



Abdominal Masses of Childhood

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History

- **A child is seen by the PCP.**
- **The mother has noticed the child's abdomen was different upon bathing**



History Discussion

- **What other points of the history do you want to know?**
 - **Age:** a crucial factor that may adjust the differential diagnosis
 - **Mass:** duration, associated pain, changes in eating and elimination patterns, history of trauma
 - **Birth hx:** prematurity, difficult birth, prenatal care
 - **Medical hx:** associated medical illnesses. Previous concerns for hematuria or hypertension?
 - **Family hx:** syndromes (Beckwith-Wiedemann, WAGR, Gardner)
 - **ROS:** night sweats, malaise, bleeding or bruising, skin changes



BWS

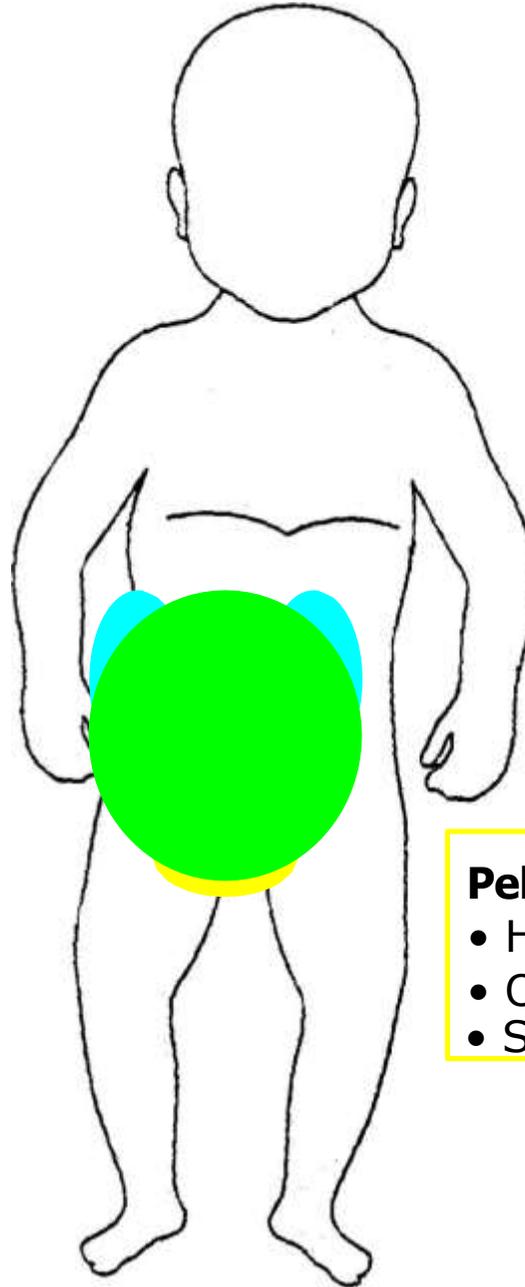


Macroglossia, hemihyperplasia, omphalocele, neonatal hypoglycemia, macrosomia, embryonal tumors (e.g., Wilms tumor, hepatoblastoma, neuroblastoma, and rhabdomyosarcoma), visceromegaly

INFANTS

Flank - 65%

- **Renal - 55%**
- Hydronephrosis
- Polycystic kidney
- Mesoblastic nephroma
- Renal ectopic
- Renal vein thrombosis
- Nephroblastomatosis
- Wilms tumor
- **Nonrenal - 10%**
- Adrenal hemorrhage (Waterhouse Friderichsen syndrome)
- Neuroblastoma



Intraabdominal - 20%

- **GI Masses - 15%**
- Duplication
- Meconium ileus
- Mesenteric-omental cyst
- **Hepatobiliary - 5%**
- Hemangioendotheloma
- Hepatoblastoma
- Hepatic cyst
- Choledochal cyst
- Hydrops of gallbladder

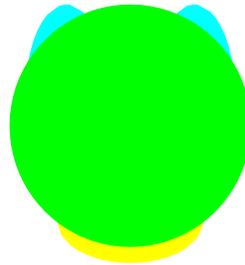
Pelvic - 15%

- Hydrometrocolpos
- Ovarian cyst
- Sacrococcygeal teratoma

CHILDREN AND ADOLESCENTS

Flank - 78%

- **Renal – 55%**
- Wilms tumor
- Hydronephrosis
- Cystic disease
- **Nonrenal – 23%**
- Neuroblastoma
- Teratoma
- Other neoplasms



Intraperitoneal – 18%

- **GI Masses - 12%**
- Appendiceal abscess
- Other neoplasms
- **Hepatobiliary – 6%**
- Hepatoblastoma
- Hepatocellular ca
- Choledochal cyst

Pelvic – 4%

- Ovarian cyst
- Hydrometrocolpos

Physical Exam

- **What specifically would you look for?**
 - Vital Signs: some tumors can cause elevated HR, BP; some masses may push up on diaphragm and limit breathing
 - NB: This is a neoplasm of neural crest origin, arising in the adrenal medulla and along the sympathetic ganglion chain from the neck to the pelvis
 - Appearance: look for overgrowth
 - H/N: aniridia, raccoon eyes, proptosis, Homer's syndrome
 - Chest: Rapid and shallow breathing



