nailetpek

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rattibha.com

Hi #MedTwitter
A recent case- discussion on #ASPNeph webinar #kidneymass in children intrigued me. Here's what I learned.
#Nephtwitter #tweetorial

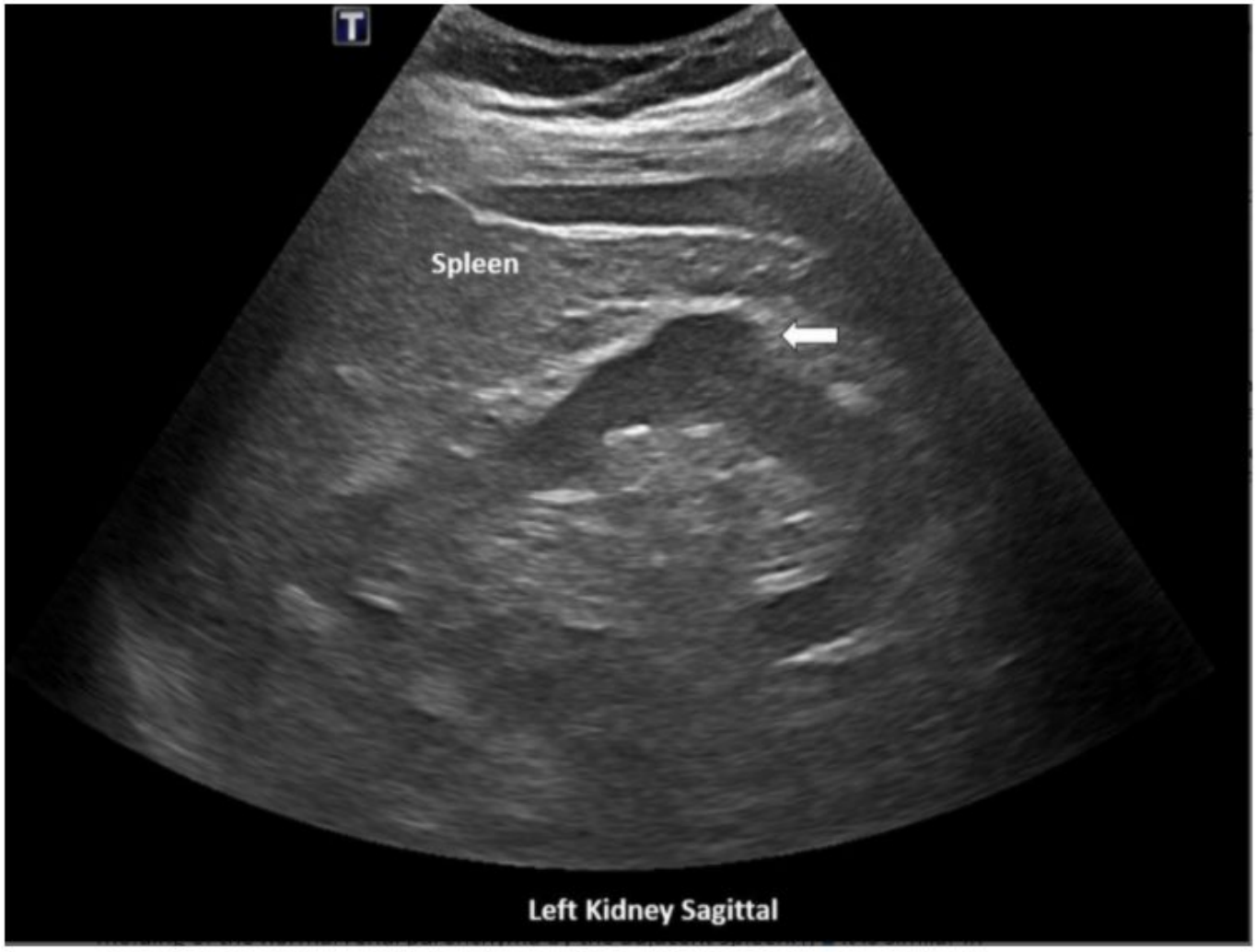
Do you know that most of the intra-abdominal masses originate from kidneys in children! (Potisek et al 2017)
Kidney masses
📌accounts for 60 % of abdominal masses
📌mostly benign and urologic in origin (Nelson 2018)

What is the first step when you recognize a kidney mass in children?

