2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

CASE – 1

- 2 year old child presented with complaints of mass per abdomen x 1 month, gradually progressive in size - no associated pain over the swelling or sudden increase in size
- No h/o haematuria.
- O/E ,Right upper abdominal fullness

➢Umbilicus pushed downwards

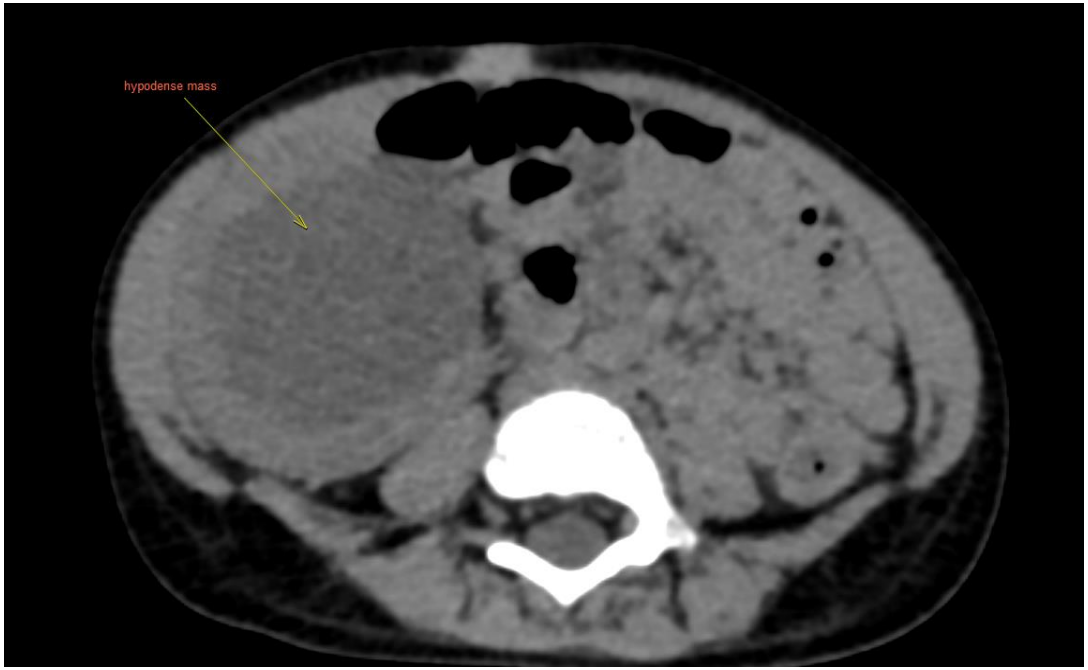
➢Skin over the swelling normal

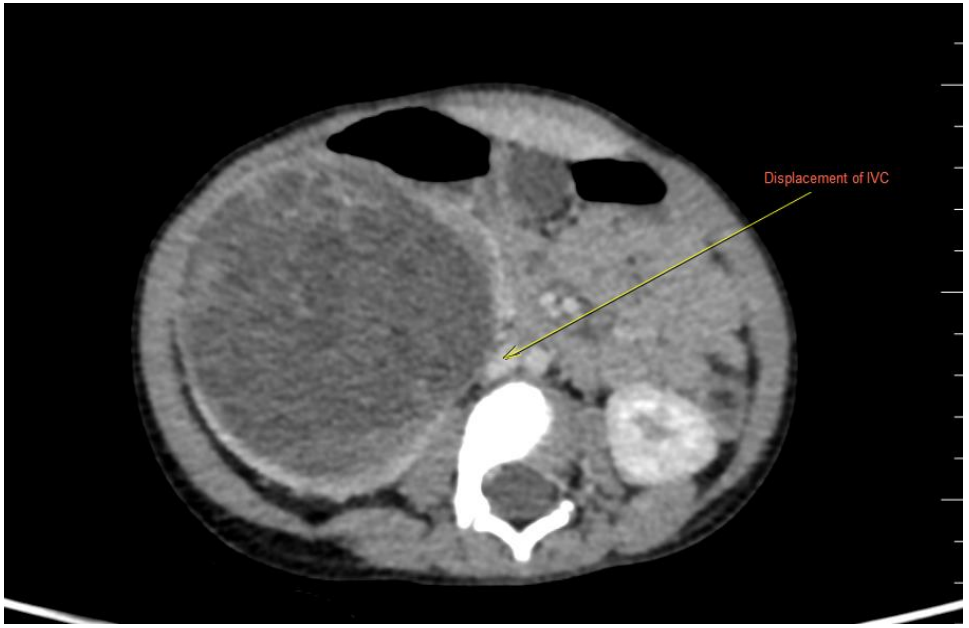
➢Firm to hard solitary lesion in the right hypochondrium extending from - 5th intercostal space till 4cm above the right iliac spine (measuring 18cm) - from posterior axillary line to nearly the midline.