Physical Exam

- **What specifically would you look for?**
 - Cardiac: congestive heart failure (SCT=Vascular Steal Syndrome)
 - Lymphadenopathy
 - Abdomen:
 - Omphalecele, hepatosplenomegaly
 - Mass – location, configuration, size, consistency, mobility, tenderness
 - GU: ambiguous genitalia, hypospadias, cryptorchidism



Studies (Labs)

- **What labs are needed?**
 - CBC and differential
 - Lytes, BUN, Cr
 - Liver function tests
 - Amylase, lipase
 - Urine: U/A, Vanillylmandelic acid (VMA), Homovanillic acid (HVA)
 - Markers: alpha-fetoprotein (AFP), β -HCG



Studies (Imaging)

- **Investigations:**
 - X-rays: not usually helpful
 - US: good first test
 - CT: good to help plan surgery and for staging
 - MRI: limited application
 - Nuclear scans: selective use based on diagnosis



CT Scans

Wilms Tumor



- “Claw sign: Sharp angles on either sides of the mass”: Wilms mass arising from kidney

Neuroblastoma



- Calcifications more likely seen in neuroblastoma



Case Discussion

- **Diagnosis**
 - See flowchart
- **Plans**
 - See flowchart



Approach to the Abdominal Mass

History, Physical, Lab Tests

ABDOMINAL MASS

Abdominal Radiograph

REFER { Bowel Obstruction
Bowel gas displacement
Consider: BE or UGI
Calcifications

Abdominal Ultrasound

Confirms

FLANK

PELVIC

INTRAPERITONEAL

Renal

Adrenal

Other

Cystic

Solid

Complex

Solid

Cystic

Cystic
normal ureter
dilated ureter

Solid Cystic Solid Cystic Solid
Mesenteric Omental
Choledochal
Intussusception

CT SCAN OF ABD.
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Renal Scan
MCKD
UPJ
VCUG
VUJ
VUR
PUV

WT
RCC
Adrenal Hemorrhage
NB
Cystic Hygroma
SCT

HIDA

SCT
NB
RMS
Duplication
Mesenteric
Omental
Lymphoma

PEDIATRIC GENERAL SURGERY REFERRAL

Operation

- Goals of surgery is:
 - Staging (Invasion and LNs)
 - Obtain tissue for diagnosis only, if not resectable
 - Complete resection (avoid disruption of the margins to avoid tumor rupture)
 - Assistance with radiotherapy, or assistance with chemotherapy (Debulking)



Complications

- **Peri-operative**

- Ileus is common after any abdominal surgery.
- Post-op intussusception is well reported, particularly after retroperitoneal dissection.

- **Long Term**

- Is dependent on the tumor type and whether rupture has occurred.
- Potential for adhesive bowel obstruction.



Questions

1. Where do most abdominal masses arise?
 - A. Flank
 - B. Intraperitoneal
 - C. Pelvic
 - D. None of the above



Questions

1. Where do most abdominal masses arise?

A. **Flank**

B. Intraperitoneal

C. Pelvic

D. None of the above



Questions

2. Which is the most useful first test to order to help determine the type of abdominal mass?
- A. X-ray
 - B. Ultrasound
 - C. CT scan
 - D. MRI



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Questions

3. With regards to abdominal masses, the goal of surgery may include?
- A. Staging
 - B. Obtain tissue for diagnosis
 - C. Resection of mass
 - D. Help adjuvant therapy
 - E. All the above



Questions

3. With regards to abdominal masses, the goal of surgery may include?
- A. Staging
 - B. Obtain tissue for diagnosis
 - C. Resection of mass
 - D. Help adjuvant therapy
 - E. **All the above**



Questions

4. With regards to abdominal masses, avoiding tumor rupture is critical to?
- A. Avoiding pathologic misinterpretation
 - B. Intraoperative blood loss
 - C. Not upstaging the patient
 - D. Decreasing likelihood of postoperative intussusception
 - E. Spuriously increasing tumor markers after surgery



Questions

4. With regards to abdominal masses, avoiding tumor rupture may be critical to?
- A. Avoiding pathologic misinterpretation
 - B. Intraoperative blood loss
 - C. Not upstaging the patient**
 - D. Decreasing likelihood of postoperative intussusception
 - E. Spuriously increasing tumor markers after surgery



Review

- History and physical are key to help determine the type of abdominal mass.
- Most masses arise from the flank.
- US is the first test to do to determine the source.
- CT is the next test to help surgical planning.



