
-Hematogenous to lung_(75%), bone (20%), liver (18%), and brain (8%).

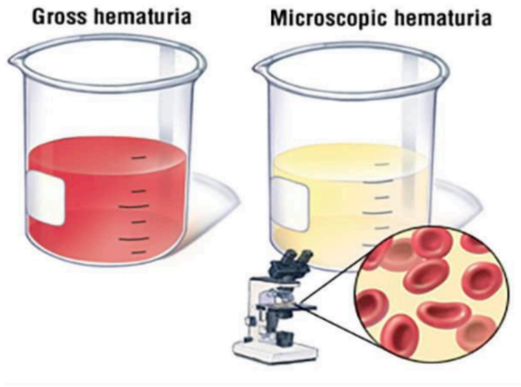
# Clinical presentation

**In the early stage, when the mass is small (‹3cm), renal cell cancer is typically asymptomatic. Approximately 25% of patients are asymptomatic, and the solid renal mass is an incidental finding during a routine radiological study. Symptoms usually do not appear until late, when the may already be large and metastatic**

**-Flank pain**
**- Fever of unknown origin**
**- Palpable mass**
**- Varicocele (2% of males) usually left-sided due to obstruction of the testicular vein.**
**-Nonspecific symptoms fatigue, weight loss, and early satiety**
**-mets symptoms**






**-Hematuria (most common manifestation)**

- **Microscopic: more common.**
- **Gross: less common, blood visible.**
- **Blood throughout the stream or vermiform clots → suggests upper urinary tract origin.**

associated with localized RCC are definitively treated with nephrectomy only

# Paraneoplastic syndromes

occur in 20% of patients. They are symptoms that are caused **not directly by the tumor but rather as a body reaction to the tumor.**

## Non-Endocrine

- *Amyloidosis*
- *Anemia (hematuria, chronic disease)*
- *Neuromyopathies*
- *Vasculopathy*
- *Nephropathy*
- *Coagulopathy*
- *Prostaglandin elevation*

## Endocrine

- Hypercalcemia (PTHrP); most common
- Hypertension ( renin secretion, parenchymal compression, arteriovenous fistula, and polycythemia)
- Polycythemia (EPO)
- Stauffer syndrome (nonmetastatic hepatic dysfunction): elevation of the liver enzymes and hepatic synthetic products; resolved by nephrectomy
- Galactorrhea
- Cushing's syndrome (ectopic ACTH)
- Alterations in glucose metabolism, hypoglycemia (ectopic insulin)

# Investigations

- **Labs**

**- Urinalysis : microscopic hematuria, cancer cells**

**Urine cytology/culture should be normal**

**-CBC : with erythrocytosis/polycythemia, anemia**

**-Electrolyte : panel hypercalcemia**

**- Kidney function tests : creatinine (and BUN) unaffected unless both kidneys a reaffected.**

**- Liver function tests : (elevation of a t least 3 required to diagnose Stauffer syndrome)**

**• Alkaline phosphatase elevation indicates bone scan**

**-Needle biopsy recommended only when the diagnosis would impact treatment choice.**

# Imaging

**Ultrasound has mostly been replaced by CT .**

**Abdominal/pelvic CT scan (with+without contrast): imaging modality of choice.**

- **If a solitary mass is enhancing, the degree of confidence in diagnosing RCC is high.**
- **If mass is predominantly cystic, the confidence level decreases , US may be useful.**

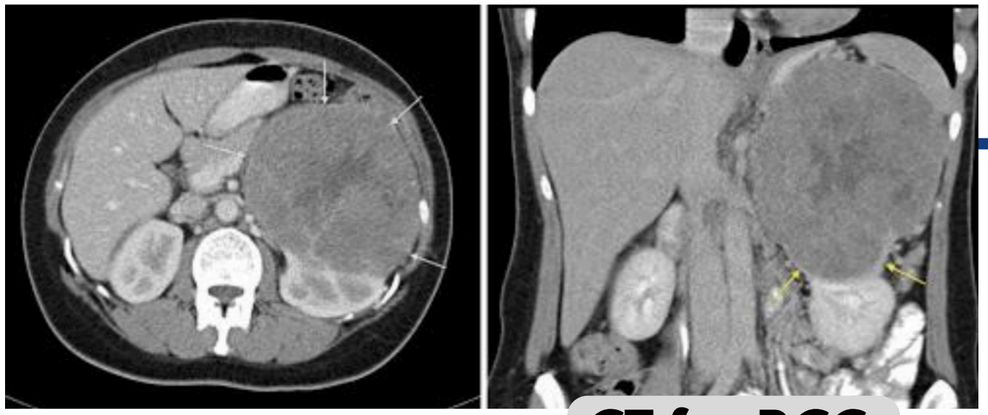
**RCC usually appears as an enhancing solid mass, sometimes with necrosis or cystic features (thick septa, wall nodules)**

**MRI an alternative to pregnancy or when contrast is contraindicated.**

- **(MRA/MRV) When inferior vena cava involvement is suspected.**
- **Brain MRI, if clinically indicated.**

