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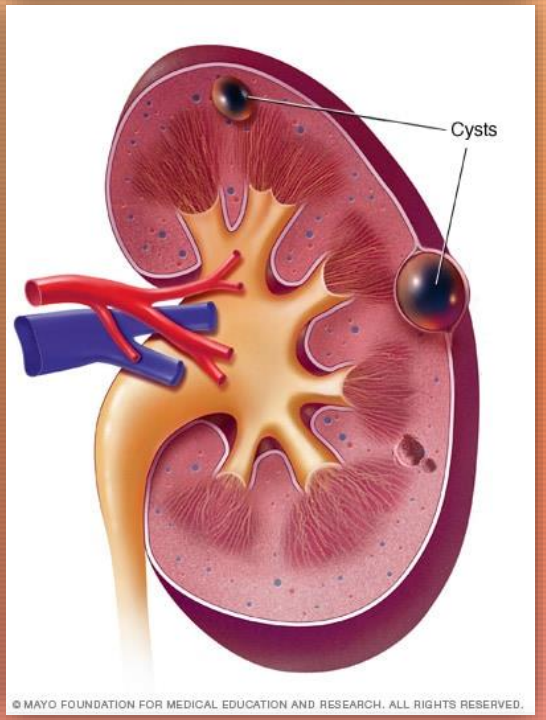
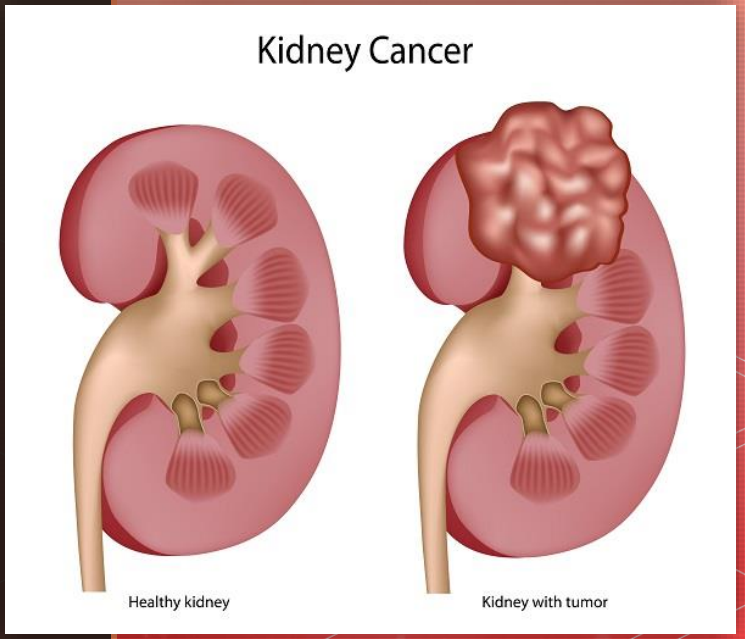
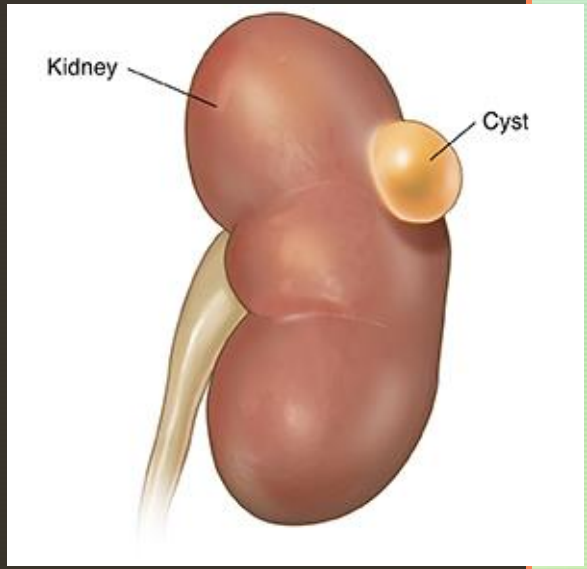
Renal cysts and Renal Tumors

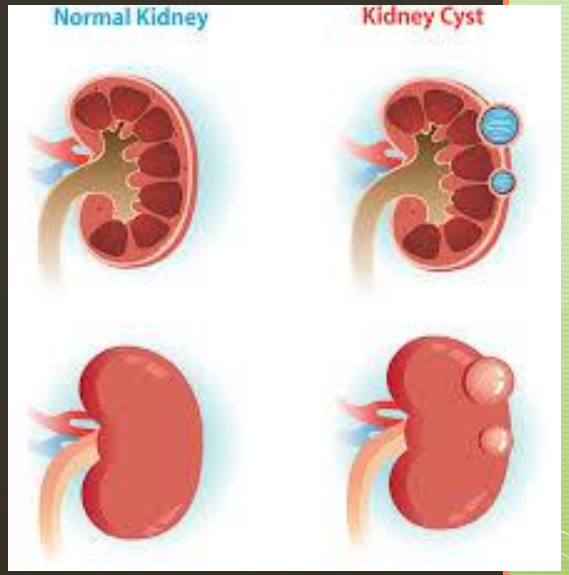




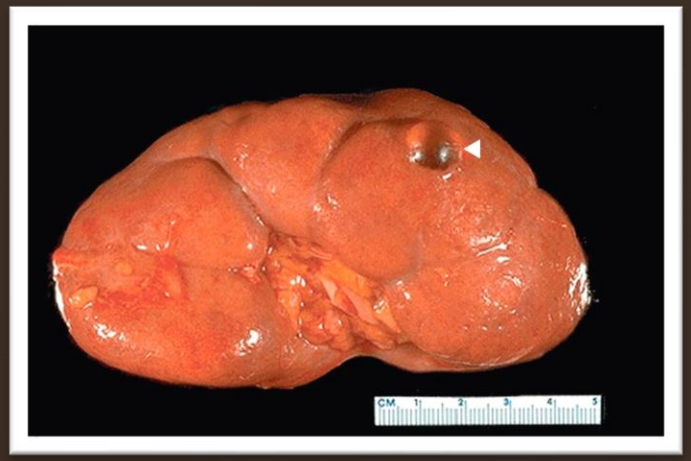
Renal Cysts

Renal cysts are fluid-filled sacs that develop within the kidneys.





Characteristics of Simple Cysts



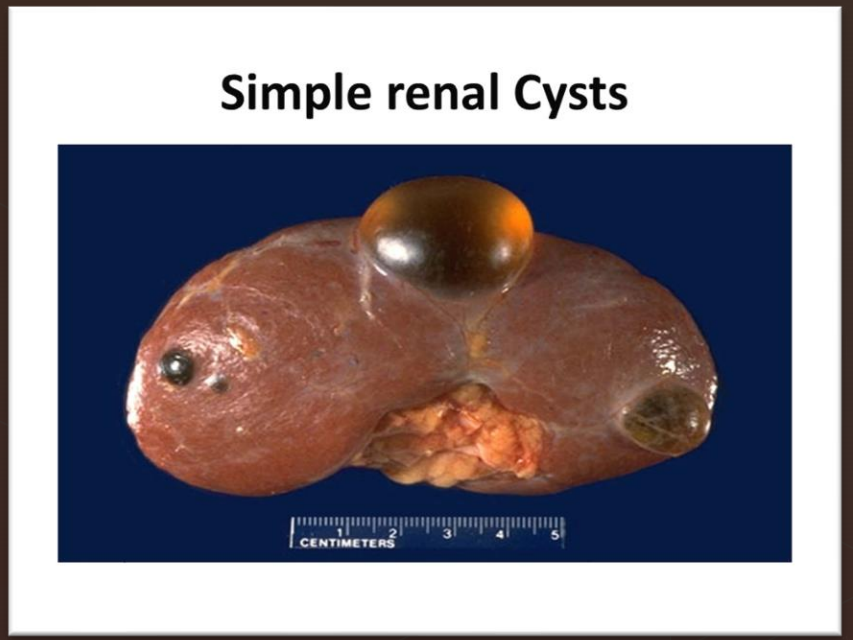
The most common (70%)

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:

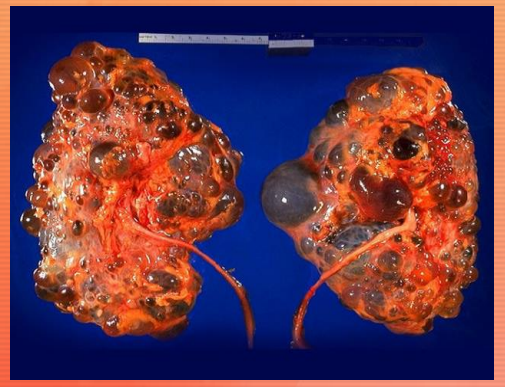
- Contains Clear Fluid & may contain altered blood
- Lined by Flat epithelium.
- Surrounded by Fibrous tissue



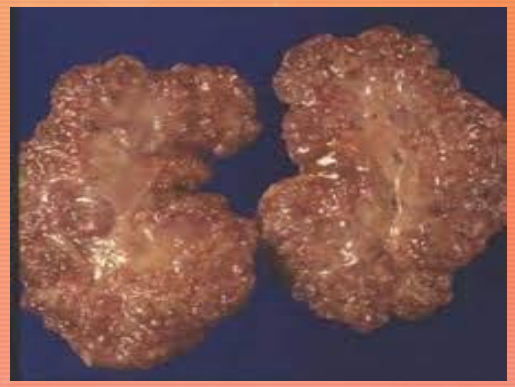
Etiology of Renal Cysts

1 **Developmental**

Multicystic dysplastic kidney (MCDK)



(ADPKD)



(ARPK)

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 **Autosomal Dominant Polycystic Kidney Disease (ADPKD)**

progressive bilateral disease leading to hypertension and renal failure associated with hepatic cysts and cerebral aneurysms

Usually Asymptomatic

Simple renal cysts are usually **asymptomatic**

Dull Aching Pain

In some cases, a dull aching pain in pain in the loin may be experienced due to the stretch of of the renal capsule by the cyst. cyst

Swelling in the Loin

A swelling may be felt in the loin loin area

Complications

- Hemorrhage
- Rupture and pressure on the ureter (Hydronephrosis)
- Infection and Calcification

Cysts Associated with Systemic Disease

1. **Von Hippel-Lindau Syndrome (VHLS)**

   Renal cysts, cerebellar and retinal hemangioblastomas, pancreatic and epididymal cysts (30-40% incidence of renal cell carcinoma)

2. **Tuberous Sclerosis (TS)**

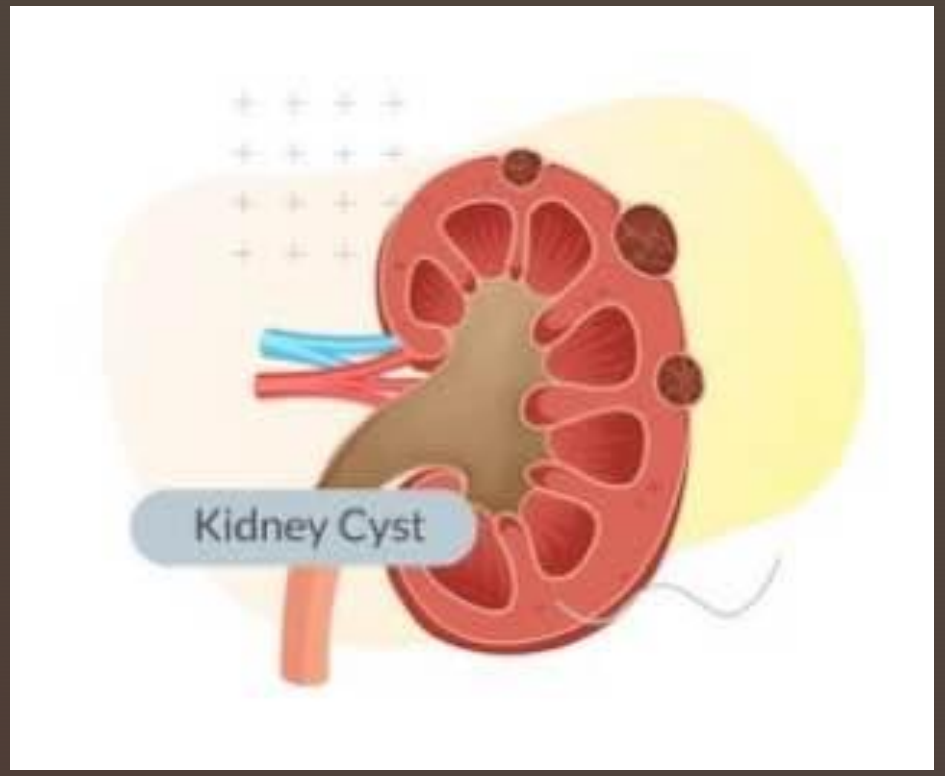
   Autosomal dominant syndrome characterized by mental retardation, epilepsy, adenoma sebacaum, and other hamartomas

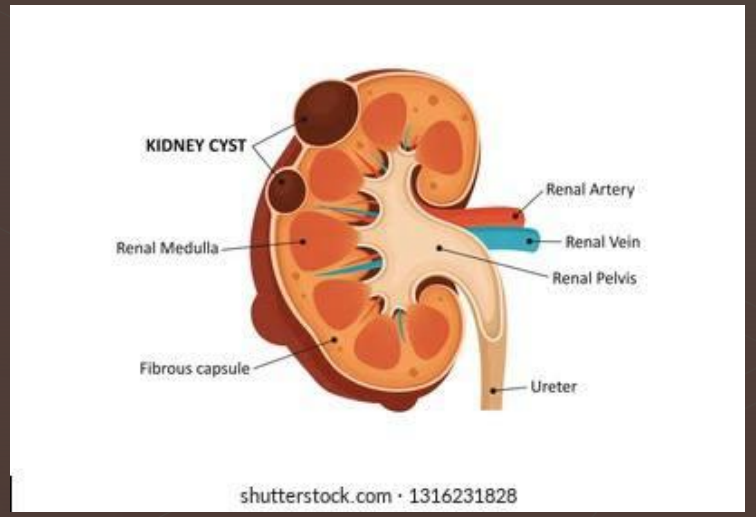
3. **Acquired Simple cysts, acquired cystic renal disease, medullary sponge kidney (MSK):**

   dilatations of the collecting ducts usually benign course, but predispose to calcium phosphate stones