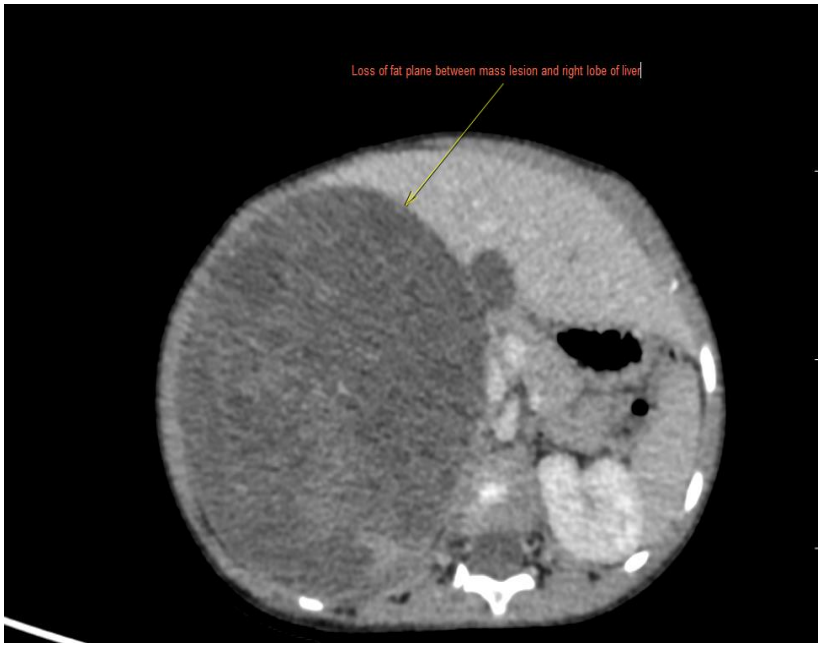
- Advised for CECT abdomen

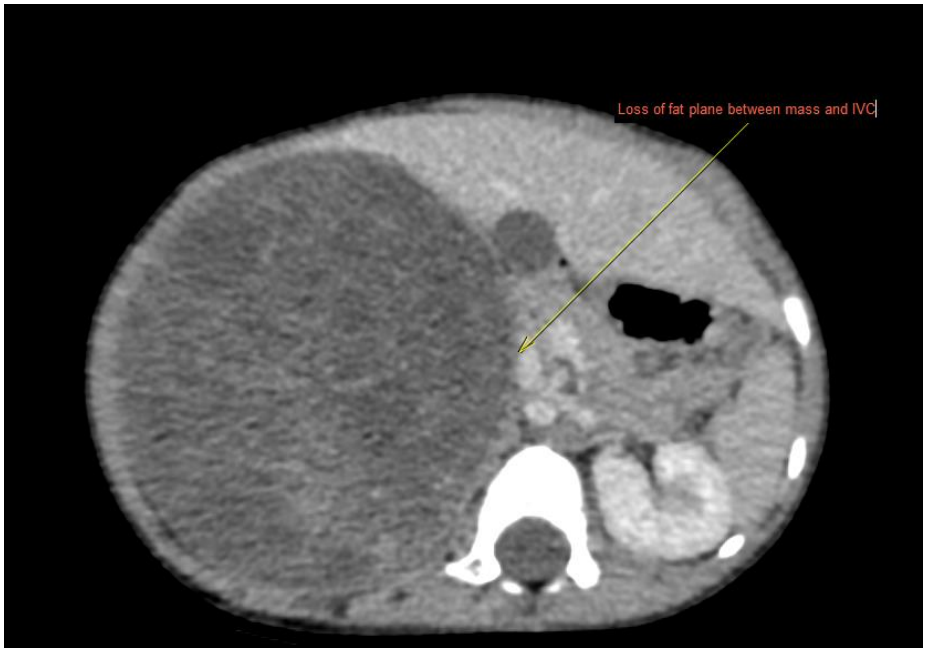
SUMMARY:

- Large well-defined heterogenously enhancing hypodense mass lesion with thick septations arising from the right kidney with mass effect noted in the form of displacement of bowel loops and IVC to left side.
- There is loss of fat plane between the mass lesion and right lobe of liver and IVC.
- No definite invasion into the distal right renal vein and IVC seen.
- No other similar density mass lesion is noted in both kidneys.


hypodense mass


Displacement of IVC


Loss of fat plane between mass lesion and right lobe of liver


Loss of fat plane between mass and IVC

DIFFERENTIALS:

- Wilms tumour
- Neuroblastoma
- PNET
- Clear cell sarcoma
- Renal cell carcinoma
- Leukemia
- Lymphoma

DEPARTMENT OF LABORATORY MEDICINE

MRN	: **10020001343258**	**Department**	: PAEDIATRIC SURGERY
Name	: **Master ABHI**	**Specimen**	: **BODY TISSUE**
Age / Sex	: 2 Year(s) / Male	**Visit Type**	: IP
Sample No	: **M22205020034**	**Collected On**	: 02/05/2022 05:31 PM
Consulting DR	: Drs Sanjay Rao/Ashley J D Cruz/Zameer M M/Vinay Chandrashekar	**Referred DR**	: Dr. Basavaraj
		Received On	: 02/05/2022 09:57 PM
Patient Address	: KALKERI, Bijapur Kalkeri Karnataka,India	**Reported On**	: **07/05/2022 08:34 AM**
		Report Type	: **Final**

HISTOPATHOLOGY	
REGULAR HP SMALL BIOPSY	
Lab No	H-2886/2022
Specimen details	Right renal mass biopsy.
Clinical Details	Right renal mass. ? Wilms tumor. HPE.
Gross Examination	Three tissue cores range from 0.6 cm to 1.0 cm in length. Entire tissue processed in one capsule.
Microscopy and Impression	These cores show areas of loose and dense collagenous stroma with variably cellular bland spindle cells and areas containing cells showing rhabdomyoblastic differentiation. There are some scattered crushed groups of blue cells, which are likely to represent blastemal areas. Scattered variable sized tubular / cystic structures lined by flattened cells are seen also admixed (? vessels or epithelial component). COMMENT - The findings are consistent with a Wilms tumour (nephroblastoma). Immunohistochemistry can be done for confirmation and is available after payment for 6 to10 markers. Also correlate with clinical and radiological findings.

