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What is the most common pediatric renal tumor?
A Wilms tumor is the most common pediatric renal tumor comprising around 85% of all pediatric renal tumors.

What is the most common neonatal renal tumor?
Mesoblastic nephroma is the most common neonatal renal mass.

True or false? A pediatric neuroblastoma is a pediatric renal tumor.
False. Neuroblastomas involve the adrenal gland and tissues of neural crest origin. Besides adrenal glands, neuroblastomas can occur elsewhere in the retroperitoneum such as the celiac axis or the organ of Zuckerkandl as well as in the posterior mediastinum.

True or false? Renal cell carcinomas are rare in the pediatric population.
True. Renal cell carcinomas rarely occur in the pediatric population. A Wilms tumor is far more common in the pediatric population. When renal cell carcinomas do occur in the pediatric population think older children over 10 years of age and teenagers.

A Wilms tumor is most common at what pediatric age?
Most Wilms tumors occur in early childhood, most typically around ages 3-5 years old.

True or false? Most Wilms tumors are bilateral.
False. Over 90% of Wilms tumors are unilateral.

What is a classic clinical presentation for a Wilms tumor?
Wilms tumors classically present as a painless mass in the upper lateral abdomen with possible associated hematuria and hypertension.

What are some classic genetic syndromes associated with Wilms tumor?
There are many but among the most common genetic syndromes are Beckwith-Wiedemann, WAGR, Denys-Drash, and Sotos syndromes.

What does WAGR stand for in WAGR syndrome?
Wilms tumors
Aniridia
Genitourinary Anomalies
Retardation (mental disability)

What are classic features of Beckwith-Wiedemann syndrome?
Beckwith-Wiedemann syndrome is associated with congenital overgrowth including macroglossia, hemihypertrophy, organomegaly (kidneys, liver, spleen), ear pits, omphalocele, and cardiac, renal, and genitourinary anomalies. Risk of malignancy is elevated with classic associations of Wilms tumors, neuroblastoma, hepatoblastoma, rhabdomyosarcoma, adrenal tumors, pancreatoblastomas, and others.

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What are classic features of Denys-Drash syndrome?
Wilms tumor, congenital nephropathy with glomerulonephritis, and male pseudohermaphroditism.

What are classic imaging findings of Wilms tumors?
Classic imaging features of a Wilms tumor include a large solid renal mass with heterogeneous appearance internally including heterogeneous enhancement and smooth margins that may be partially cystic in appearance with areas of central necrosis if large. Most have no calcifications, but internal calcifications can occasionally be present. CT can sometimes show areas of internal fat density and MRI can sometimes show areas of internal blood products. These cause mass effect on adjacent organs and may manifest on radiography as a soft-tissue, bowel displacing abdominal mass. Invasion and/or extension into the renal vein and/or inferior vena cava is possible.

What are the most common sites of metastases with Wilms tumors?
The most common site of distant metastasis is the lung followed by liver. Local extension into the adjacent renal vein and IVC and sometimes right atrium can be seen and locoregional spread to abdominal lymph nodes is common. Note that skeletal metastases are unusual for a Wilms tumor.

What is the hallmark pathologic finding of nephroblastomatosis?
Persistent nephrogenic rests are a hallmark pathologic feature of nephroblastomatosis. More specifically, persistent nephrogenic rests are sites of metanephric blastema that persist longer than 36 weeks of gestation. This can have multifocal or diffuse involvement of the pediatric kidney.

True or false? Nephroblastomatosis has risk of malignant degeneration into a Wilms tumor.
True. Wilms tumors can arise from sites of nephroblastomatosis. In cases of bilateral Wilms tumors, these nearly always arise from nephroblastomatosis with persistent nephrogenic rests.

What are classic imaging findings of nephroblastomatosis?
If diffuse, nephroblastomatosis manifests on imaging as diffuse reniform enlargement of the kidney with a peripheral rind of thick tissue with striated enhancement characteristics. If focal, one would expect to see multiple peripheral nodules which are hypoechoic on ultrasound, hypodense on CT, low signal on both T1 and T2 MRI, and enhance less than normal renal parenchyma on both CT and MRI. Note that renal lymphoma can appear similarly but would not be expected to occur in neonates and infants to the degree of something like nephroblastomatosis.

What is the classic clinical presentation of a mesoblastic nephroma?
As the most common neonatal renal tumor, the most common presentation of a mesoblastic nephroma is a palpable abdominal mass in a neonate with symptoms such as hematuria being less common. Most of these will be diagnosed within the first 3 months of life and essentially all are found in the first year of life.