4. **Malignancy Cystic renal cell carcinoma (RCC)**

Diagnosis


Kidney Cyst

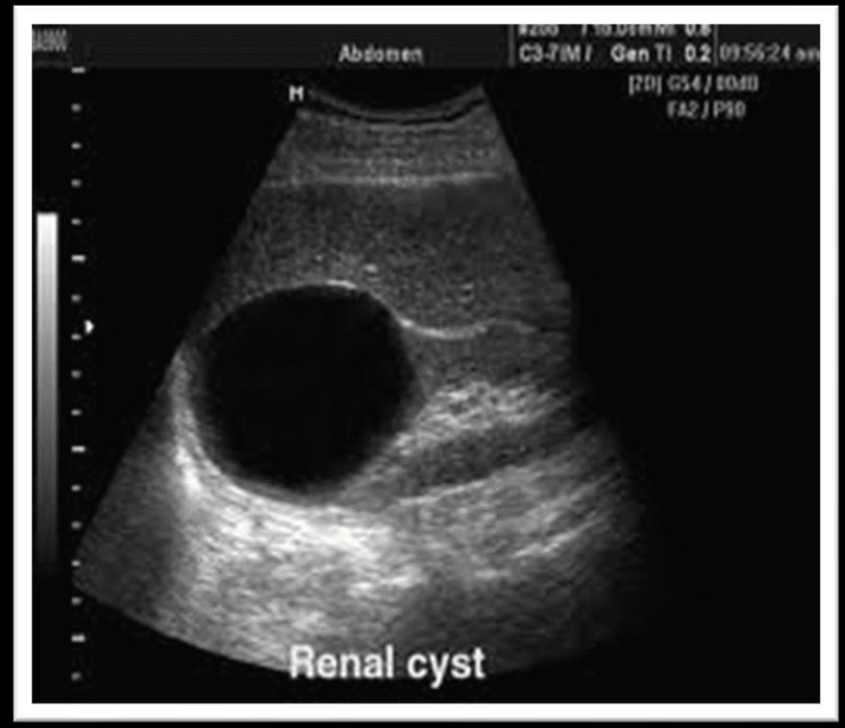

KIDNEY CYST
Renal Artery
Renal Vein
Renal Pelvis
Renal Medulla
Fibrous capsule
Ureter
shutterstock.com · 1316231828

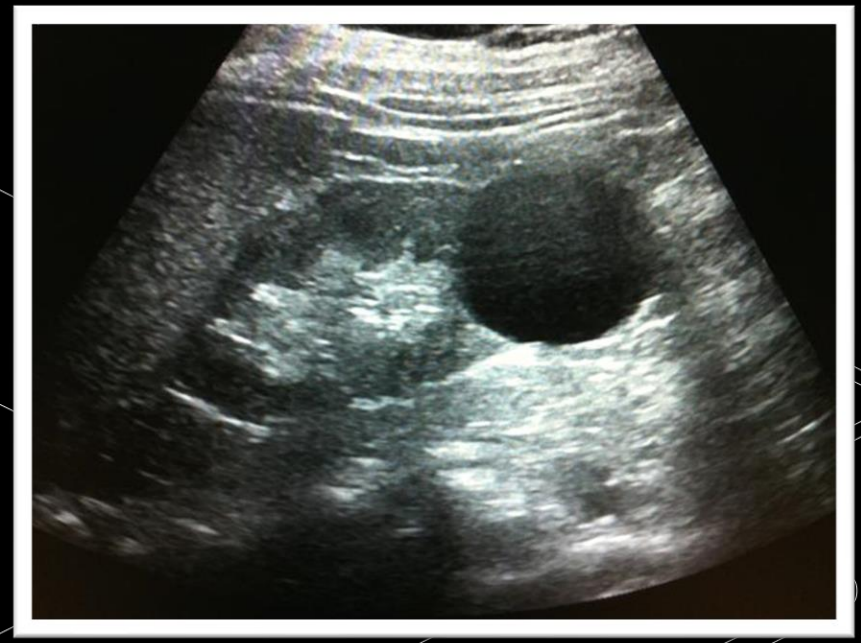
Ultrasonography

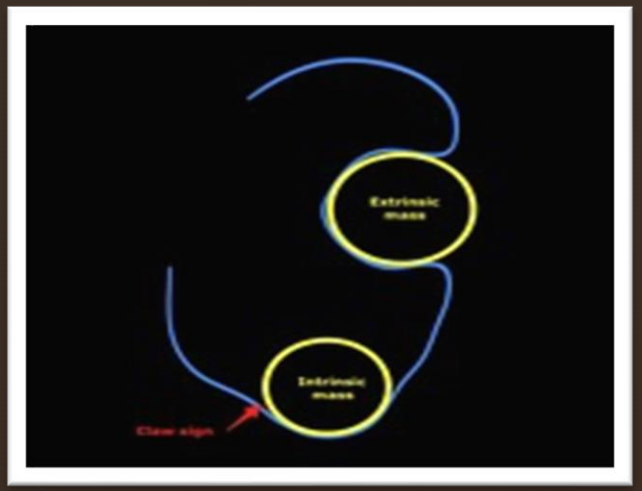
- It is very helpful
- **Differentiates between <u>SOLID and CYSTIC lesions</u>**.

- Features of simple cyst are:

1. Anechoic (echo lucent ; absence of echoes)
2. Posterior acoustic enhancement
3. Sharply marginated thin smooth walls
4. round/oval shape
5. no septation, no calcification





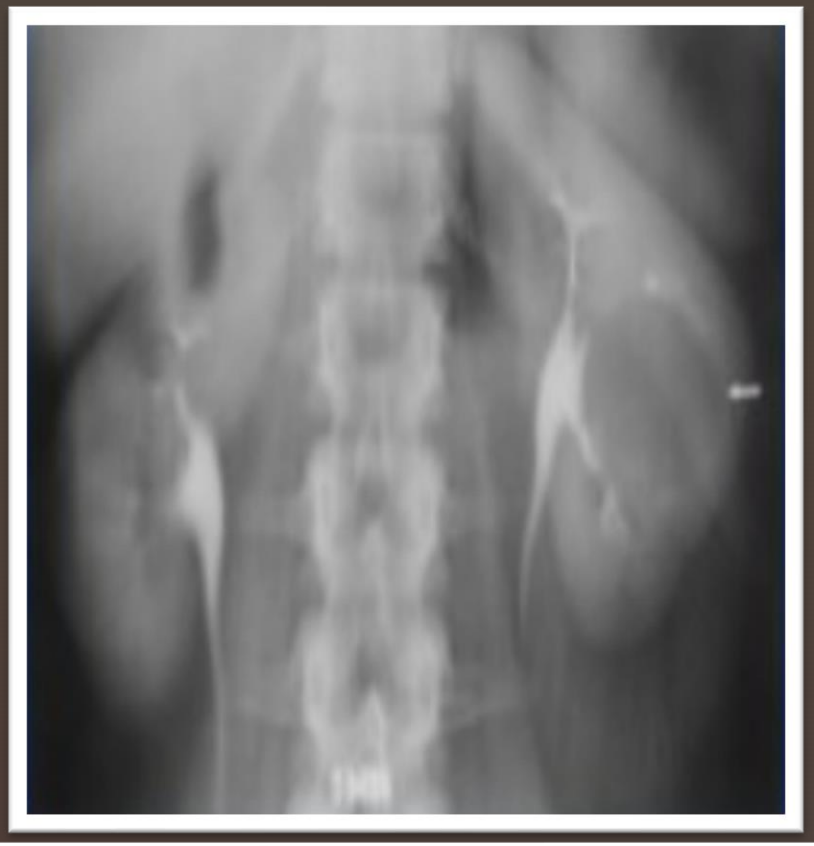


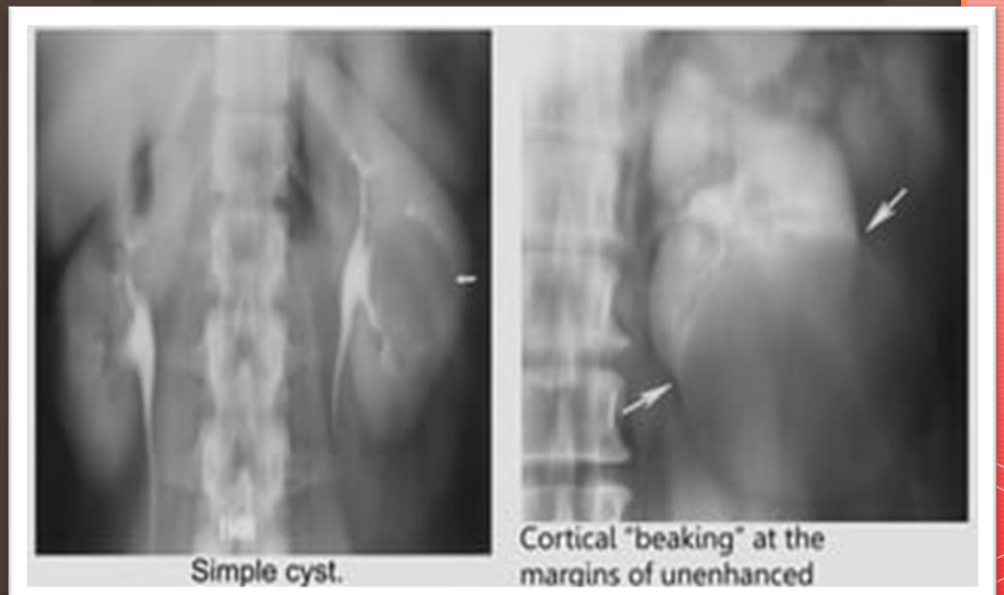
IVU Signs

smooth amputated calyx

“beak”, “claw” sign

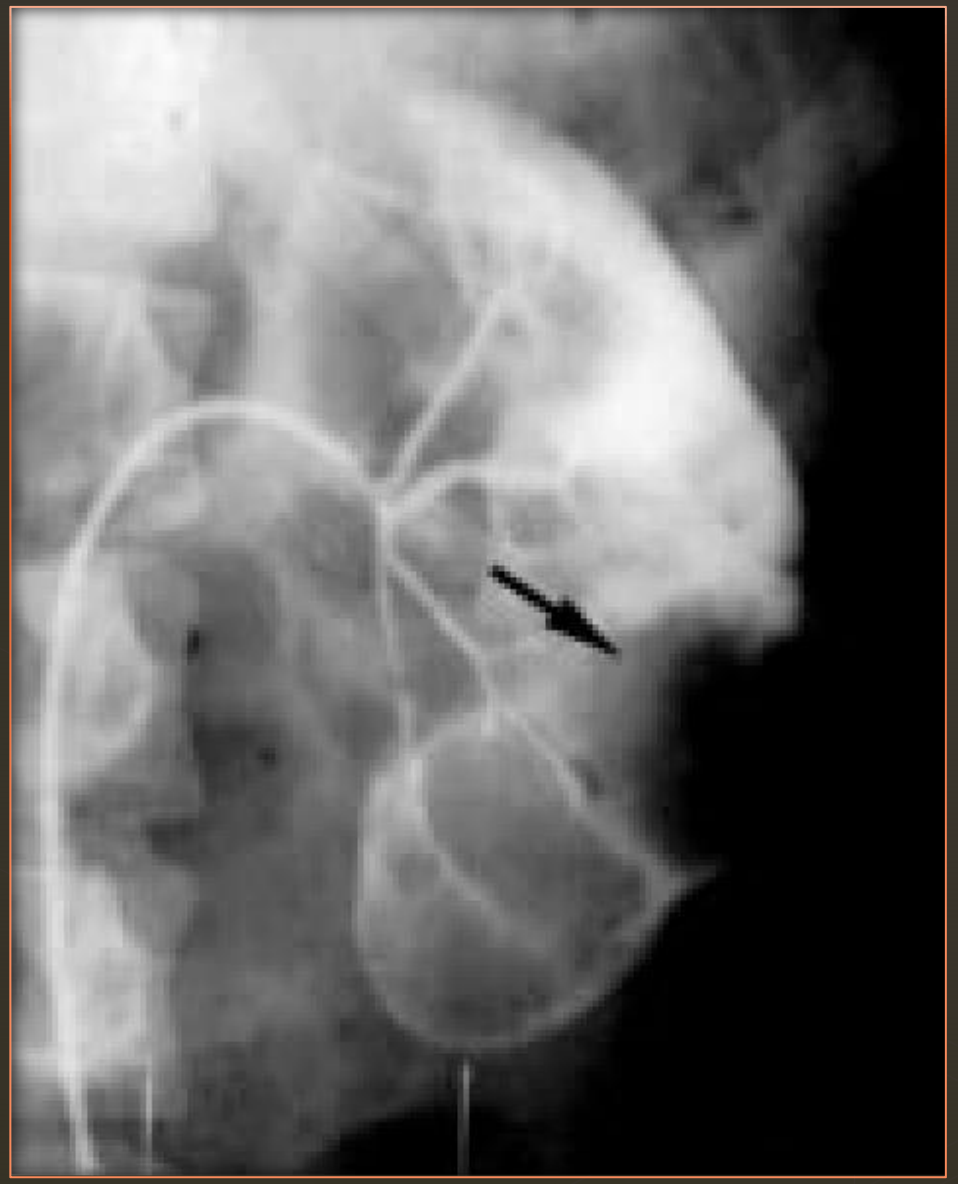
In cases when some masses or cysts grow and bulge from renal parenchyma, the normal tissue covers their contours with the formation of acute angles – the "claw" sign or the "beak" sign. This results in a smooth amputated calyx appearance on IVU.