Ans: B &C

💥It is important to exclude pseudo mass/ tumor
💥Most common causes-

📌Congenital anomalies: Prominent renal columns of Bertin, dromedary humps
📌Inflammatory: Focal pyelonephritis, abscess
📌Vascular: Renal artery aneurysm, AV fistula
📌Trauma/ Hematoma



Renal sonogram demonstrating dromedary hump (arrow), which is a focal bulge on the lateral border of the left kidney, caused by molding of the normal renal parenchyma by adjacent spleen
Koratala et al 2018 PMID: 30147922

💥Once pseudo masses are ruled out, differentiating benign from malignant is important

📌Cystic masses-usually benign

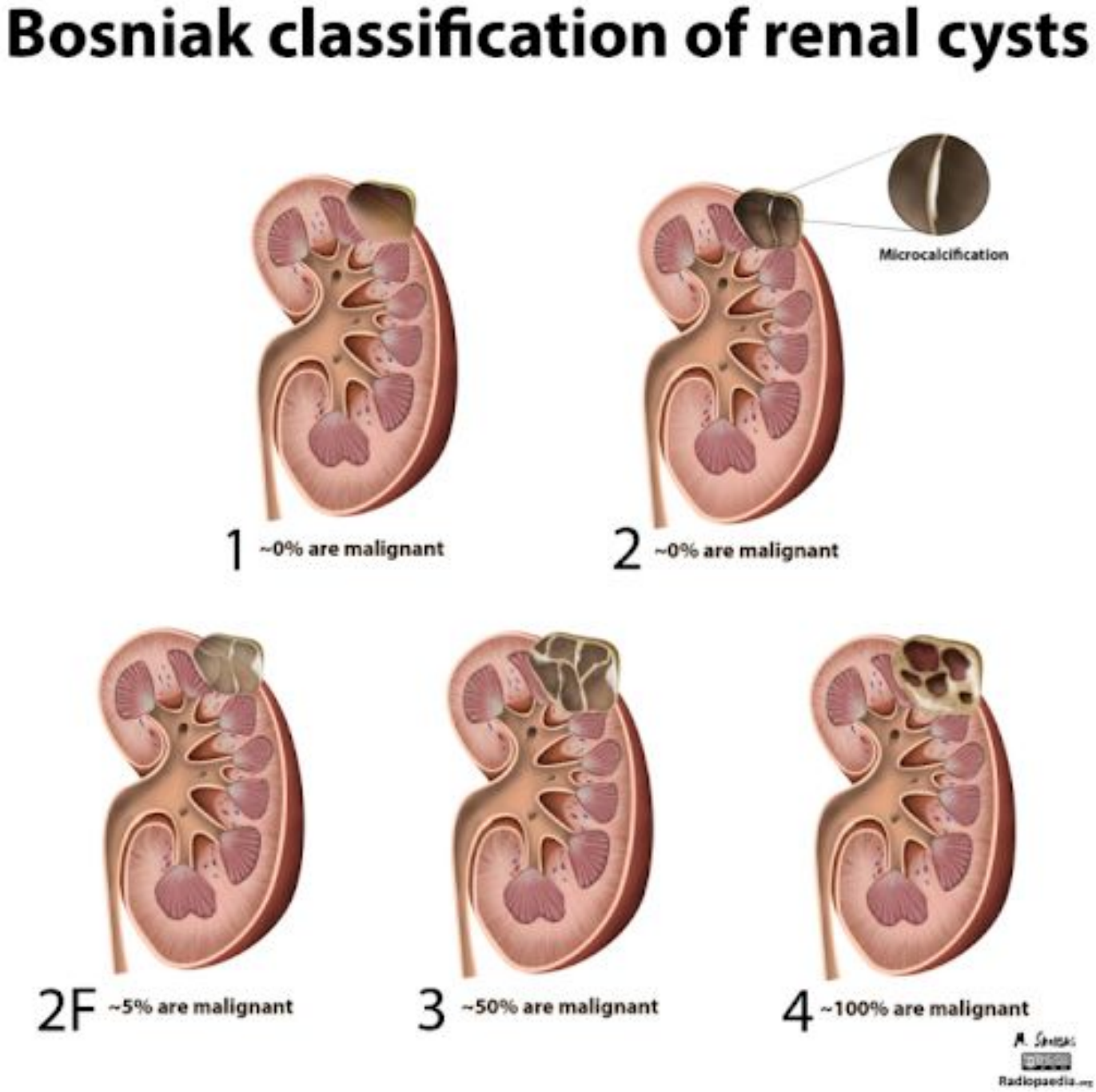
📌Imaging differentiates solid and cystic masses

📌Some cysts can be malignant

📌Bosniak Classification is helpful and is reliable in children

Which of the following has high malignant potential?