Note:
All histopathology reports need to be correlated with clinical findings.
Additional remaining surgical specimens will be preserved for a period of 3 months.

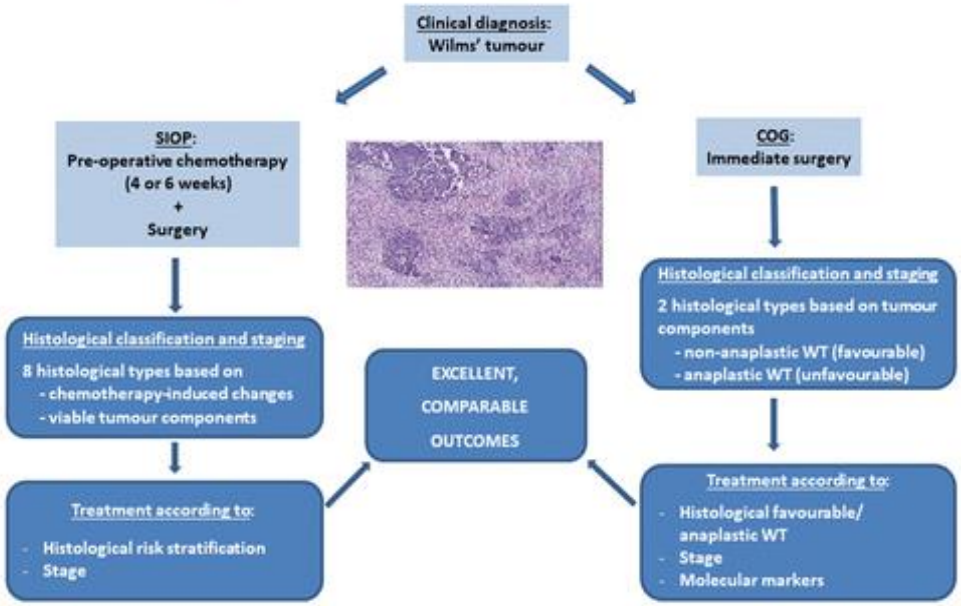
-- End of Report --

Result Entered by

Dr. Srinivas Ramaiah
Consultant
MD Histopathologist

Pathology of Wilms' tumour in International Society of Paediatric Oncology (SIOP) and Children's Oncology Group (COG) renal tumour studies: similarities and differences

Patients with Wilms' tumour (WT) are treated according to two different approaches which contain different criteria for WT histological sub-classification and staging, but their outcomes are remarkably similar



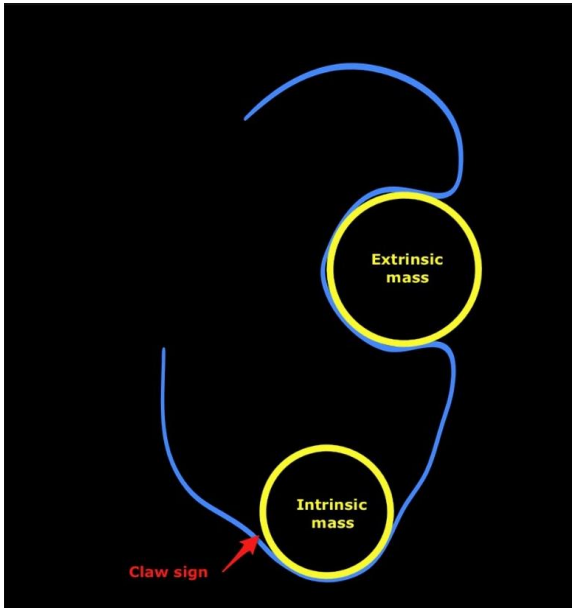
MANAGEMENT:

- Started siop protocol on 14-05-22. 4 cycles of chemotherapy (Vincristine and Actinomycin D).
- CT scan on 11.06.2022 showed no change in the size of the mass .Hence, complete surgical excision (Right radical nephroureterectomy) done on 13-07-22.