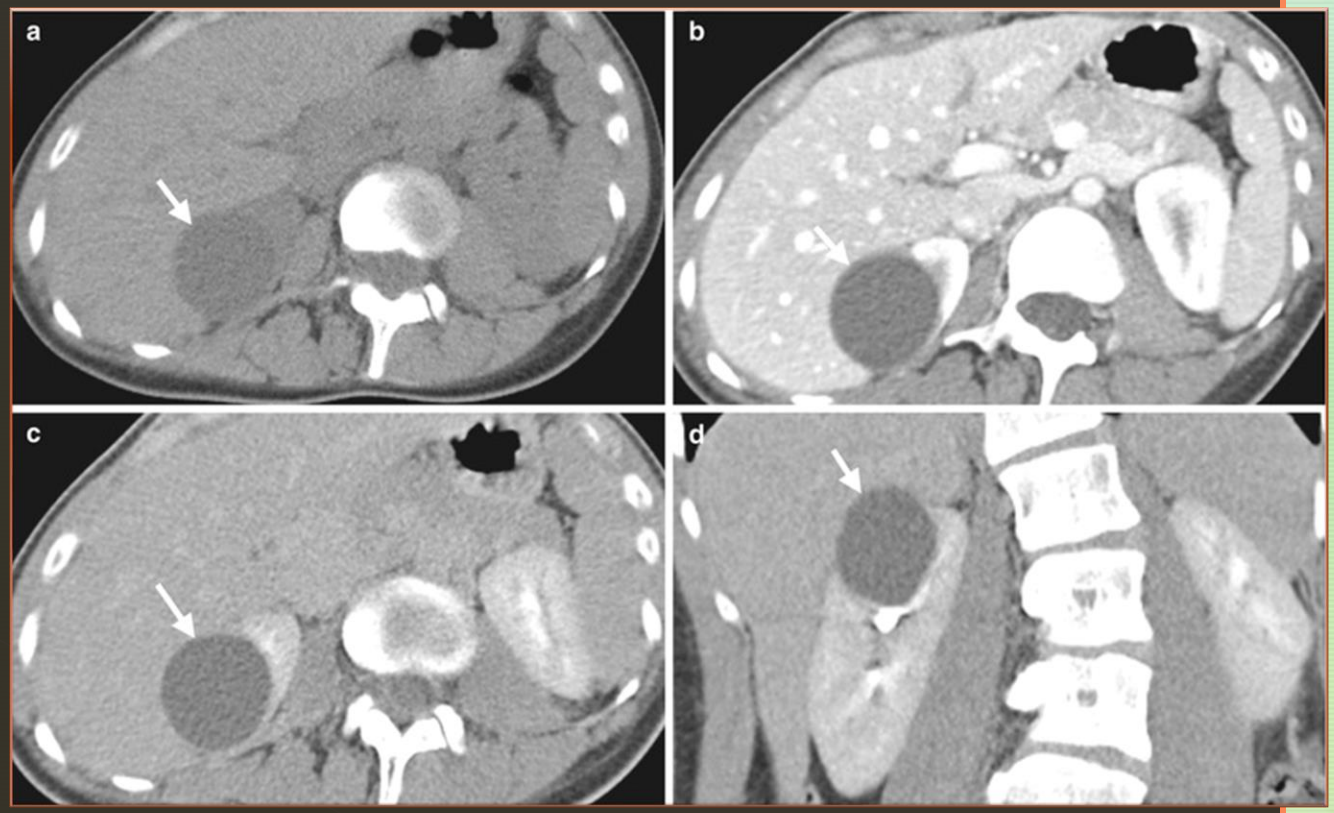
Renal Angiography

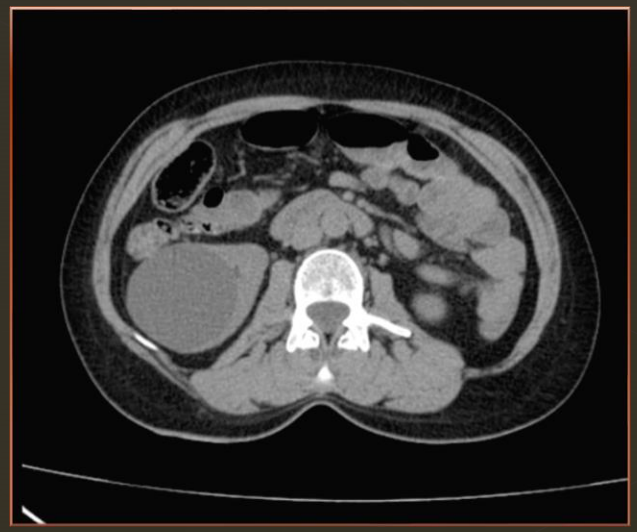
- Renal angiography is not commonly performed now
- it can be useful to differentiate between a cyst and a tumor.
- Cysts are typically: avascular
- tumors are :hypervascular

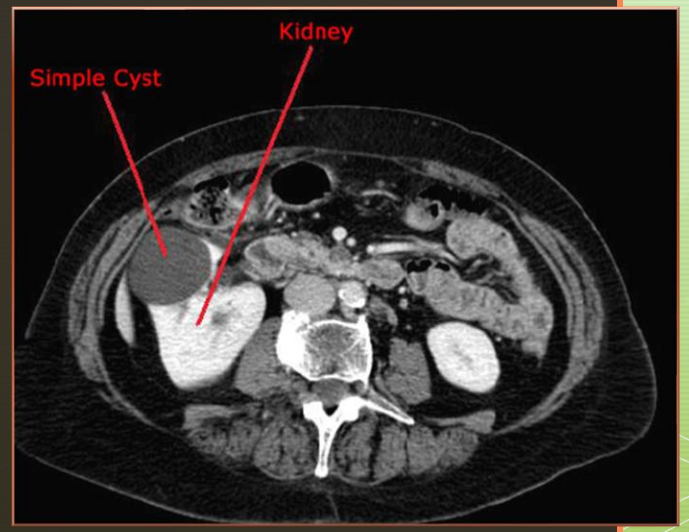


CT

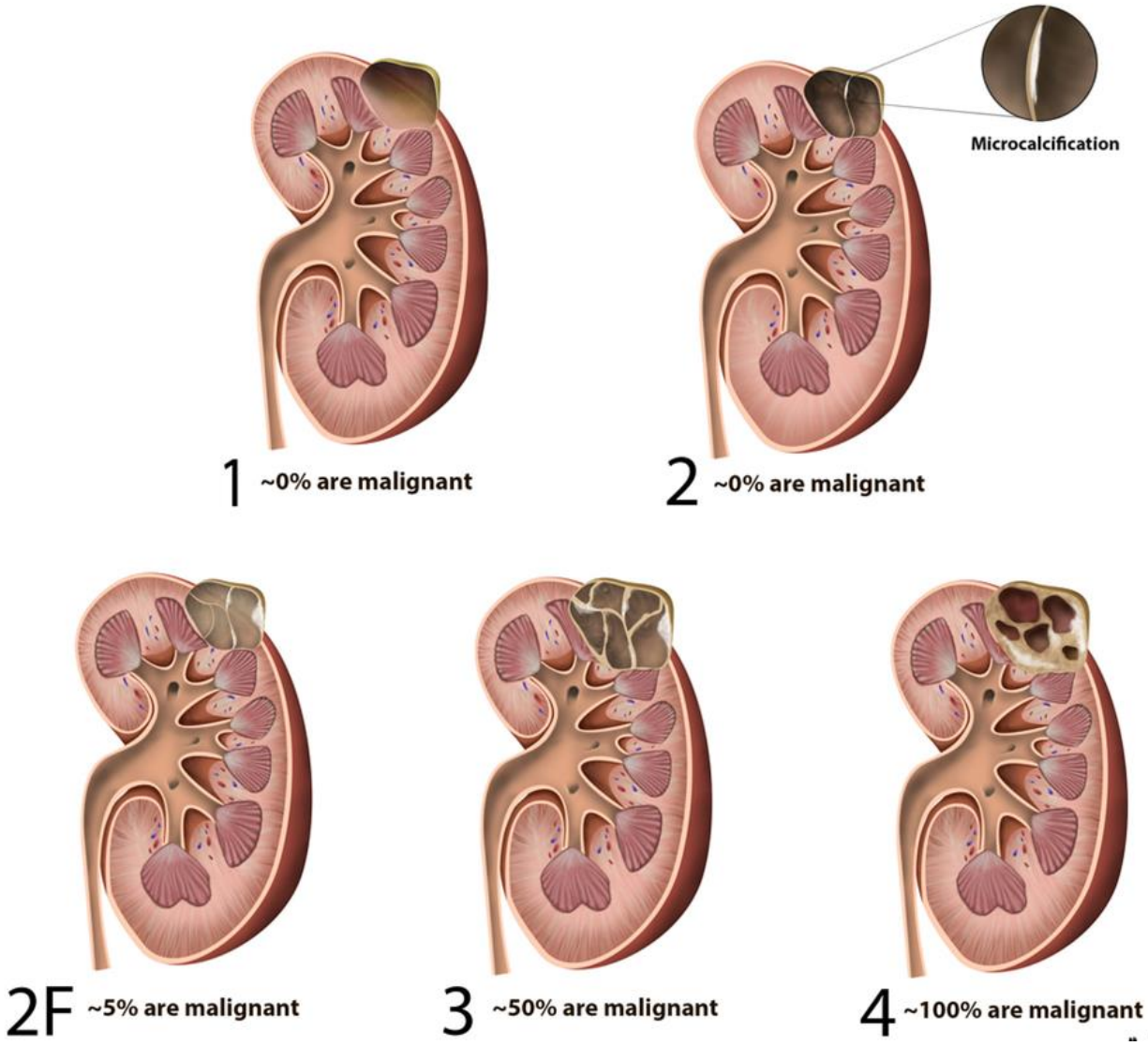
- **Distinguishes fluid – only filled cysts from solid masses**