Ans: D. Bosniak 3



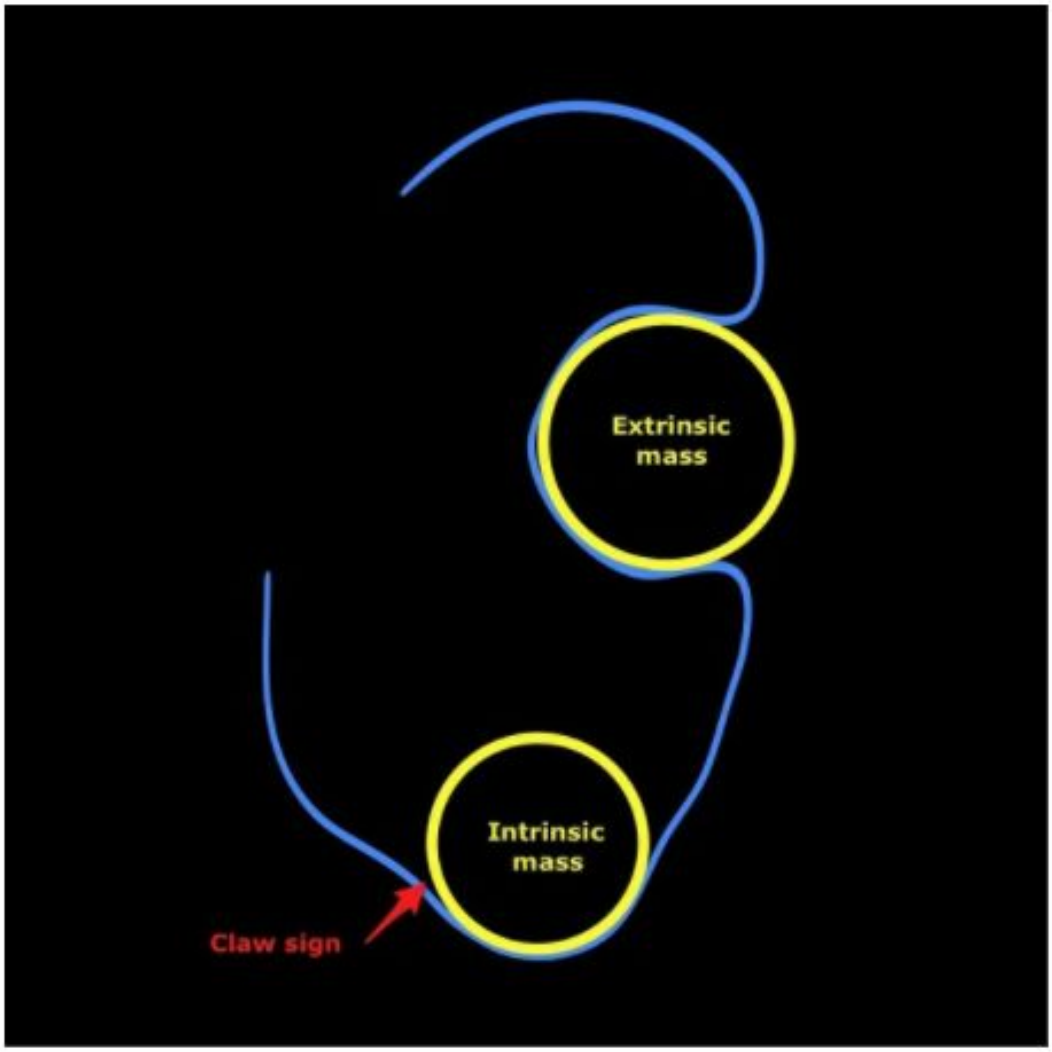
Class	Current Bosniak Classification
I	Hairline-thin wall; water attenuation; no septa, calcifications, or solid components; nonenhancing
II	Two types: 1. Few thin septa with or without perceived (not measurable) enhancement; fine calcification or a short segment of slightly thickened calcification in the wall or septa 2. Homogeneously high-attenuating masses ≤ 3 cm that are sharply marginated and do not enhance
IIF	Two types: 1. Minimally thickened or more than a few thin septa with or without perceived (not measurable) enhancement that may have thick or nodular calcification 2. Intrarenal nonenhancing hyperattenuating renal masses > 3 cm
III	Thickened or irregular walls or septa with measurable enhancement
IV	Soft-tissue components (ie, nodule[s]) with measurable enhancement