**Chest X-ray: Abdomin.**
**Chest if abnormal chest ct indication.**

**IVU : loss of renal function.**



CT for RCC

**Definitive diagnosis → by surgical pathology after partial or radical nephrectomy.**

# Grading:

**Fuhrman System Nuclear grading system that concurrently evaluates nuclear size+shape and nucleolar prominence to assess aggressiveness o f neoplastic cells.It has been replaced by the ISUP 4-tiered grading system, validated for clear cell renal cell carcinoma (ccRCC) + papillary renal cell carcinoma (pRCC) and has been implemented by the World Health Organization (WHO)**

Worse prognosis

well differentiated

Moderately differentiated

Poorly differentiated

| Grading system | Fuhrman (AJSP, 1982) (historic) | | WHO / ISUP (2022) (current) | |
|---|---|---|---|---|
| | Nuclei (shape & size) | Nucleoli | Nuclei (shape & size) | Nucleoli |
| **Grade 1** | Small, round, uniform (10 um) | Absent / inconspicuous | - | Absent / inconspicuous at 400x |
| **Grade 2** | Larger (15 um) | Visible at 400x | - | Eosinophilic and visible at 400x |
| **Grade 3** | Larger, irregular (20 um) | Visible at 100x | - | Eosinophilic and prominent at 100x |
| **Grade 4** | Pleomorphic, bizarre, giant | Chromatin clumps | Pleomorphic, giant, rhabdoid, sarcomatoid | - |

# Staging

it is very important for :

- prognosis.
- determining treatment modality.

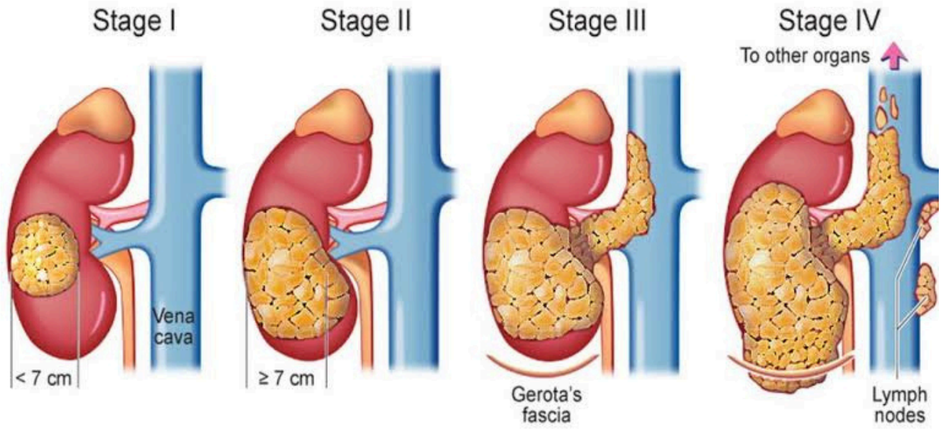
**I:** Limited to kidney, completely excised

**II:** Extending outside kidney, completely excised

**III:** Extending outside kidney, incompletely excised

**IV:** Distant metastases

**V:** Bilateral renal tumors




- **T : 1ry tumor**
  - T1:tumor less than 7cm , limited to the kidney.
  - T2: tumor more than 7cm , limited to the kidney.
  - T3: spread to major veins , adrenal gland or perinephric tissues .
  - T4:Spread beyond Gerota's fascia .
- **N: Regional lymph nodes**
  - N0: no regional LNs metastases
  - N1: one regional LNs metastases
  - N2: more than one regional LNs metastases
- **M : Distal metastases**
  - M0:No distal metastases .
  - M1: distal metastases .

## Secondary Renal Tumors

**The most common cancers that mets. to the kidney are :**

- **lung (20%)**
- **breast (12%)**
- **stomach (11%)**

**Melanoma and other solid tumors also possible.**

- **early RCC, surgical treatment, active surveillance, or thermal ablation.**
- **advanced RCC, palliative therapies or experimental protocols.**

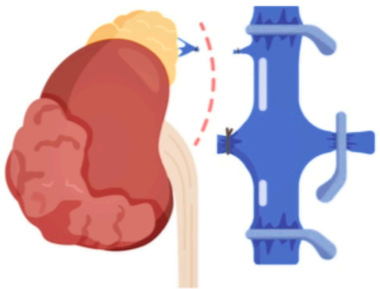
# Treatment

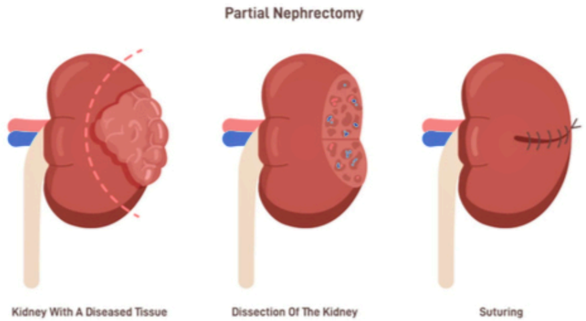
## Stage I:

- Thermal ablation: nonsurgical destruction of renal tumors via freezing (cryosurgery) or thermal energy (radiofrequency ablation), often percutaneously; considered for tumors <3 cm.