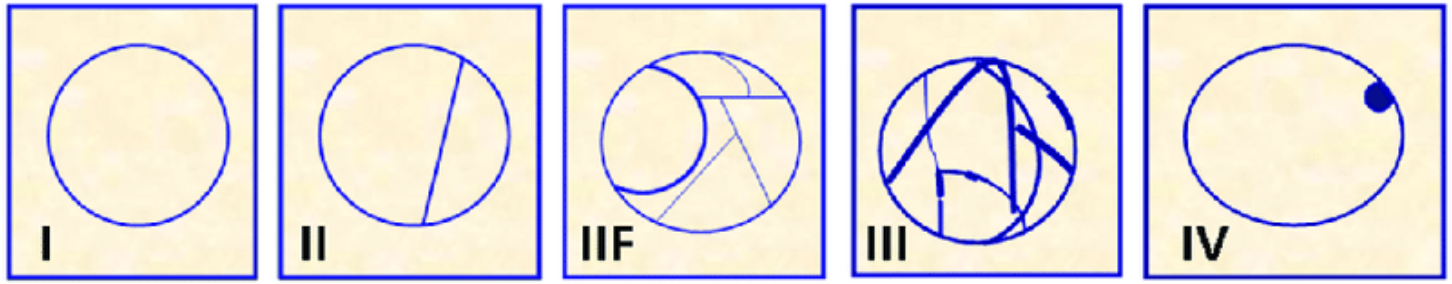




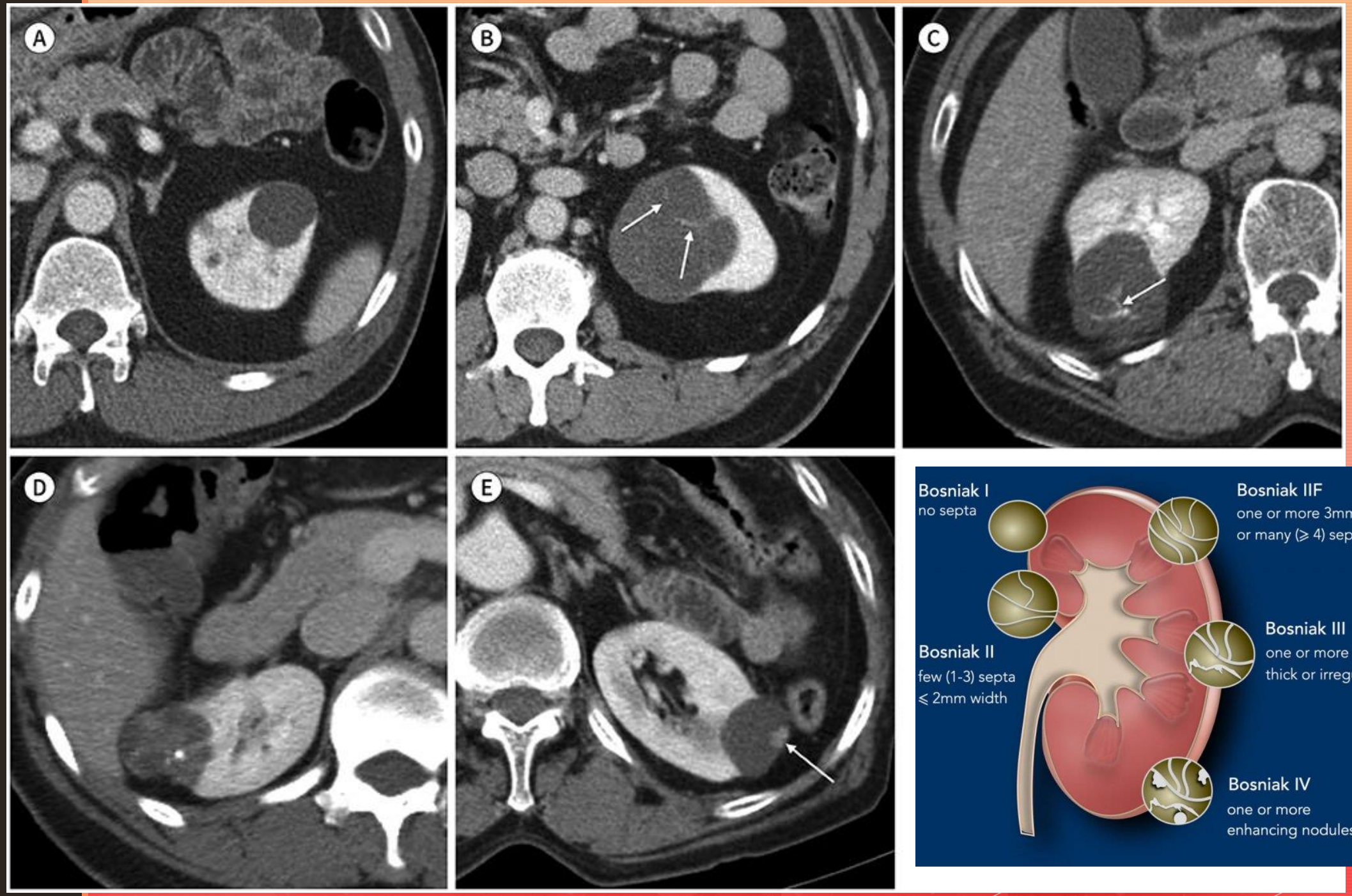


Bosniak classification of renal cysts

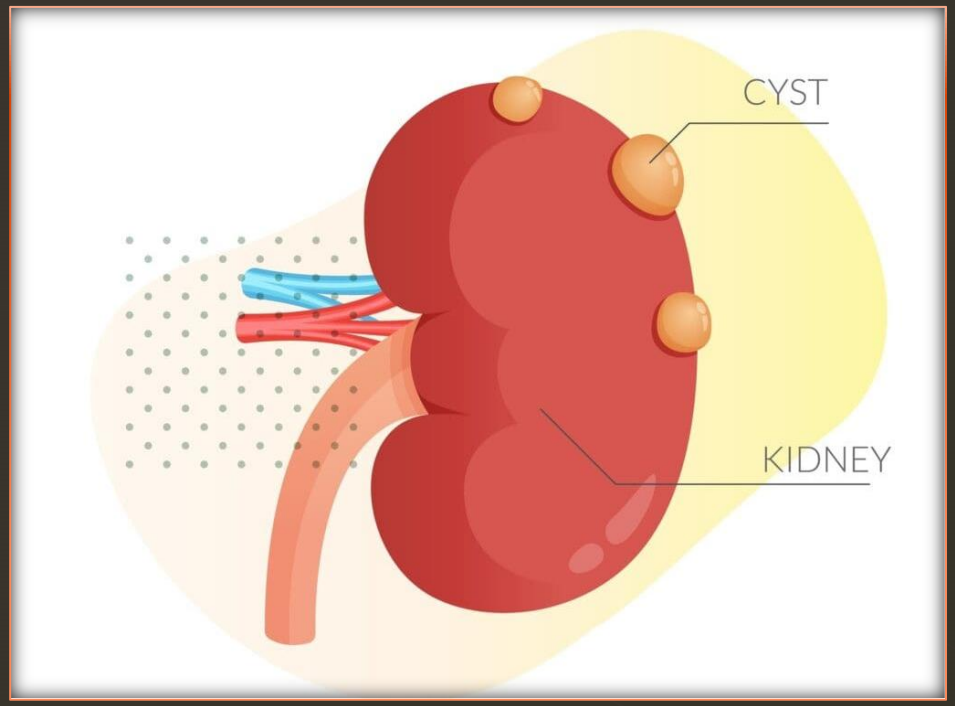


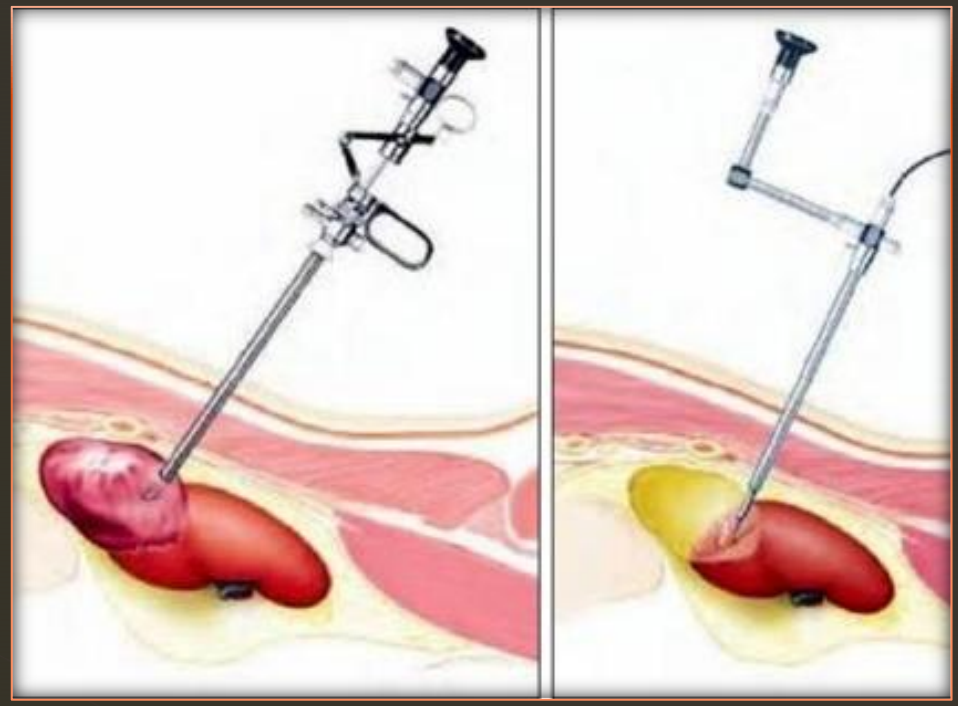


Bosniak stage	Risk of malignancy (%)	Features
I	0	Thin wall without septa or solid components. No internal enhancement.
II	0	Few (≤3) thin septa. It may show minimal enhancement of the septa. Hyperintense on T1 without enhancement.
IIF	5	Multiple (>3) thin septa. Smooth mild thickening (3 mm) of the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


A
B
C
D
E

Bosniak I
no septa
Bosniak IIF
one or more 3mm septa
or many (⩾ 4) septa ⩽ 2mm
Bosniak III
one or more ⩾ 4mm
thick or irregular septa
Bosniak II
few (1-3) septa
⩽ 2mm width
Bosniak IV
one or more
enhancing nodules





Treatment

Usually no need for treatment but follow up is required

if too much pressure or the cyst affects kidney function; (hydronephrosis):

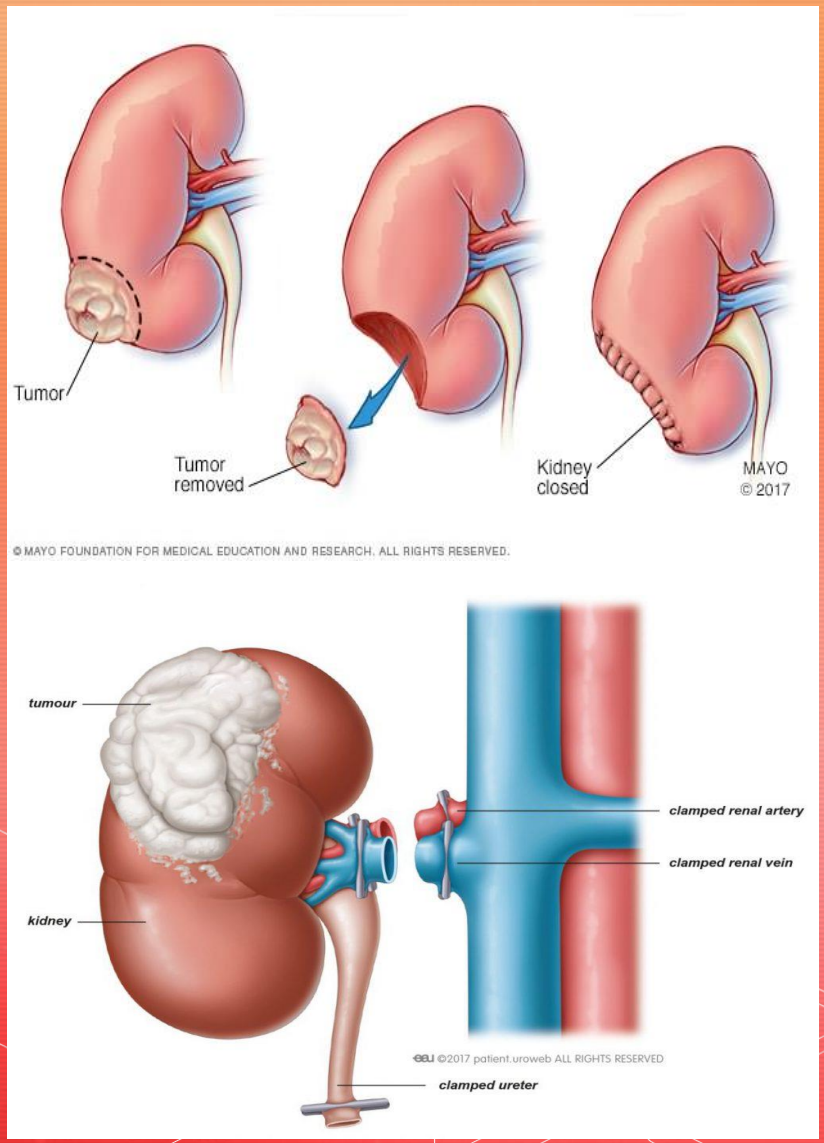
- Aspiration and sclerotherapy by 95% alcohol
- Excision

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 maybe considered .

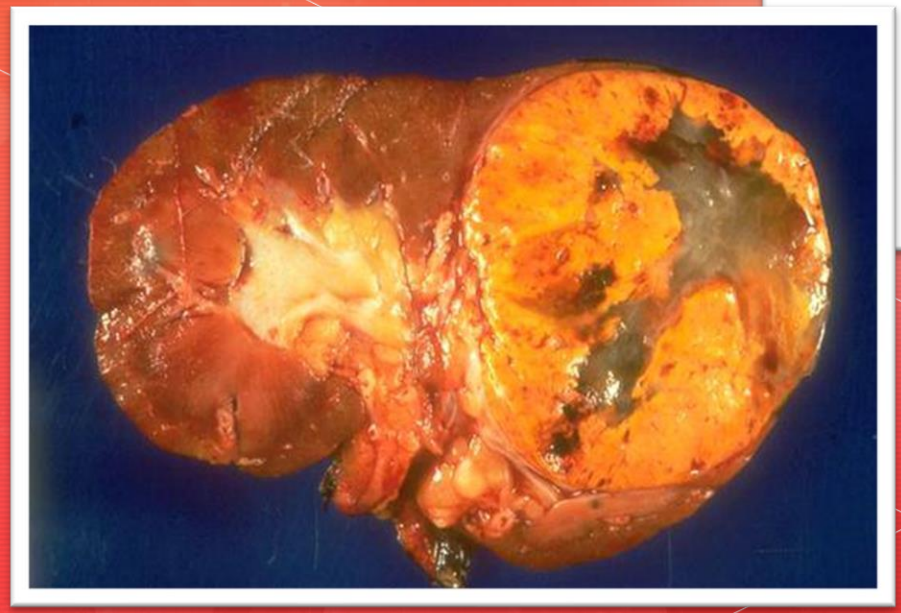
PNA : percutaneous needle aspiration

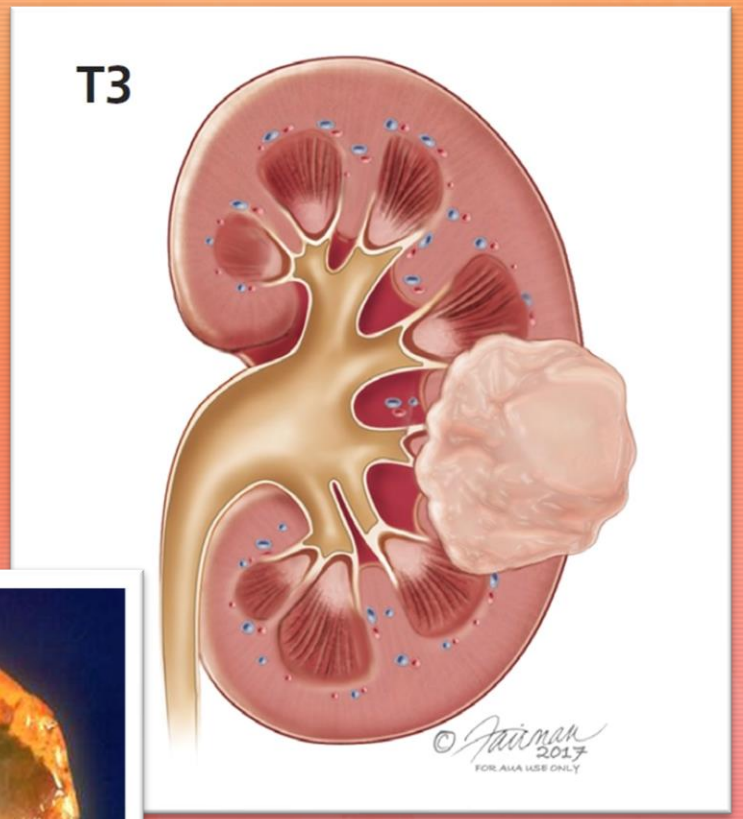
- **Partial nephrectomy:** also called kidney-sparing (nephron-sparing) surgery, the surgeon removes diseased tissue from a kidney and leaves healthy tissue in place.

- **Radical (complete) nephrectomy:** the surgeon removes the entire kidney and often some additional structures, such as part of the ureter or other adjacent structures such as the adrenal gland or lymph nodes



RENAL TUMORS





BENIGN RENAL TUMORS

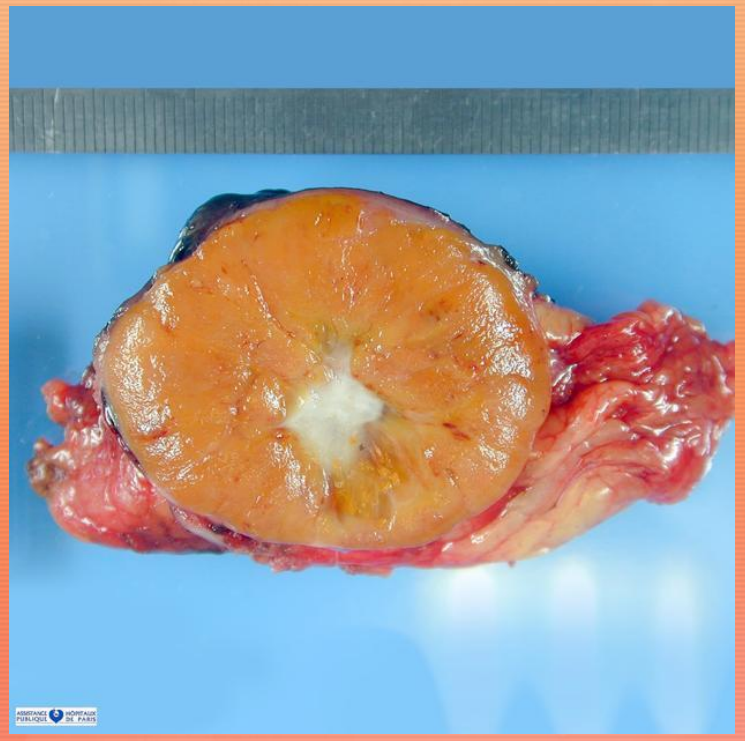
1Oncocytoma
2-Angiomyolipoma
3-Papillary adenoma