What are classic imaging findings of a mesoblastic nephroma?
Like other pediatric renal tumors, x-ray may show a soft tissue mass displacing the bowel. Ultrasound classically shows a circumscribed mass that may have concentric rings of varying echogenicity with some

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rings appearing hypoechoic and others echogenic. Internal vascularity may be noted on color Doppler imaging. If there is internal hemorrhage or necrosis, internal heterogeneity can be seen with ultrasound. CT is typically not performed due to radiation risk in a neonate, but if performed, would show solid hypodense renal lesions without calcifications. MRI classically shows variable T1 signal and variable T2 signal within the renal lesions but would be expected to show at least partial restricted diffusion in the renal tumor(s).

What is the key differentiator between a mesoblastic nephroma and a Wilms tumor?
Age may be the key differentiator between these entities with mesoblastic nephroma occurring in the neonatal period and 1st year of life, whereas Wilms tumors have peak incidence around 4 years of age.

True or false? Nearly all neuroblastomas contain calcifications.
True. Up to 90% of neuroblastomas contain calcifications.

True or false? Nearly all Wilms tumors contain calcifications.
False. Around 10-15% of Wilms tumors contain calcifications.

True or false? Neuroblastomas are usually unilateral.
False. Most neuroblastomas are bilateral. On the other hand, as previously specified, most Wilms tumors are unilateral.

True or false? Neuroblastomas typically encase but do not invade vascular structures.
True. Neuroblastomas encase rather than invade vascular structures. A classic imaging appearance would be a mass lifting the aorta off the vertebral column. Note, however, that more aggressive neuroblastomas can invade structures like the adjacent kidney and muscle.

What age group is most common for neuroblastomas?
Think young kids such as those 2 years and under.

What tumor is more common to extend into the chest: Wilms or neuroblastoma?
Neuroblastomas more commonly has extension into the chest.

True or false? A neuroblastoma is the most common childhood malignancy.
False. Leukemia is the most common childhood malignancy and brain tumors are the most common solid tumor in kids. However, neuroblastomas are the most common extracranial solid tumor of childhood.

What is so called Pepper syndrome of neuroblastoma?
Pepper syndrome is an adrenal neuroblastoma with hepatomegaly due to extensive hepatic metastases.

What is the stage of neuroblastoma for a pediatric patient with Pepper syndrome?
Stage 4S.

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What are the features of stage 4S neuroblastoma?
The "S" in 4S stands for "special". This is the "special" type of neuroblastoma. As such, expect questions on this on a board exam. Stage 4S neuroblastoma occurs in pediatric patients under 1 year of age with localized primary tumors and distant metastases only involving liver, skin and/or bone marrow.

Stage 4S tumors have an up to 90% 3-year survival, which is approximately the same as stage 1 or 2 disease, compared to a worse 3-year survival for stage 4 disease. Note with the non-special stage 4 that those over 1 year of age have dismal survival at 3-years at only 15%. Whereas patients under 1 year of age with stage 4S neuroblastoma have a 3-year survival up to 75%.

True or false? Most neuroblastoma patients present with stage 1 or 2 disease.
False. Up to 75% of neuroblastomas present with stage 3 or stage 4 tumors.

What is so called blueberry muffin syndrome?
Blueberry muffin syndrome is broader than neuroblastoma but includes neuroblastoma. Blueberry muffin syndrome is a classic skin appearance in pediatric patients with multiple blue to purple cutaneous lesions associated with neuroblastomas, leukemias, and hematological disorders as well as viral infections like cytomegalovirus and congenital rubella. This is a form of extramedullary hematopoiesis of the skin.

What is so called Hutchinson syndrome?
Hutchinson syndrome is neuroblastoma with bone metastases causing clinical symptoms of bone pain and limping, as well as proptosis and periorbital disease involvement which can include so-called raccoon eyes. If you do not know what raccoon eyes look like with neuroblastoma, please look this up.

What is the term for the jerking eye movements, cerebellar ataxia, and myoclonic jerking that in a pediatric patient is strongly associated with neuroblastoma?
Opsomyoclonus. This is essentially an autoimmune-like paraneoplastic phenomenon that can occur in pediatric patients with neuroblastoma.

What are key features that can help differentiate a rhabdoid tumor of the kidney from a Wilms tumor?
Like neuroblastomas, rhabdoid tumors also are more common in children less than 2 years of age whereas Wilms tumors most commonly present in slightly older children about 3-5 years of age. About 2/3 of rhabdoid tumors have calcification compared to around 10-15% of Wilms tumors. Rhabdoid tumors tend to be very aggressive so one may expect to see vascular invasion, central necrosis, hemorrhage and subcapsular fluid collections.