- Radical Nephrectomy: standard treatment for localized RCC and provides a reasonable chance for cure; considered in patients with a normal contralateral kidney, normal kidney function, and more advanced primary tumors (cT1b through cT4)▪ Removal of: kidney, perirenal fat, Gerota fascia, upper hald of ureter, +/- ipsilateral adrenal gland

- Partial Nephrectomy: standard of care for clinically staged T1a tumors (cT1a, < 4 cm) and should be considered in patients with T1b or T2 tumors, and absolute indications for nephron preservation:Both kidneys affected, CKD, solitary kidney

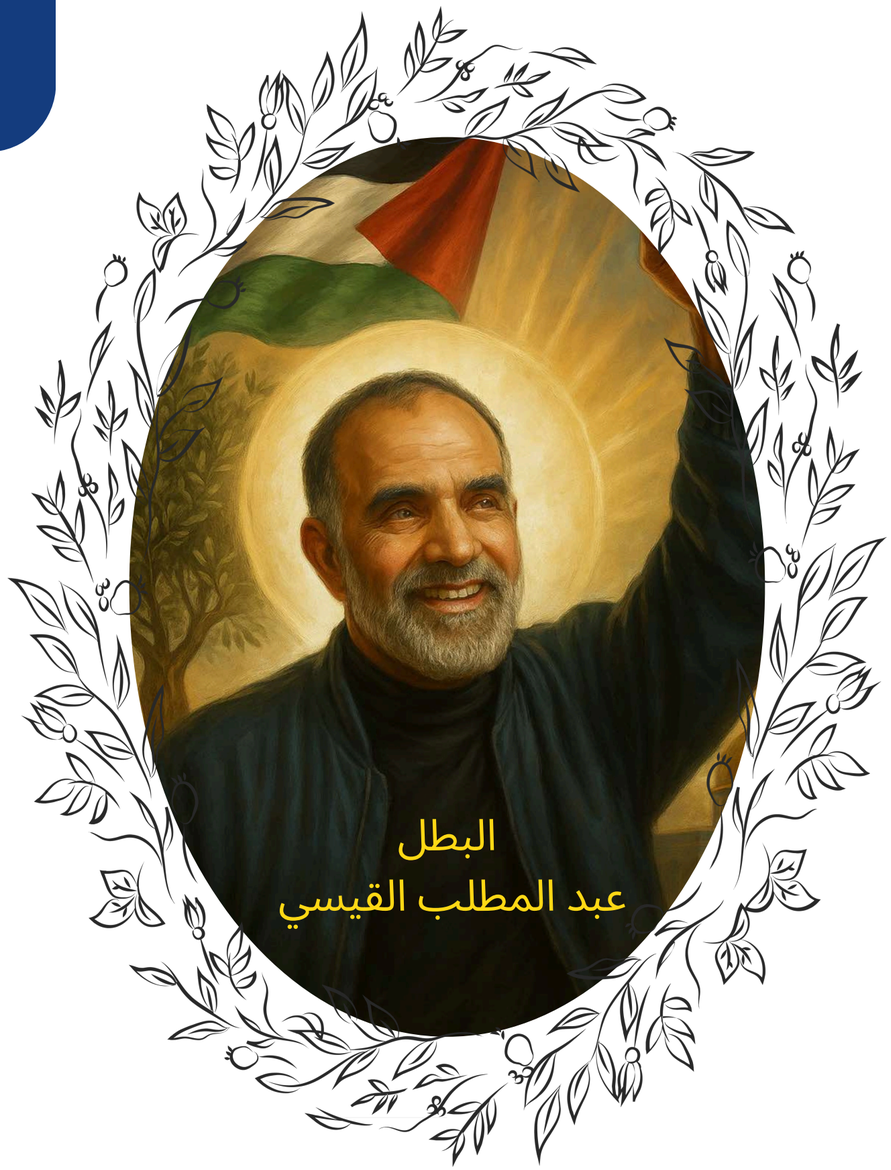


- stage II renal cell cancer, laparoscopic radical nephrectomy is the treatment of choice.

- stage III renal cell cancer, open radical nephrectomy is the standard of care. Adrenalectomy or extensive lymph node dissection is only recommended when abdominal CT shows evidence of adrenal or lymph node invasion.

- Stage IV renal cell cancer is not curable. Treatment is palliative
  - Tumor embolization
  - External-beam radiation
  - Nephrectomy
  - Drug therapy
  - Immunotherapy: interferon alfa-2b or IL-2 (Aldesleukin)
  - Molecular-targeted therapy: tyrosine-kinase inhibitors (sunitinib, sorafenib, bevacizumab, pazopanib), mTOR inhibitors (temsirolimus and everolimus)
  - immune checkpoint inhibitors: monoclonal antibodies against PD-1 or PD-L1
- Although metastatic RCC is traditionally characterized as radioresistant, radiation therapy can be palliative when RCC is oligometastatic, particularly to the bone.




# Thank you

سَتُسَألُ وحدَك فالزمْ ثغرَك وأغِث أمّتك
وكلٌّ بما ملكَتْ يداه !