1.Renal Vs extra-renal mass

➢CLAW SIGN

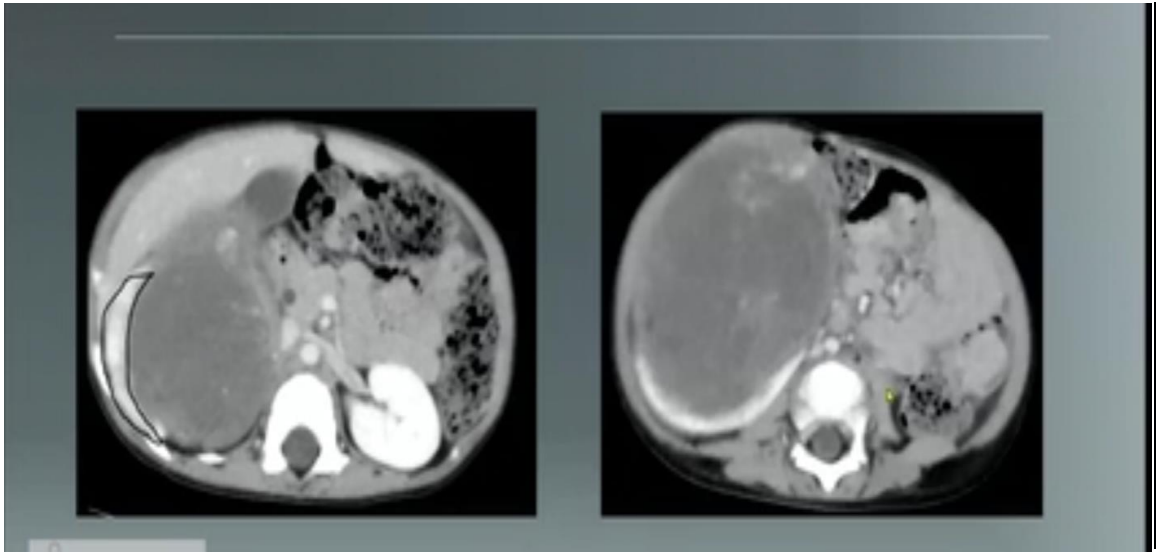
- Useful in determining that a mass arises from a solid structure rather than is located adjacent to it and distorts the outline.
- It refers to the sharp angles on either side of the mass, which the surrounding normal parenchyma forms when the mass has arisen from the parenchyma



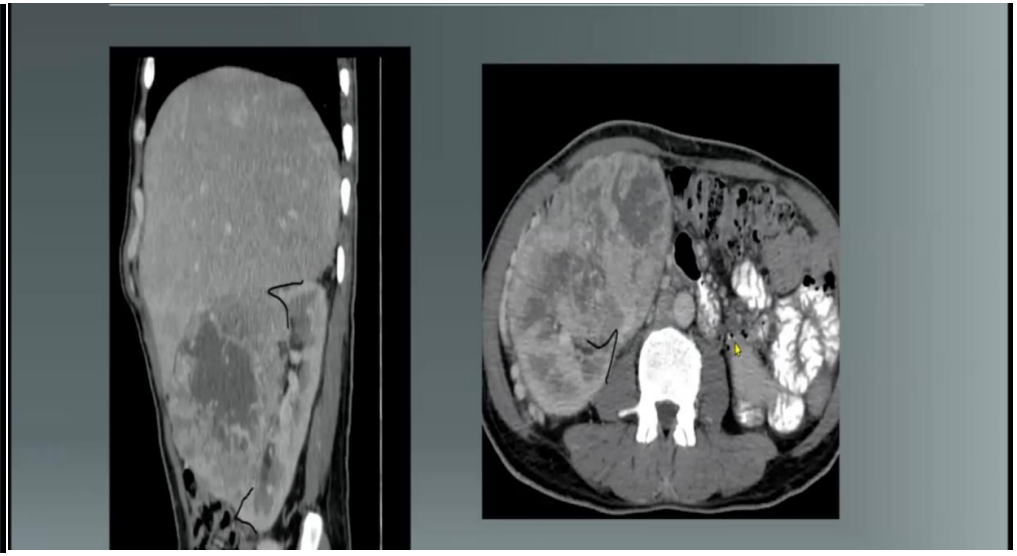
Positive signs:

- Crescent sign
- Beak sign
- Prominent feeding renal artery sign

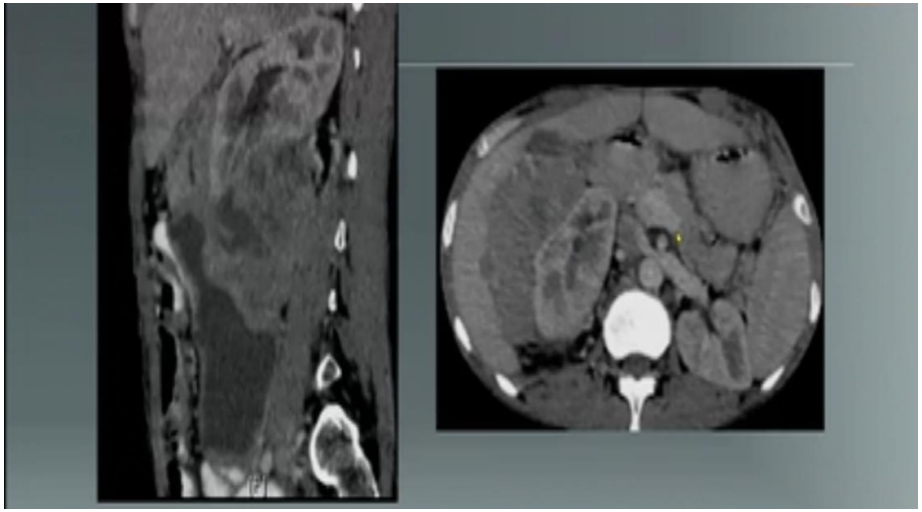
CRESCENT SIGN –Kidneys are Compressed like crescent