Silverman et al, Radiology, Vol. 292, No.2

How to differentiate if a mass is originating from the kidney or not?

Ans: Claw sign

📌Determines if mass arises from solid structures
📌Refers to sharp angles on the either side of mass, formed by the surrounding normal parenchyma

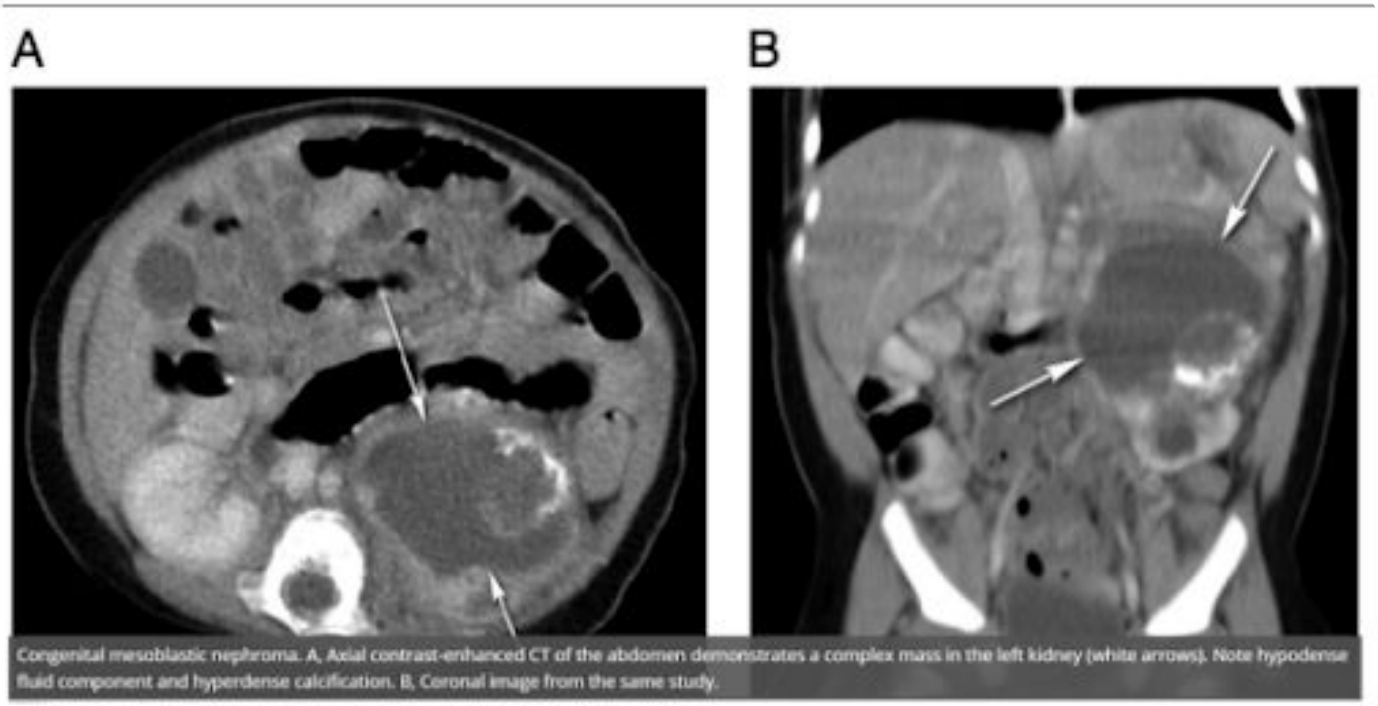


https://radiopaedia.org/articles/claw-sign-mass?lang=us

What is the most common benign kidney tumor in neonates?