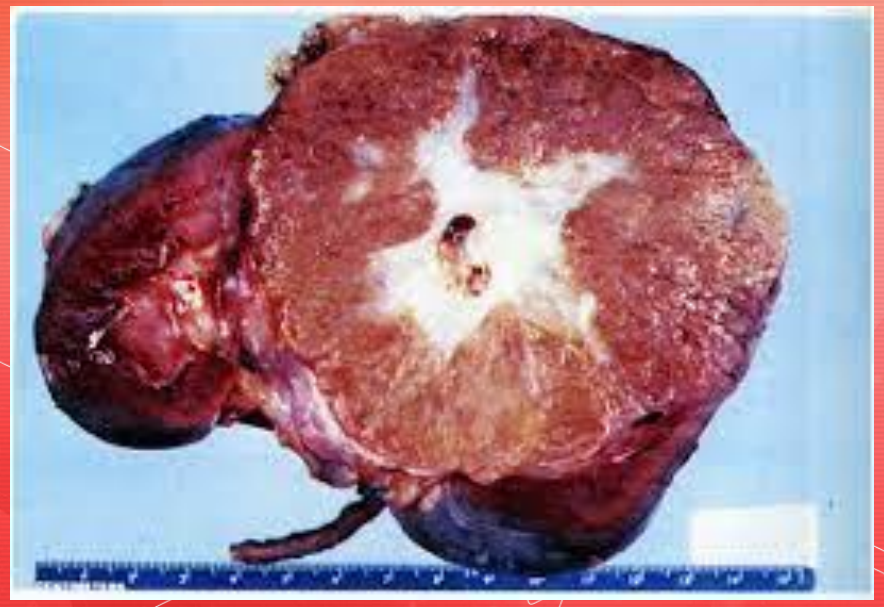
Table 1. WHO Classification of Benign Renal Masses

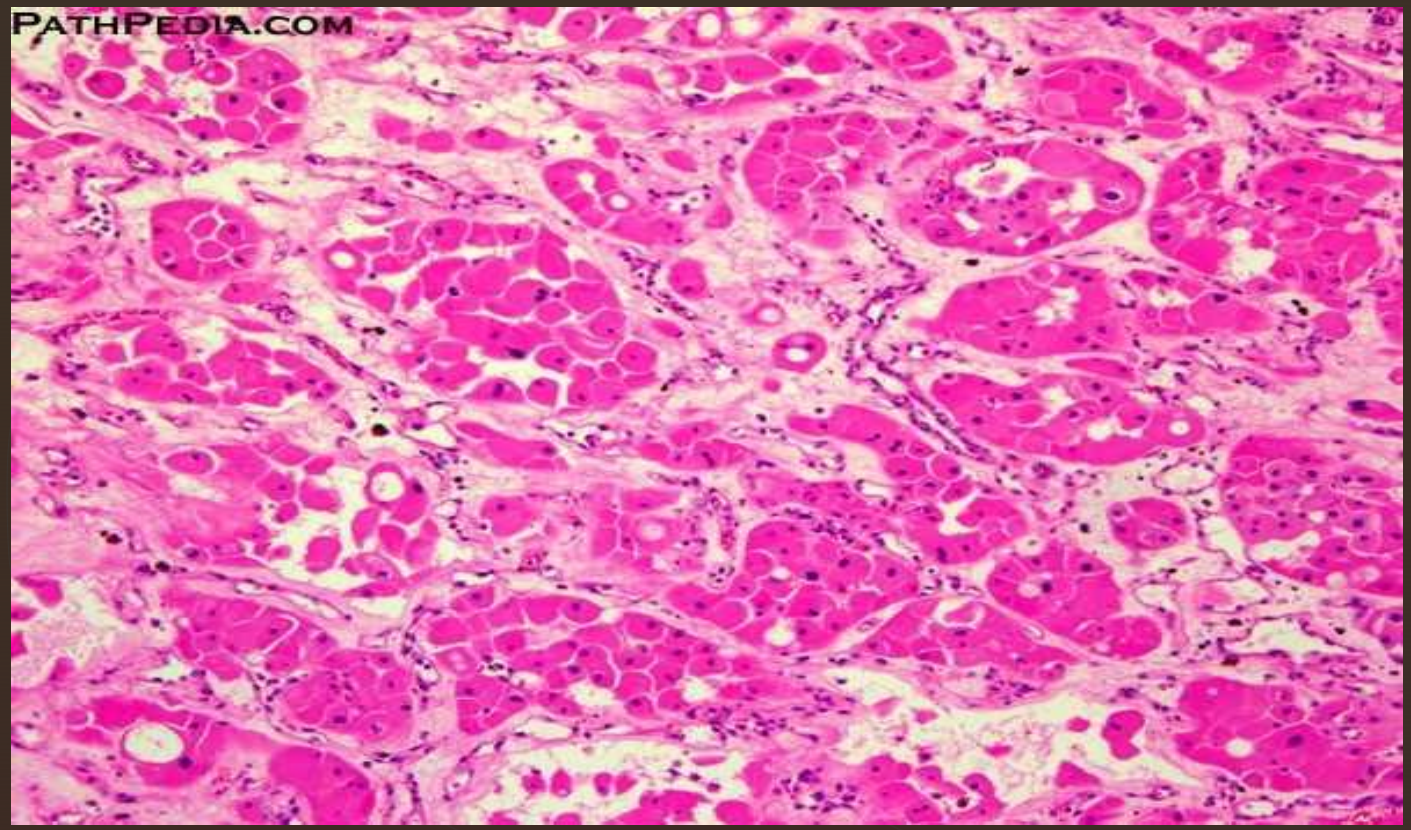
Classification	Types
Epithelial Tumors	Onocytoma Papillary adenoma
Mesenchymal Tumors	Angiomyolipoma Leiomyoma Hemangioma Reninoma Schwannoma Lymphangioma
Mixed Epithelial and Mesenchymal Tumors	Mixed epithelial and stromal tumor Cystic nephroma
Metanephric Tumors	Metanephric adenoma Metanephric adenofibroma Metanephric stromal tumor

Oncocytoma:

- BENIGN RENAL EPITHELIAL NEOPLASM that arise from the **intercalated cells** of collecting ducts that comprises approximately 5-9% of renal **tubular epithelial** tumors
- accounting for 3-7% of renal 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

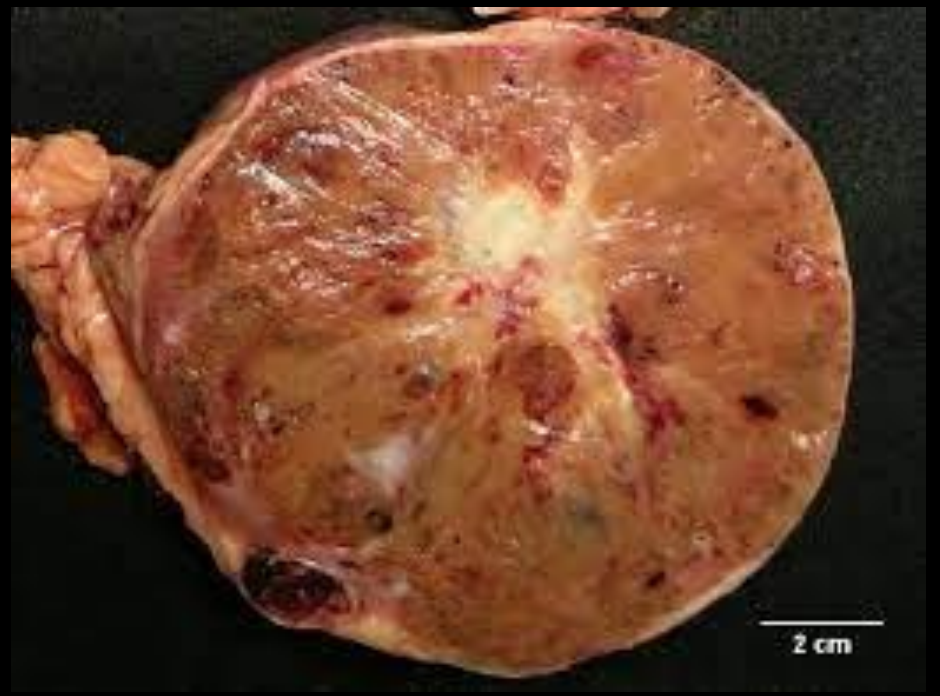






- **Histological**: organized eosinophilic cells originating from intercalated cells of collecting duct , mitoses are rare and they are considered benign , not known to metastasize.

Oncocytoma



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

4 **Shape:**

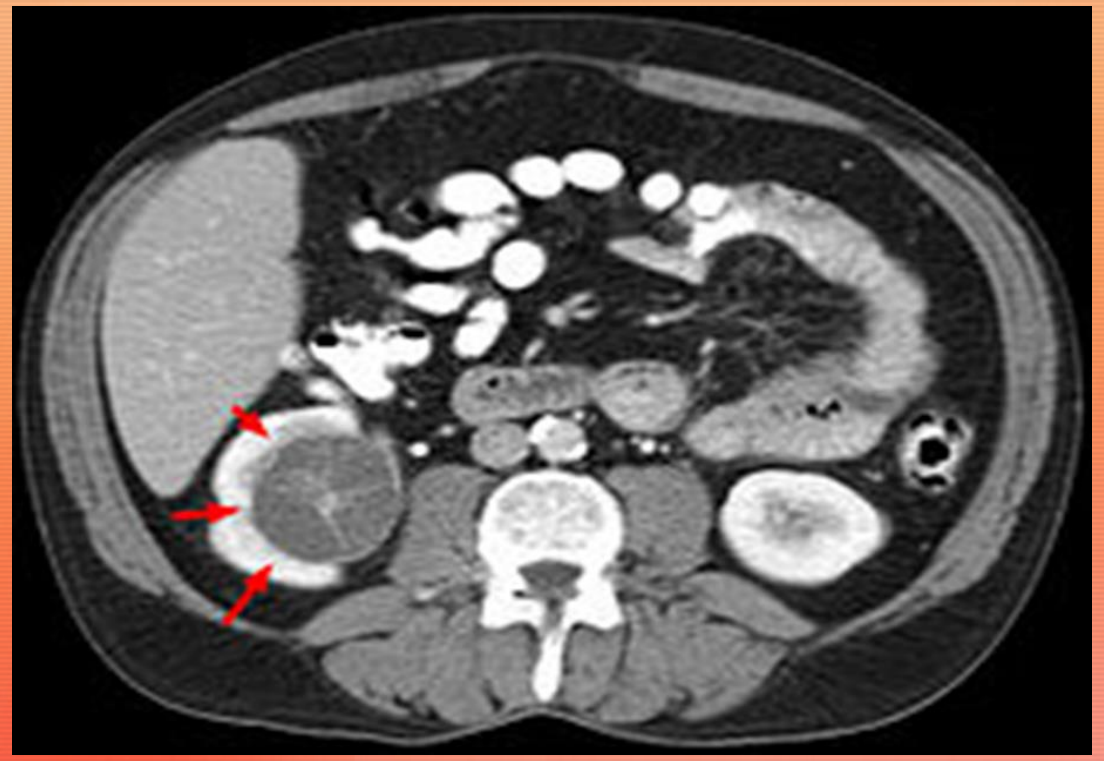
Spherical, solitary lesion

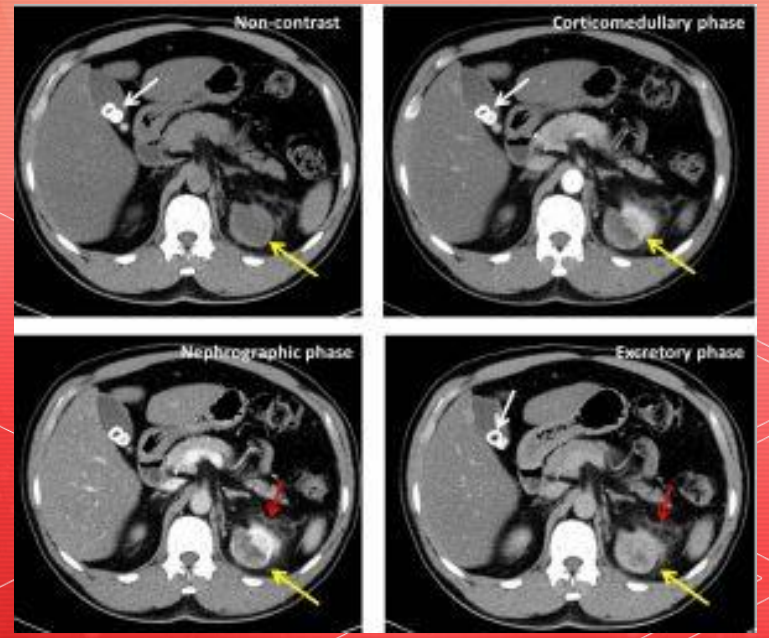
3 **Color and Texture:**

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

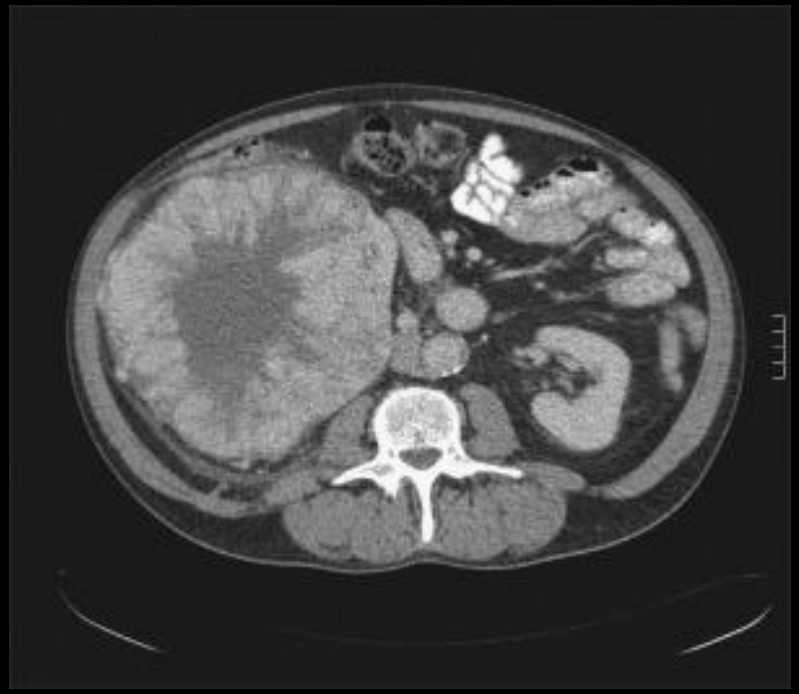
Diagnosis :