Wilms Tumor

Discussion of History Elements

- Congenital abnormalities?
 - Associated with WAGR syndrome, Beckwith-Wiedemann syndrome (BWS), Denys-Drash syndrome (DDS=WT+DSD), hemihypertrophy and Perlman syndrome (low muscle tone+DD)
- Abdominal pain?
 - Most commonly asymptomatic or no abdominal pain
- Hematuria?
 - Gross hematuria (18%)
 - Microscopic hematuria (24%)
- Weight loss/Appear ill?
 - Patients with Wilms tumors typically healthy appearing compared to Neuroblastoma – ill appearing due to typical metastatic disease
- Hypertension?
 - 20-25% of patients

Physical Exam

- Healthy appearing toddler with palpable mass in abdomen/flank, non tender.

Physical Exam

- Left sided varicocele – tumor has extended into left renal vein obstructing left testicular vein

Studies (Labs, Imaging)

- What labs are needed?
 - CBC, BMP, PT/INR – rule out anemia, coagulopathy
 - Urinalysis – evaluate for hematuria
- What imaging is needed?
 - Abdominal ultrasound
 - Including doppler to evaluate mass extension into renal vein/IVC
 - CT scan abdomen/pelvis
 - Evaluate for location of disease, whether the masses are bilateral, metastatic disease.

CT scan abdomen/pelvis



-
- Diagnosis
 - Left sided Wilms' tumor
 - Management
 - Staging – two systems:
 - Children's Oncology Group (COG)
 - Focuses on surgery as primary therapy, followed by chemotherapy
 - Société Internationale d'Oncologie Pédiatrique (SIOP)
 - Focuses on neoadjuvant chemotherapy, followed by surgery

Staging

COG Wilms' Tumor Staging

<i>Stage</i>	<i>Criteria</i>
I	<p>The tumor is limited to the kidney and has been completely resected.</p> <p>The tumor was not ruptured or biopsied prior to removal.</p> <p>There is no penetration of the renal capsule or involvement of renal sinus vessels.</p>
II	<p>The tumor extends beyond the capsule of the kidney but was completely resected with no evidence of tumor at or beyond the margins of resection.</p> <p>There is penetration of the renal capsule or invasion of the renal sinus vessels.</p>
III	<p>Gross or microscopic residual tumor remains postoperatively, including inoperable tumor, positive surgical margins, tumor spillage surfaces, regional lymph node metastases, positive peritoneal cytology, or transected tumor thrombus.</p> <p>The tumor was ruptured or biopsied prior to removal.</p>
IV	<p>Hematogenous metastases or lymph node metastases outside the abdomen (e.g., lung, liver, bone, brain).</p>
V	<p>Bilateral renal involvement is present at diagnosis, and each side may be considered to have a stage.</p>

Case Discussion

- Management
 - Preop
 - CT scan of chest → rule out lung metastasis
 - Most common metastatic sites are lung and liver
 - Laboratory studies
 - CBC, PT/INR, PTT, Type and Screen
 - Operative
 - Radical vs partial nephrectomy

Operation – Goals – Radical Nephrectomy

- Safely resect entire tumor
- Avoid upstaging tumor by complications
 - Capsular tears, gross tumor spillage, biopsy of tumor
- Adequately stage tumor
 - Evaluate for metastasis, removal/biopsy of appropriate lymph nodes (most common operative error), evaluating vascular invasion
- Adequately documenting pre-operative vs intraoperative tumor rupture
 - Changes post operative management with radiation

Operation - Contraindications

- Extension of tumor thrombus above the level of the hepatic veins
- Tumor involves surrounding structures, requiring removal of those structures to remove complete tumor
- Tumor involves bilateral kidneys
- Tumor involves a solitary kidney
- Pulmonary metastasis leading to respiratory compromise

- ****All indications for neoadjuvant chemotherapy****

Operation – Complications

- Tumor Spill (9.7%)
 - Break in tumor capsule
 - Includes pre-operative or intraoperative needle or core biopsy (COG protocol)
 - Transection of ureter or renal vein where tumor exists
- Bleeding (2%)
- Vascular or bowel injuries

Post-Op Management

- Peri-operative management
 - Monitor for ileus – particularly in patients with extensive retroperitoneal dissection
 - Monitor for complications:
 - Wound infection
 - Bowel obstruction
 - Intussusception
 - Prepare for radiation and chemotherapy

Questions

The most common presenting abnormality in patient's with a Wilms' tumor is:

- a. Hypertension
- b. Hematuria
- c. Asymptomatic abdominal mass
- d. Abdominal pain

Questions

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- a. Hypertension
- b. Hematuria
- c. **Asymptomatic abdominal mass**
- d. Abdominal pain

Questions

Which of the following does NOT increase recurrence of tumor?

- a. Pre-operative core needle biopsy
- b. Resection lymph nodes at renal hilum, along iliac vessel and para-aortic regions
- c. Transecting ureter containing tumor
- d. Sustaining minor renal capsular tear during dissection from surrounding structures

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Final Discussion/Review

- Wilms' tumor is the most common pediatric renal mass and 2nd most common pediatric abdominal mass
- Most common presentation: asymptomatic abdominal mass
- Treatment for unilateral tumors: Radical nephrectomy followed by chemotherapy +/- radiation
- Histology and stage of tumor main prognostic indicators for Wilms tumors