Ans:💥 Mesoblastic Nephroma

📌Most common neonatal kidney mass
📌M>F
📌polyhydramnios, hydrops & preterm birth
📌US: the “ring” sign- concentric hyper & hypoechoic rims surrounding tumor
📌Path: interlacing fibro/myofibroblastic cell bundles interspersed with collagen & vessels



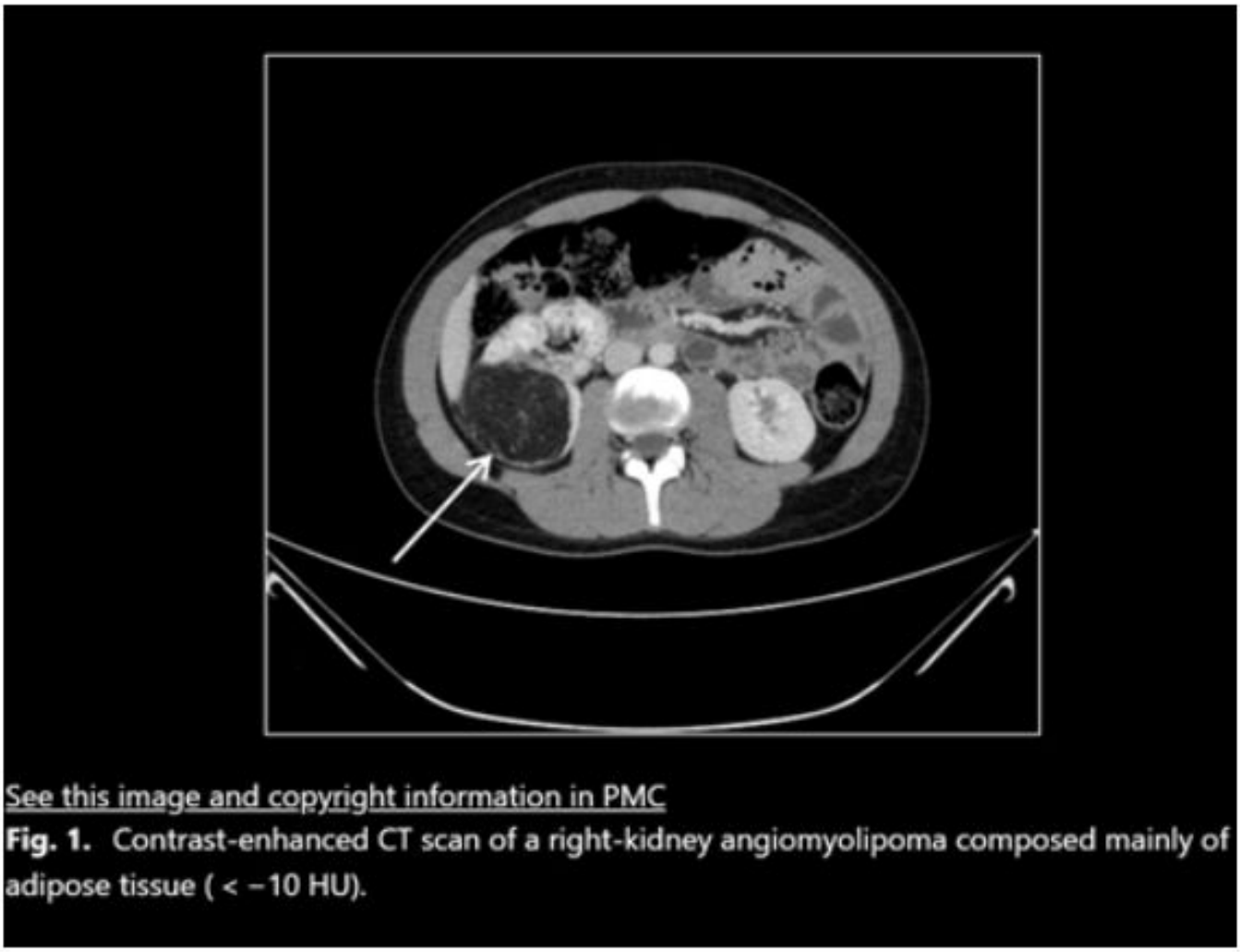
Congenital mesoblastic nephroma. A, Axial contrast-enhanced CT of the abdomen demonstrates a complex mass in the left kidney (white arrows). Note hypodense fluid component and hyperdense calcification. B, Coronal image from the same study.