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





Oncocytoma Management



1. **Huge Oncocytomas:**

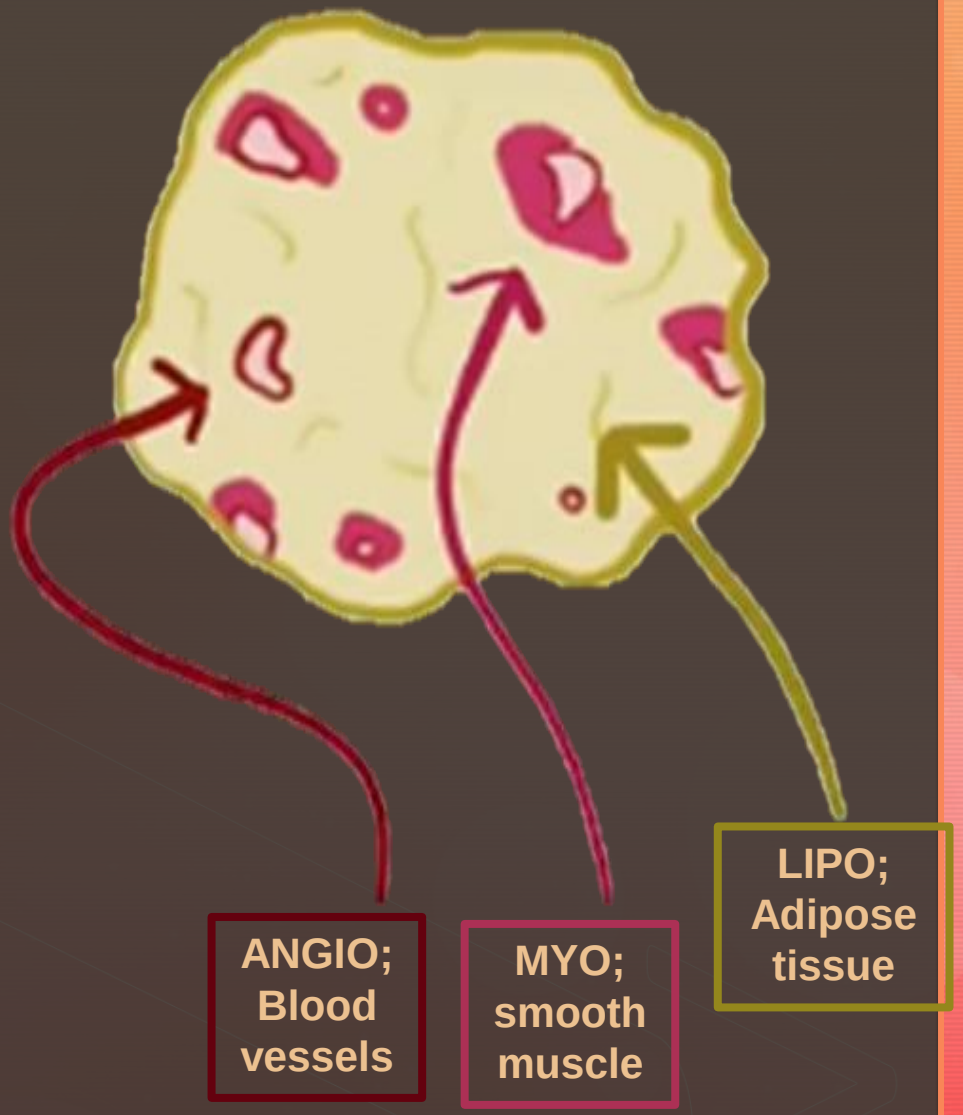
   Partial or radical nephrectomy

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 waves

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.**

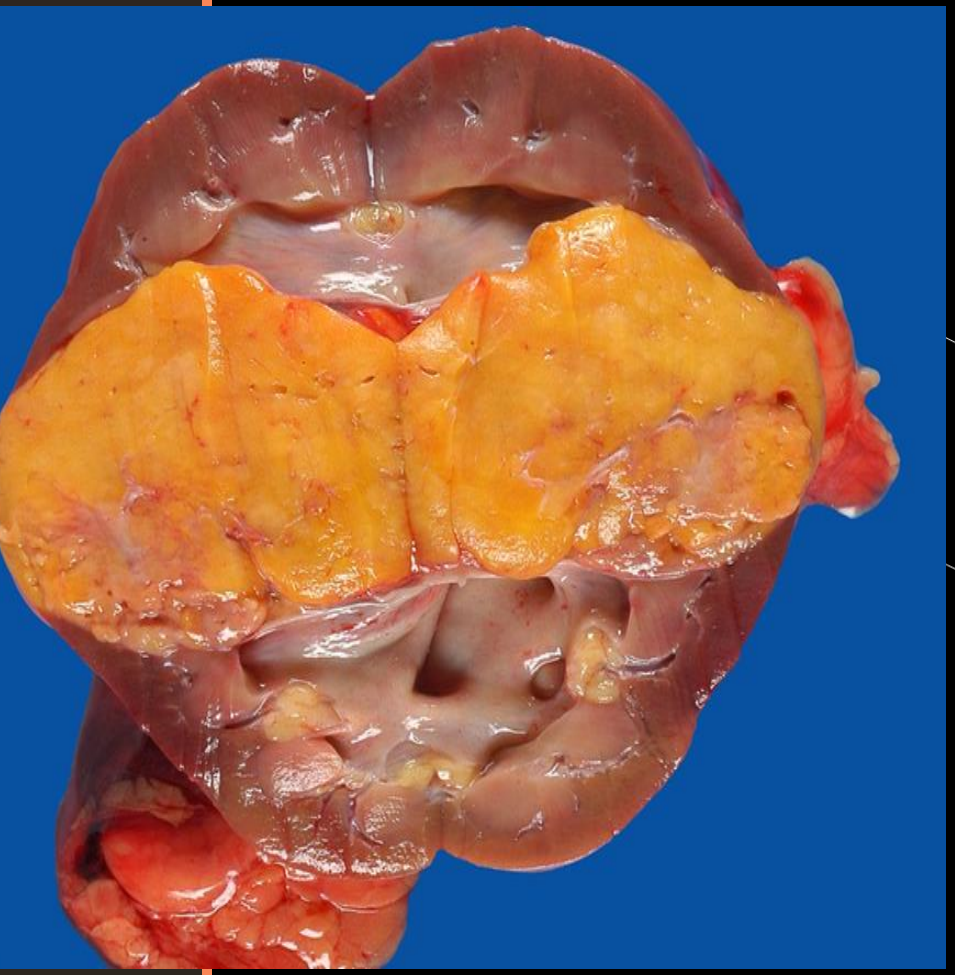


Presentation:

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 in up 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.

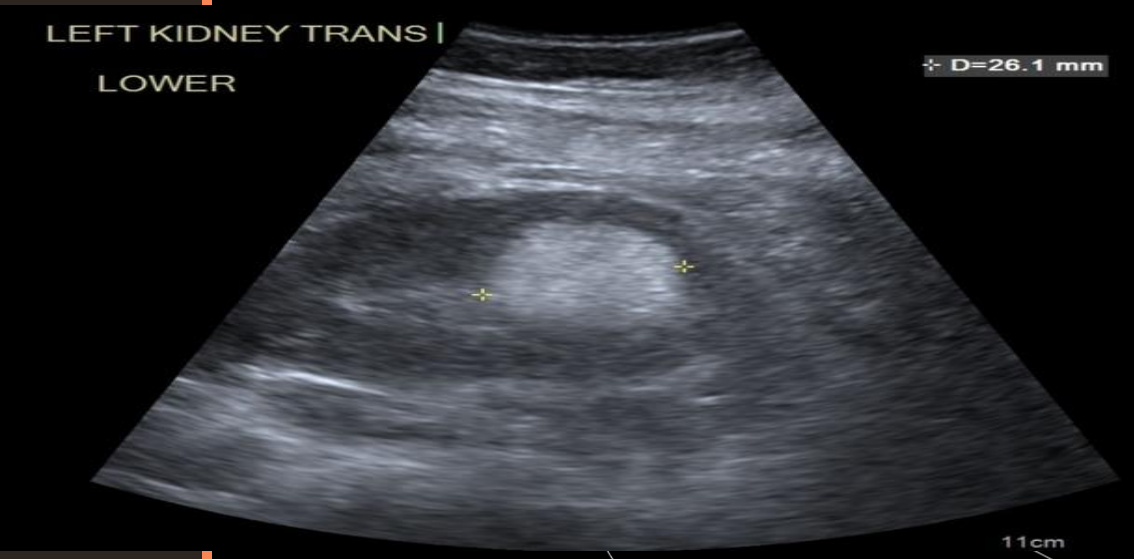
Pregnant women appear to be at an increased risk for hemorrhage .

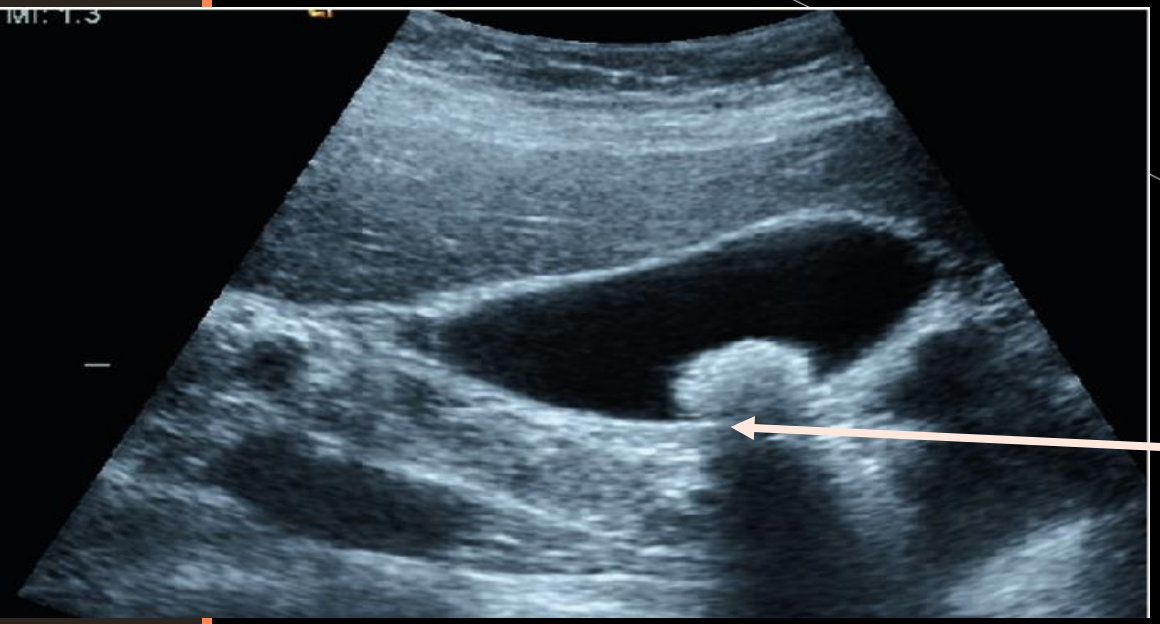


AML is composed of PERIVASCULAR epithelioid cells (PEC)

Gross appearance:

- non-capsulated
- oval, yellow to gray tumor
- a well circumscribed lump of fat





➢ Investigation :

Ultrasound

Homogenous
Well- defined
characteristic hyperechoic ;
bright echo pattern (fat)

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

ANGIOMYOLIPOMA

IF LARGE

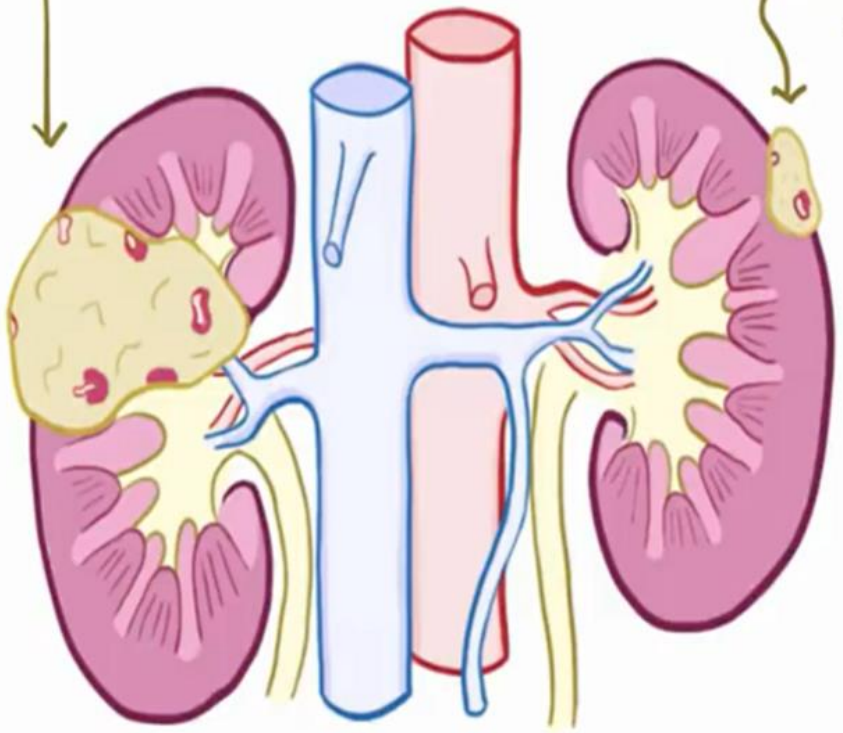
MASS EFFECTS may IMPAIR FUNCTION

↓

CHRONIC KIDNEY DISEASE

↓

END-STAGE RENAL DISEASE



IF SMALL

* usually HARMLESS
* do not require TREATMENT

MANAGEMENT

asymptomatic AMLs if size <4 cm

Follow up with serial US

symptomatic 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 .