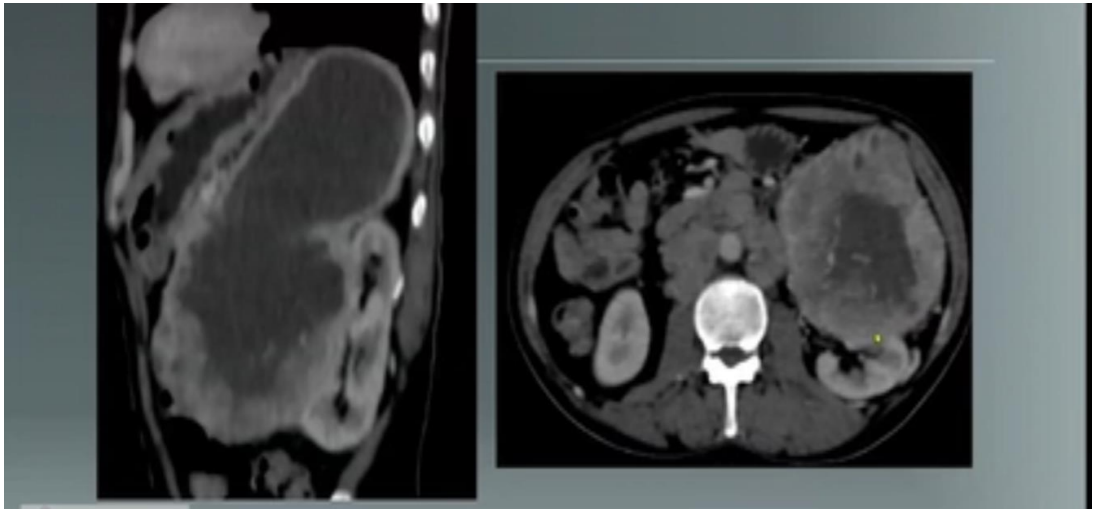


BEAK SIGN-Beak like projection from kidney impinging on the tumour



- NEGATIVE SIGNS:
- INTEGRITY OF KIDNEY-If the integrity of kidney is maintained ,it rules out renal mass.
- RENAL DISPLACEMENT:R/O renal mass





CLASSIFICATION OF PAEDIATRIC RENAL MASSES:

- Normal anatomy: Mimicking a mass-column of bertin, fetal lobulations
- Benign conditions: Simple renal cyst, calyceal diverticula
- Primary benign neoplasms: Mesoblastic nephroma,multilocular cystic renal tumour, Angiomyolipoma, Ossifying renal tumour of infancy.
- Primary malignant neoplasm
- Secondary malignant neoplasm

- CLASSIFICATION OF PEDIATRIC RENAL MASSES

Benign neoplasm

- Congenital mesoblastic nephroma
- Multilocular cystic renal tumour
- Angiomyolipoma
- Renal ossifying tumour of infancy
- Nephroblastomatosis

Primary malignant Neoplasm

- Wilms tumour
- Renal clear cell sarcoma
- Rhabdoid tumour
- Renal cell carcinoma
- PNET

Secondary involvement by malignant tumour

- Neuroblastoma
- Leukemia
- Lymphoma

WILMS TUMOUR:

- Most common cause of pediatric renal masses.
- Arise from mesodermal precursors known as metanephrons.
- Peak incidence at 3 to 4 yrs.

CLINICAL FEATURES:

- Palpable mass
- Hematuria,pain,hypertension
- Constitutional symptoms(Fever,anorexia and weight loss)

- Genetic predisposition of WILMS:

1. WT 1 gene

- WAGR syndrome(Wilms tumor,aniridia,GU malformations and mental retardation)
- Denys-drash syndrome(Wilms tumour,male pseudohermaphroditism,progressive glomerulonephritis)

2.WT 2 gene

- Beckwith-wiedemann syndrome (growth disorder, embryonal tumours-wilms tumour,hepatoblastoma,neuroblastoma and rhabdomyosarcoma, visceromegaly,adrenocortical cytomegaly.

Syndromes associated with wilms: Screening at 6 months of age with CT followed by serial USG every 3 monthly upto 7 yrs.

Staging Wilms tumor

Stage 1 (a) Tumor is limited to kidney and is completely resected
(b) The tumor may be protruding into the pelvic system and ureter
(c) The vessels of the renal sinus are not involved
(d) Intrarenal vessel involvement may be present

Stage 2 (a) The tumor extends beyond kidney or penetrates through the renal capsule into perirenal fat but is completely resected.
(b) The tumor infiltrates the renal sinus and/or invades blood and lymphatic vessels outside the renal parenchyma but is completely resected
(c) The tumor infiltrates adjacent organs or vena cava but is completely resected

Stage 3 (a) Incomplete excision. The tumor extends beyond resection margins
(b) Any abdominal lymph nodes are involved
(c) Tumor rupture before or intraoperatively
(d) The tumor has penetrated through the peritoneal surface
(e) Tumor thrombi present at resection margins
(f) The tumor has been surgically biopsied (wedge biopsy) prior to preoperative chemotherapy or surgery

Stage 4 Hematogenous metastases (lung, liver, bone, brain, etc.) or lymph node metastases outside the abdominopelvic region

Stage 5 Bilateral renal tumors at diagnosis

- Imaging features of WILMS:

- Well circumscribed mass due to pseudocapsule
- Enhancement :less than adjacent renal parenchyma
- Heterogenous: Solid with areas of central necrosis and cystic changes
- Calcifications in 15 %(curvilinear / coarse)
- Hemorrhage and fat attenuation may be present.
- Distorts / invade the renal parenchyma and collecting system.
- Vascular invasion of renal vein and IVC
- Spreads by direct extension and displaces adjacent structures and does not encase them.
- Metastasis to lungs (85 %) , liver and regional lymph nodes.