Malkan AD, Loh A, Bahrami A, Navid F, Coleman J, Green DM, Davidoff AM, Sandoval JA. An approach to renal masses in pediatrics. Pediatrics. 2015 Jan;135(1):142-58. doi: 10.1542/peds.2014-1011. Epub 2014 Dec 1. PMID: 25452658

What is the most common heritable benign tumor?

Ans: Angiomyolipoma

📌Rare Benign renal masses
📌80% with syndromes: TS, VHL, SW, NF, ADPKD
📌Macroscopic fat in CT/MRI
📌asymptomatic & <4cm➡️biannual/ annual follow-up with US/CT
📌Symptomatic or ≥4cm or B/L➡️surgery or arterial embolization



See this image and copyright information in PMC

Fig. 1. Contrast-enhanced CT scan of a right-kidney angiomyolipoma composed mainly of adipose tissue (< −10 HU).

https://pubmed.ncbi.nlm.nih.gov/29225800/

Other benign tumors are-

📌Metanephric stromal tumors
📌Metanephric adenoma
📌Cystic nephroma
📌Ossifying Renal Tumor of Infancy (ORTI)
📌Reninoma

A 2.5 years old boy was brought to the pediatrician because mother noticed right sided abdominal distension. He was asymptomatic. What is the most common kidney tumor in the first decade?

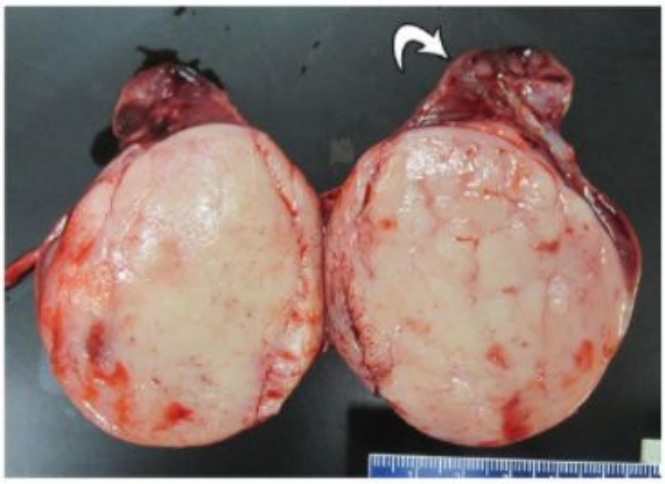
Ans: Wilms tumor

📌90% of childhood kidney cancers
📌2-5 yrs of age
📌90% Sporadic
📌10% Hereditary: WAGR (WT1), Denys-Drash (WT1, 11p13), Beckwith Wiedemann (WT2, 11p15), Perlman, & Sotos
📌unilateral-75-95%
📌Symptoms: Abdominal pain, distention, HTN, malaise or hematuria

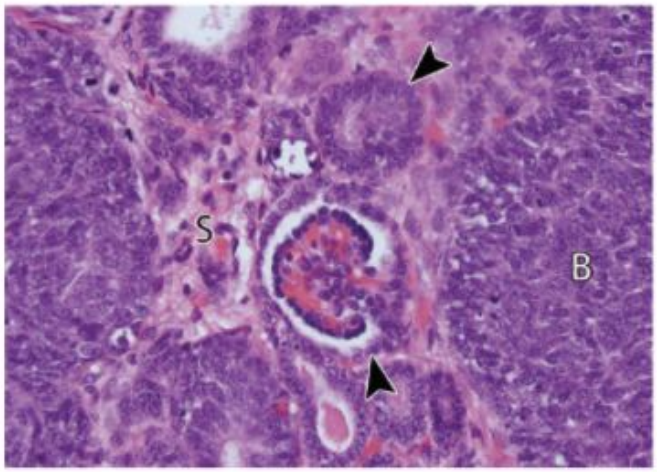
💥 Imaging: Large, well defined intrarenal mass with uniform echogenicity in the US (often with necrosis, old hemorrhage, & Calcification ~15%