ANGIOMYOLIPOMA

ANGIOMYOLIPOMAS have a high density of BLOOD VESSELS

↓

difficult to RESECT

alternatively DESTROY the BLOOD SUPPLY with EMBOLIZATION

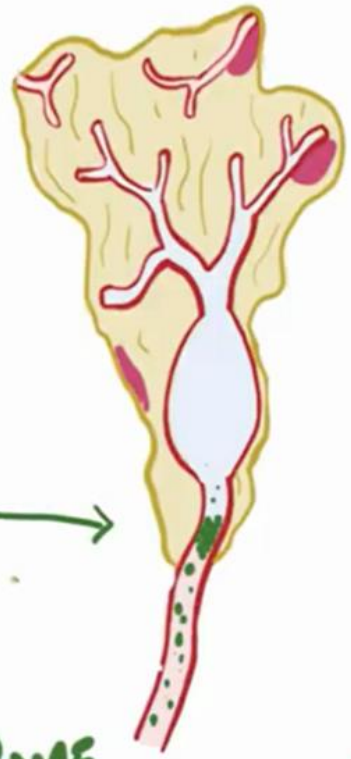
↓

TUMOR can become NECROTIC & SHRINK

↓

POSTEMBOLIZATION SYNDROME

└ fever, flank pain & malaise



LARGE TUMORS can develop IRREGULAR BLOOD VESSELS

↓

that can DILATE

↓

forming an ANEURYSM

↓

that can RUPTURE

↓

can cause FLANK PAIN & HYPOVOLEMIC SHOCK (esp. if near major BLOOD VESSEL)

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 neoplasm)
2. renal tubular adenoma
3. alveolar renal adenoma

7 – 23 % at autopsy

M:F 3:1

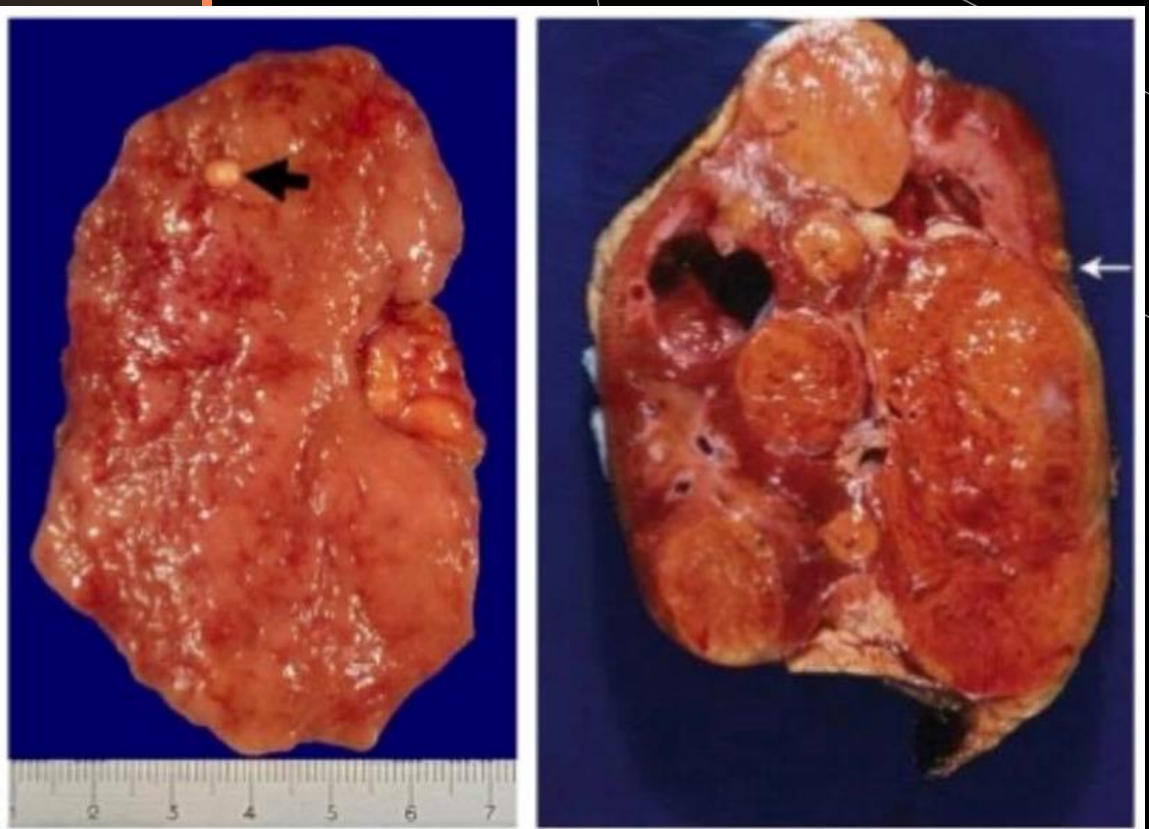
Cytogenetics : which may exhibit trisomy of chromosome 7 & 17

Rarely symptomatic

Grossly :

Solitary cortical solid ,non-capsulated pale yellow-gray discrete nodules < 2 cm

(Majority are solitary but may be multifocal)



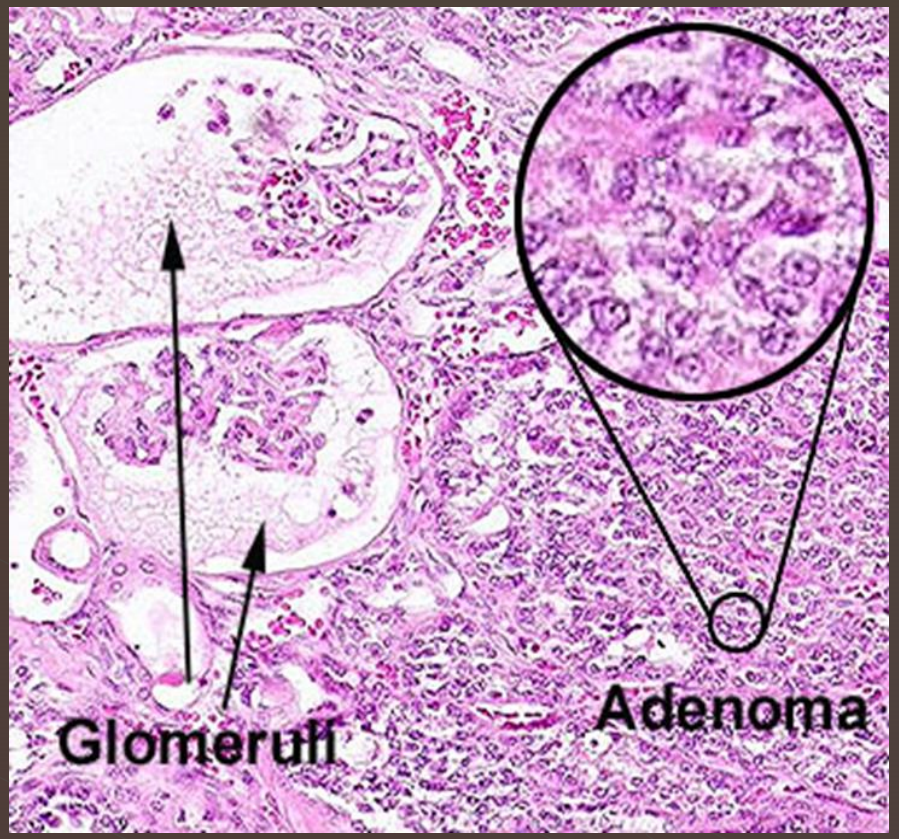
Adenoma cells look much like **low-grade RCC** cells under a microscope. In fact, while they are considered benign, there is presently no known cellular classification to differentiate them from RCCs. Many researchers and physicians regard them as early stage precancers, to be treated accordingly.

Controversy as to whether this represent benign or pre-malignant neoplasm

Incidental finding on CT

Histologically :

organized cells with
no <u>or</u> little atypia



Other rare tumors:

- Leiomyoma
- Hemangioma
- Lipoma
- Juxtaglomerular cell tumors
- Fibroma
- Rhabdomyoma

Malignant renal tumors

- Renal cell carcinoma
- Nephroblastoma (wilm’s tumor)
- Neuroblastoma

Renal cell carcinoma (hypernephroma/ Grawitz’s tumor)

- 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 :**

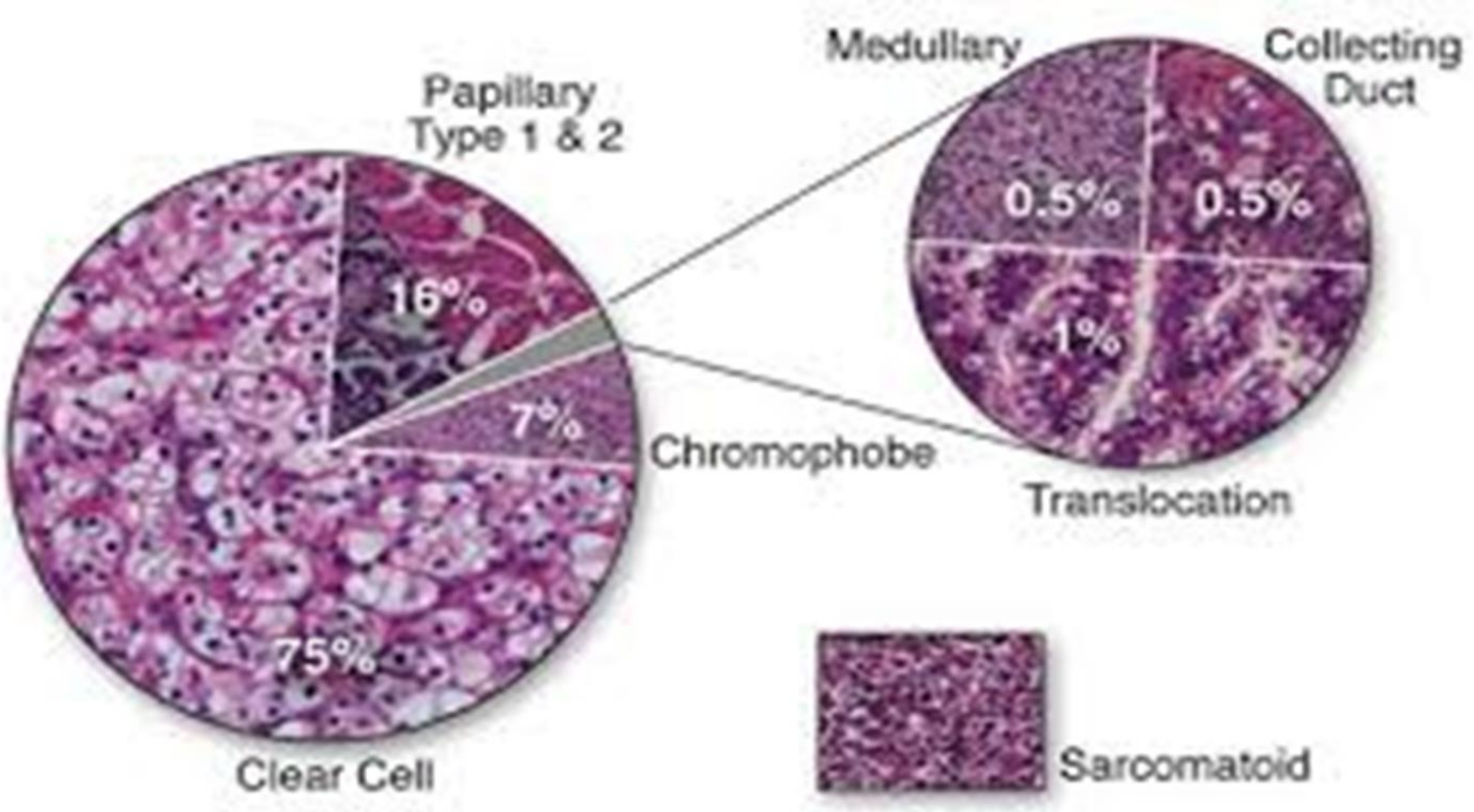
- Usually **unilateral** mainly from upper pole.
- Bilateral tumor (1-2%)
  - Von-Hippel Lindau disease
  - hereditary papillary renal carcinoma

- **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 differentiated variant of any type, it is <u>carcinoma</u> !!

- **Macroscopic picture :**

- **Mass** (mainly from upper pole of kidney) **infiltrating edge**
- area of hemorrhage ,necrosis and scarring
- **golden yellow color** due to **high lipid content**
- They are usually circumscribed by a **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 stages, 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 tumor may already be large and metastatic.

- Gross/microscopic hematuria (most common manifestation)
  - Gross occurs less commonly microscopic. Blood visible throughout the stream suggests an origin in the upper urinary tract, as do vermiform clots.
- 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

Only 10% of patients will present with the classic triad of flank pain, hematuria, and flank mass. If present, it usually indicates advanced disease.

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.

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)

Non-endocrine

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

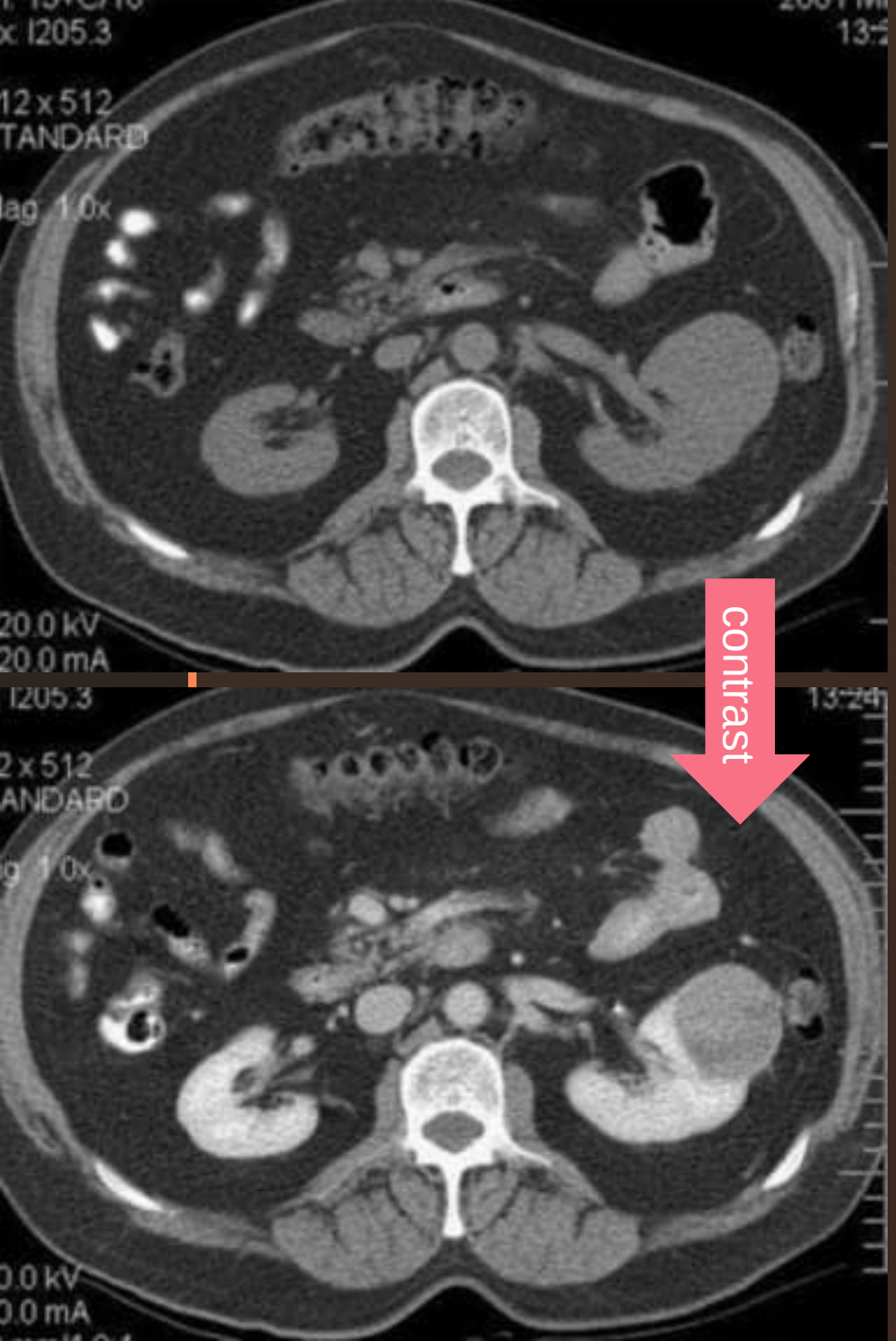
Most paraneoplastic syndromes associated with localized RCC are definitively treated with nephrectomy only; the recurrence of a previous paraneoplastic syndrome should alert the physician to possible disease progression.