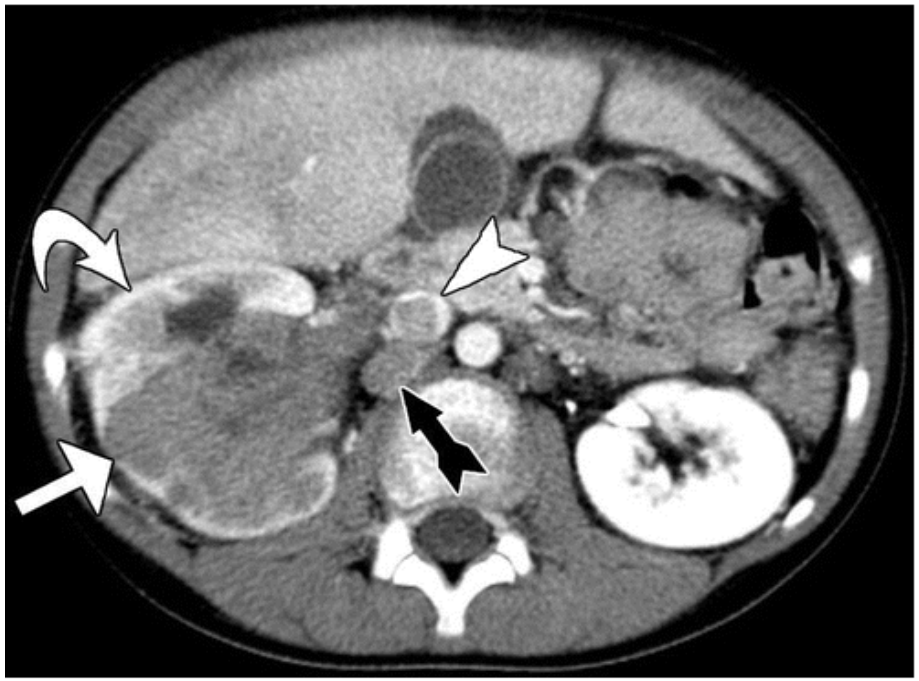


Figure 3a. Wilms tumor with rhabdomyomatous differentiation in a 12-year-old boy. **(a)** Axial postcontrast CT image shows a partially circumscribed slightly heterogeneous tumor (straight white arrow), which is hypoattenuating compared with enhancing renal cortex (curved arrow). Hypoattenuating tumor thrombus surrounded by contrast material is seen in the IVC (arrowhead). Also note the enlarged regional lymph node (black arrow). **(b)** Oblique sagittal image from pulmonary embolism CT shows hypoattenuating tumor thrombus in the right ventricle (arrow) and left lower lobe pulmonary artery (arrowhead).

- WILMS vs NEUROBLASTOMA:

	WILMS	NEUROBLASTOMA
Age	3-4 yrs	1-2 yrs
Claw sign	present	absent
Margins	Well defined due to pseudocapsule	Ill - defined
Calcifications	Curvilinear / coarse	M.C Stippled
Vessels	Displaced rather than encased	Aorta encased and uplifted
Nodes	Present	Conglomerate
Spinal extension	absent	present

- **NEPHROBLASTOMATOSIS**

➢ Nephrogenic rests are foci of persistent embryonal renal tissue, or metanephric blastema, in the kidney after 36 weeks of gestation.

➢ Multiple or diffuse nephrogenic rests are known as nephroblastomatosis.

➢ Nephrogenic rests are found in up to 40% of kidneys resected for Wilms tumor

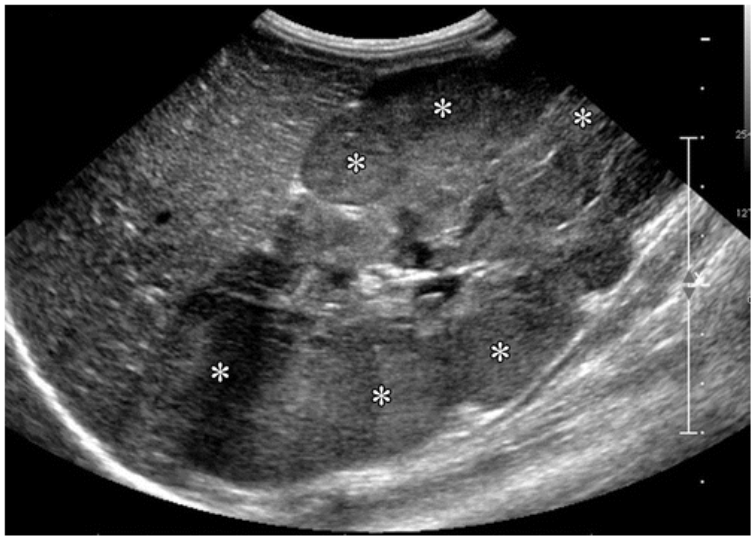


Figure 6a. Diffuse nephroblastomatosis in a 2-year-old boy. **(a)** Longitudinal US image shows multiple round to ovoid peripheral masses (*) that are hypoechoic compared with the central renal parenchyma. **(b)** Coronal contrast-enhanced CT image shows that the masses (*) are hypoattenuating compared with the adjacent parenchyma compressed in the center of the kidney.