💥Path: Triphasic appearance -stromal, epithelial, & blastemal elements

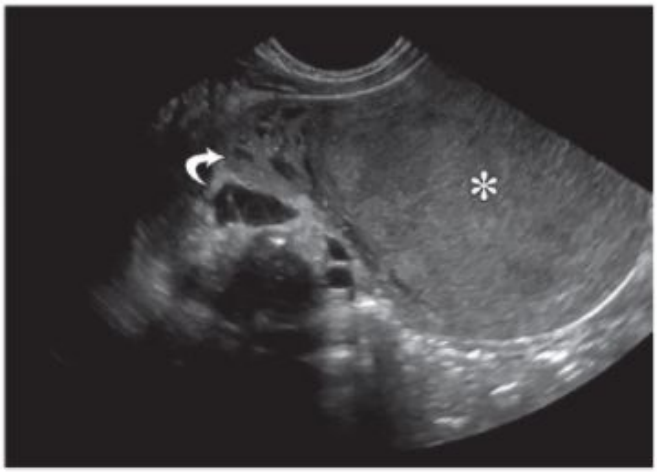
💥Rx: Nephrectomy + adjuvant chemotherapy



a.



b.



c.

Figure 1. Wilms tumor in a 12-week-old infant. **(a)** Bivalved nephrectomy specimen shows a yellow-tan soft cut surface with small foci of hemorrhage. Arrow = adjacent kidney. **(b)** Photomicrograph shows the triphasic pattern of Wilms tumor. Nests of small round to ovoid hyperchromatic cells represent the blastemal component *(B)*. Epithelial elements form tubular and glomeruloid elements (arrowheads). Surrounding these components is an immature spindle cell stroma *(S)*. (Hematoxylin-eosin [H-E] stain; original magnification, ×40.) **(c)** Longitudinal US image shows a large circumscribed tumor of fairly homogeneous echotexture (*), which is slightly hypoechoic compared with the cortex of the adjacent kidney (arrow).

Chung EM, Graeber AR, Conran RM. Renal Tumors of Childhood: Radiologic-Pathologic Correlation Part 1. The 1st Decade: From the Radiologic Pathology Archives. Radiographics. 2016 Mar-Apr;36(2):499-522. doi: 10.1148/rg.2016150230. PMID: 26963460.

Stage	Criteria
Stage I	Confined to kidney Complete excision with renal capsule intact and negative resection margins Lymph nodes negative for Wilms tumor spread
Stage II	Regional extension beyond kidney capsule, but confined to flank May include: Tumor penetration through capsule but confined to Gerota's fascia Infiltration into renal vein Complete excision with negative resection margins Lymph nodes negative for Wilms tumor spread
Stage III	Residual tumor, but confined to abdomen May include: Regional lymph node involvement Peritoneal contamination: Biopsy Pre- or intraoperative tumor rupture Tumor growth through peritoneal surface Positive resection margins
Stage IV	Distant metastases: Lung, liver, bone, brain
Stage V	Involvement of bilateral kidneys at diagnosis

Adapted from Davidoff (2012) [4].

Phelps, H.M.; Kaviany, S.; Borinstein, S.C.; Lovvorn, H.N., III. Biological Drivers of Wilms Tumor Prognosis and Treatment. *Children* **2018**, *5*, 145.

A 14 year old AA boy presents with painless gross hematuria. Which of the following associations should concern you?

Ans: D - s/o renal medullary carcinoma

📌Most common tumor of second decade

📌Sickle cell trait or heterozygous sickle cell disease

📌Painless hematuria, flank pain & palpable mass

📌Overexpression of transcription factor E3 (TFE3):Xp11.2 translocation involves gene fusion

Fig. 1A —17-year-old boy with renal medullary carcinoma. CT image shows right upper pole renal medullary carcinoma. Mass (*arrow*) is infiltrative, and kidney retains its reniform shape.

Renal Medullary Carcinoma: CT and MRI Features

Netta M. Blitman, Robert G. Berkenblit, Alla M. Rozenblit, and Terry L. Levin

American Journal of Roentgenology 2005 185:1, 268-272

Malignant tumors are rare in children (6-7% of all childhood tumors)

Other malignant kidney tumors are-

📌Clear cell sarcoma

📌Nephroblastomatosis

📌Rhabdoid tumor

That's All Folks...

For case-based discussion on this topic logon to @ASPNeph December radiology webinar #FellowFOAMgroup @drM_sudha @RoshanPGeorgeMD @priti899 #pediatricnephrology