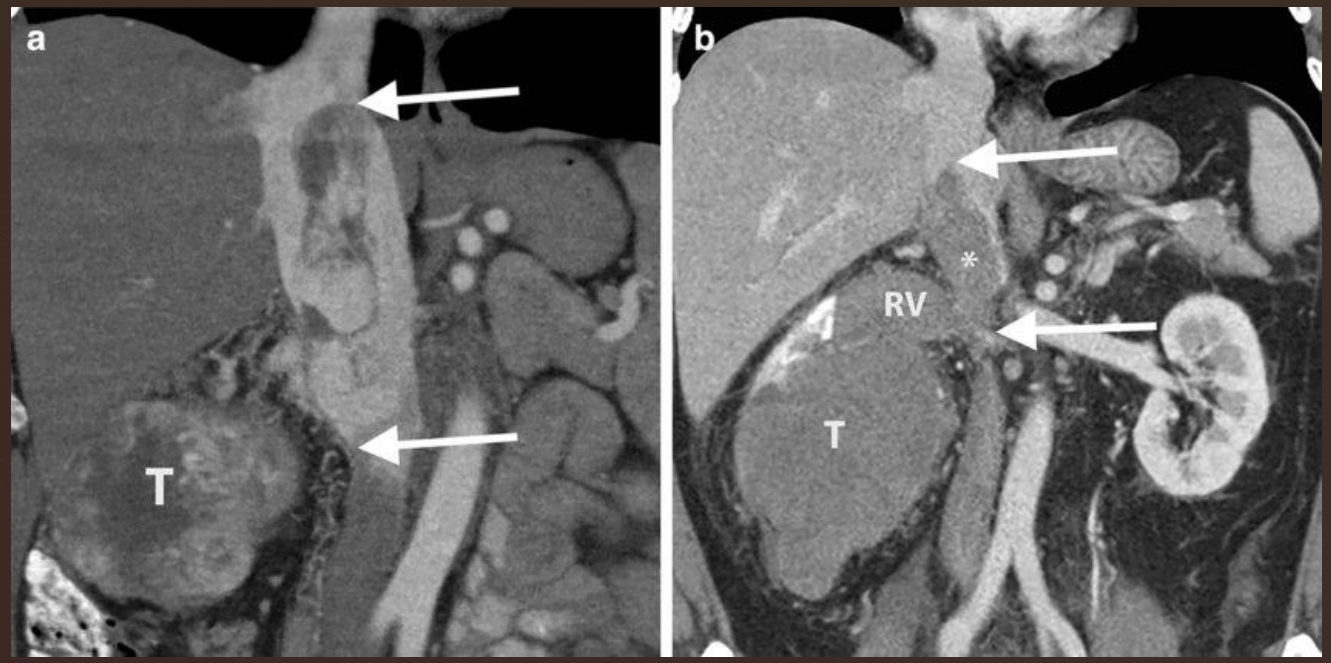
Labs

- Urinalysis → microscopic hematuria, cancer cells
- Urine cytology/culture should be normal
- CBC with differential → erythrocytosis/polycythemia, anemia
- Electrolyte panel → hypercalcemia
- Kidney function tests → creatinine (and BUN) unaffected unless both kidneys are affected
- Liver function tests → aspartate aminotransferase, alanine aminotransferase, alkaline phosphatase, and prothrombin time (elevation of at least 3 required to diagnose Stauffer syndrome)
  - Alkaline phosphatase elevation indicates bone scan
- Needle biopsy does not have sufficient sensitivity when findings are equivocal; it is recommended only when the diagnosis would impact treatment choice,

Imaging

- Ultrasound has mostly been replaced by CT, but it can be useful in evaluating questionable cystic renal lesions if CT imaging is inconclusive.
- Abdominal/pelvic CT scan (with+without contrast): imaging modality of choice
  - Upon contrast-enhancement: RCC is usually solid, and decreased attenuation suggestive of necrosis is often present. Sometimes, RCC is a predominantly cystic mass, with thick septa and wall nodularity. RCC may also appear as a completely solid and highly enhancing mass.
  - If a solitary mass is enhancing, the degree of confidence in diagnosing RCC is high. When a mass is predominantly cystic, the confidence level decreases. In these patients, US may be useful.
- MRI
  - When inferior vena cava involvement is suspected, either inferior venacavography or magnetic resonance angiography (MRA) is used. MRA is currently the preferred imaging technique. Knowledge of inferior vena cava involvement is important in planning the vascular aspect of the operative procedure.
  - Also used as an alternative to CT in cases of pregnancy or when contrast is contraindicated due to allergy or renal insufficiency
  - Brain MRI, if clinically indicated
- Chest X-ray: if abnormal→chest CT indicated


contrast


a
T
b
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Grading: Fuhrman System

Nuclear grading system that concurrently evaluates nuclear size+shape and nucleolar prominence to assess aggressiveness of 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

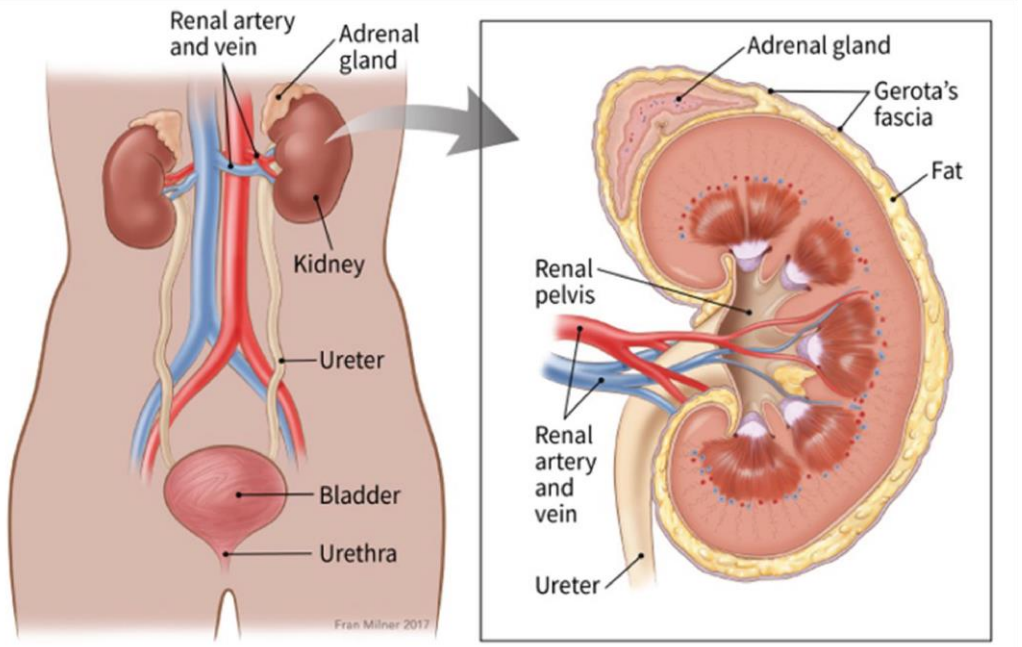
Well-differentiated →
Moderately-differentiated →
Poorly-differentiated →

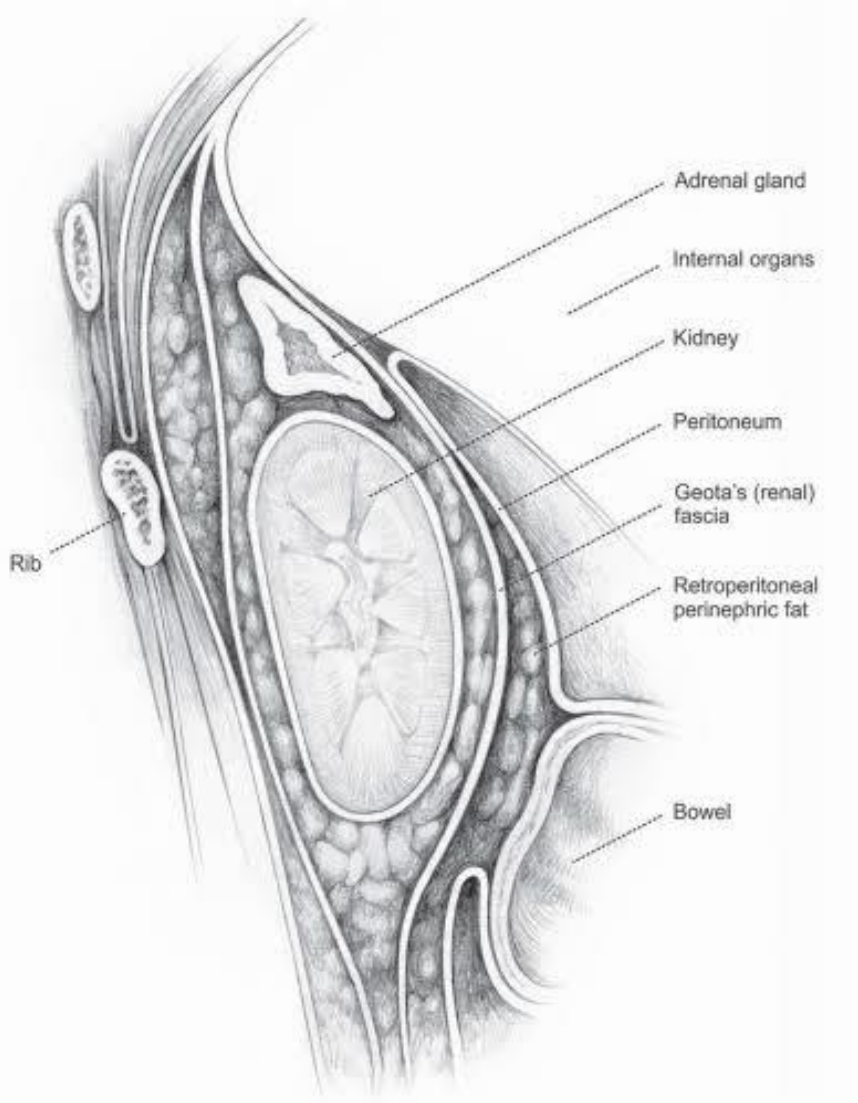
Grading system	Fuhrman (AJSP, 1982) (historic)		WHO / ISUP (2022) (current)	
	Nuclei (shape & size)	Nucleoli	Nuclei (shape & size)	Nucleoli
Grade 1	Small, round, uniform (10 um)	Absent / inconspicuous	-	Absent / inconspicuous at 400x
Grade 2	Larger (15 um)	Visible at 400x	-	Eosinophilic and visible at 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: TNM

Tumor (T) describes the size of the tumor and how far it's grown in the nearby area

T Category	Extension	T Criteria
T1	Limited to kidney	a. ≤ 4 cm in greatest dimension
		b. > 4 cm but ≤ 7 cm in greatest dimension
T2	Limited to kidney	a. > 7 cm but ≤ 10 cm in greatest dimension
		b. > 10 cm in greatest dimension
T3	Extension within Gerota's fasica, no adrenal gland involvement	a. Extends into renal veins or its segmental branches or invades perirenal and/or renal sinus fat
		b. Extends into the part of the vena cava that is within the abdomen
		c. Extends into the part of the vena cava that is within the chest or it is growing into the wall of the vena cava
T4	Extension beyond Gertoa's facsia (the fibrous layer that surrounds the kidney and nearby fatty tissue) +/- adrenal gland involvement (on top of the kidney).	


Renal artery and vein
Adrenal gland
Kidney
Ureter
Bladder
Urethra
Fran Milner 2017
Adrenal gland
Gerota's fascia
Fat
Renal pelvis
Renal artery and vein
Ureter


Adrenal gland
Internal organs
Kidney
Peritoneum
Geota's (renal) fascia
Retroperitoneal perinephric fat
Rib
Bowel

Node (N) describes whether cancer has spread to the regional lymph nodes

N Category	N Criteria
N0	None
N1	Present

Metastasis (M) describes whether the cancer has spread to a different part of the body.

M Category	M Criteria
M0	None
M1	Distant metastasis

Stage	T	N	M
I	T1	N0	M0
II	T2	N0	M0
III	T3	N0	M0
	T1-T3	N1	M0
IV	T4	Any N	M0
	Any T	Any N	M1

Staging is very important for prognosis. It is also important for determining treatment modality.

- For early RCC, surgical treatment, active surveillance, or thermal ablation
- For 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

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

Secondary Renal Tumors

The most common cancers that metastasize to the kidney are melanomas and solid tumors, particularly:

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

Albuminuria and hematuria are common

Usually latent--pain and renal insufficiency are rare