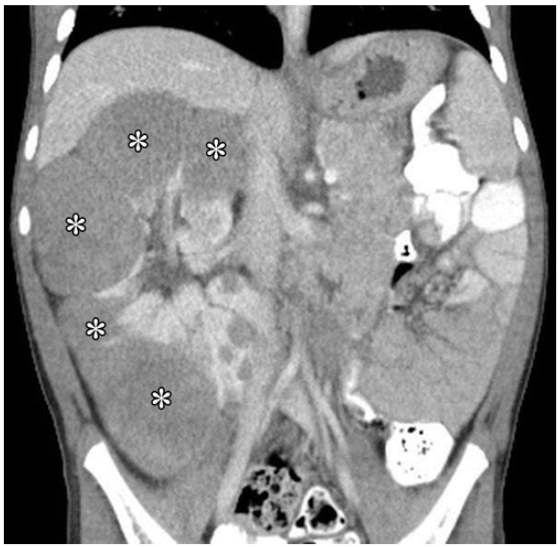


Figure 6b. Diffuse nephroblastomatosis in a 2-year-old boy. **(a)** Longitudinal US image shows multiple round to ovoid peripheral masses (*) that are hypoechoic compared with the central renal parenchyma. **(b)** Coronal contrast-enhanced CT image shows that the masses (*) are hypoattenuating compared with the adjacent parenchyma compressed in the center of the kidney.

- **CLEAR CELL SARCOMA:**

- Rare aggressive mesenchymal neoplasm
- 2nd most common pediatric renal neoplasm.
- Early metastasis common to bones(Uncommon in wilms)
- IMAGING(No definite imaging feature to distinguish from wilms)
- Predominantly solid, heterogenous with cystic /hemorrhagic/necrotic areas)
- Distant metastasis in 5 – 18 % at presentation: Common to lymph nodes and bone
- Calcifications, subcapsular hemorrhage and venous involvement can be seen.

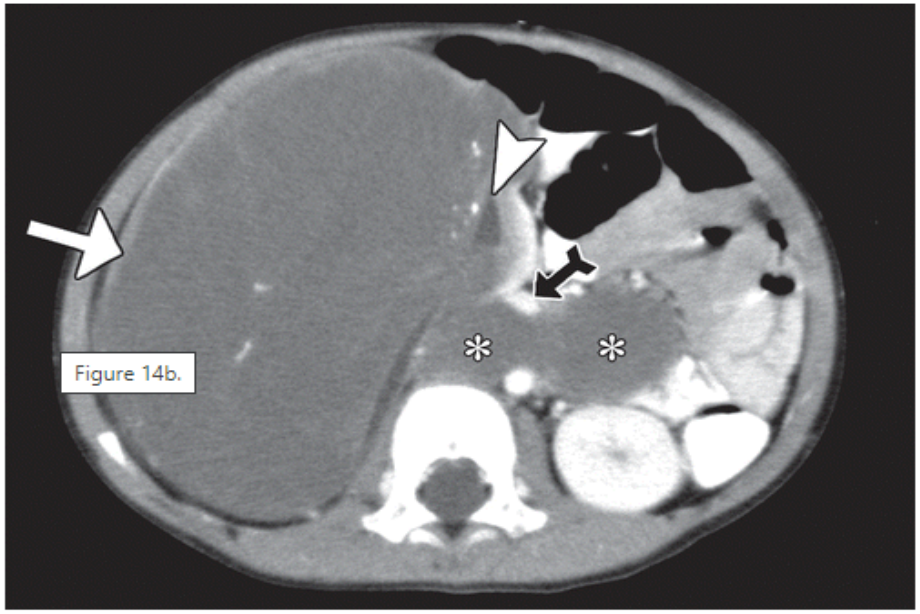


Figure 14b. CCSK in a 2-year-old girl. **(a)** Photograph of the sectioned kidney shows a large mass with a glistening cut surface (straight arrow) and a focal area of necrosis (arrowhead). A small cystic area is also seen (curved arrow). **(b)** Axial CT image shows that the large mass (white arrow) is hypoattenuating compared with the contralateral enhancing kidney, except for a few small calcifications. A focal area of fluid attenuation without enhancement corresponds to necrosis (arrowhead). Enlarged regional lymph nodes (*) separate the aorta from the leftward-deviated IVC (black arrow). **(c)** Axial T2-weighted image shows that the tumor is predominantly hyperintense compared with the contralateral kidney (arrow), except for the dark septa. Small fluid-signal-intensity cysts are seen medially (arrowhead). Involved lymph nodes (*) are similar in signal intensity to the kidney. **(d)** Axial gadolinium-enhanced T1-weighted image shows the tumor and involved lymph nodes (*) enhancing less than the contralateral kidney and lack of enhancement of the cysts (arrowhead). **(e)** Coronal T2-weighted image shows the posterior aspect of the right renal tumor (arrow) and a compression fracture of a thoracic vertebral body due to metastatic disease (arrowhead).

RENAL RHABDOID TUMOUR:

- Aggressive neoplasm
- Hematuria is common presentation.
- **Associations**: Synchronous primary / secondary intracranial neoplasms(Posterior fossa midline tumours)
- **Imaging** :similar to wilms,however more common features are

➢Subcapsular fluid collection d/t subcapsular hemorrhage / necrosis.

➢Multilobulated architexture,indistinct margins,centrally located

➢Curvilinear calcifications like wilms

➢Invasion of renal hilum and pelvis,renal vein & IVC.

➢Adenopathy,lung and liver metastasis are common.

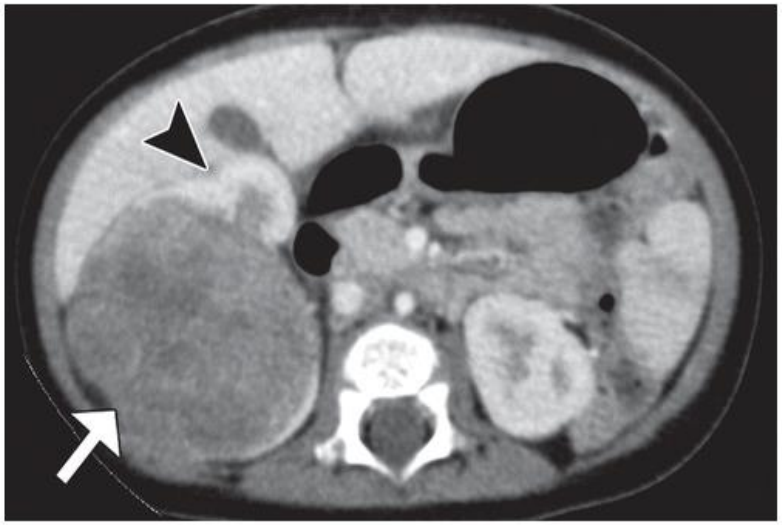


Figure 16d. Renal rhabdoid tumor in a 6-month-old boy. **(a)** Photograph of the bivalved kidney shows a gray-white, lobulated, well-demarcated tumor within the kidney *(K)*. **(b)** Photomicrograph shows discohesive cells with vesicular nuclei. A few rhabdoid cells show eosinophilic cytoplasm displacing the nuclei to one side (arrowheads). (H-E stain; original magnification, ×100.) **(c)** Transverse color Doppler image shows a tumor of heterogeneous echogenicity (arrow) with some internal flow. **(d)** Axial CT image shows a slightly heterogeneous mass (arrow) enhancing less than the adjacent kidney (arrowhead).

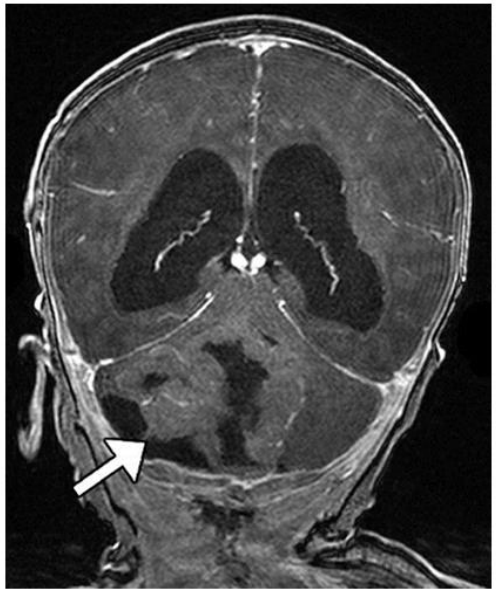


Figure 17b. Rhabdoid tumor in an 11-week-old boy. **(a)** Axial T2-weighted image shows a slightly heterogeneous mass of the left kidney (straight arrow) with a fluid-signal-intensity crescent (arrowhead). Curved arrow = adjacent kidney. **(b)** Coronal spoiled gradient-echo image after intravenous administration of gadolinium contrast material shows a posterior fossa tumor (arrow) with mild enhancement and central nonenhancing necrosis. Hydrocephalus is also seen.

- **Renal cell carcinoma:**
- Common in latter half of 2 nd decade.
- Common histogical subtypes:

➢Xp11.2 translocation RCC-M.C type in children.

➢Papillary RCC:2nd M.C

➢Clear cell RCC-Ass. With Von –Hippel lindau,relatively hypervascular mass.

➢Medullary carcinoma-In adults with sickle cell trait-infiltrative growth pattern and extensive metastasis

PRIMITIVE NEUROECTODERM OF KIDNEY:

Small round cell tumours of kidney

Age :Adolescents and young adults

IMAGING:

- Large size at presentation, diameter generally > 10 cm.
- Ill defined irregular margins: invades renal pelvis,perirenal fat and adjacent organs at the time of initial diagnosis.
- Heterogenous hypoenhancing mass with necrotic and hemorrhagic areas.
- Multiple septum like structures and peripheral hemorrhage.
- May cause tumour thrombus.
- Fine calcification.

Definitive diagnosis: On IHC and cytogenetical analysis-t(11:22)(q24:q12) is highly specific for PNET / EWING's.

- **RCC VS PNET:**

➢ **Pattern of necrosis**: Both are heterogenous(Advanced rcc's has central necrosis and PNETs has multifocal necrosis.

➢ **Enhancement pattern**: PNET shows low or moderate enhancement, in both arterial and venous phases.Clear cell RCC –increased enhancement on arterial phase andlow on venous phase.However,papillary,medullary and chromophobe RCC are hypoenhancing.

➢ PNETs are more aggressive than conventional RCCs, usually present at advanced stage.

Renal synovial sarcoma:

- Spindle cell neoplasm.
- IMAGING:

➢ Well – circumscribed heterogenous mass.

➢ Cystic or necrotic areas within.

➢ Calcifications/subcapsular hemorrhage/venous involvement less common(Usual features of WILMS)

➢ Lymphadenopathy absent.