RADY 401 Case Presentation: Extrarenal Wilms Tumor / Nephroblastoma

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Focused Patient History and Workup

- 10 y.o. male with a h/o asthma presents with
  - acute onset RLQ abdominal pain x1 day
  - 1x non-bloody, nonbilious emesis.
  - No fevers, constipation, diarrhea, urinary complaints, or testicular pain.

- Pertinent Physical Exam findings:
  - Abdominal exam shows normal bowel sounds, mildly distended but soft belly, and tenderness to palpation of RLQ w/o rebound or guarding. No hernias or masses palpated.
  - Both testicles descended appropriately.

- Labs:
  - Leukocytosis of 20.3 WBC with neutrophil predominance
  - Mild microcytic anemia: Hgb 11.8 (LLN), Hct 35 (LLN), MCV 70 (L), RDW 15.8 (H)
  - Unremarkable metabolic panel
  - Unremarkable urinalysis
  - C/F Appendicitis >>> MRI pelvis w/ and w/o IV contrast
Differential Diagnosis for School-Aged Pediatric Vomiting/Abdominal Pain

- Gastroenteritis
- Appendicitis
- Diabetic Ketoacidosis
- Ovarian/Testicular Torsion
- Foreign Body Ingestion
- Volvulus
- Trauma
- Renal Dx/Stone/Pyelonephritis
- Biliary Dx
List of Imaging Studies

• ED Outside Hospital:
  • X-Ray Abdomen KUB
  • MRI Pelvis with and without IV contrast
Findings: *Large stool is demonstrated, throughout the colon and rectum. No dilated gas filled loops of bowel are demonstrated. No pneumatosis. No suspicious calcifications. Bones are unremarkable.*

Impression: *Nonobstructive bowel gas pattern with a large colonic and rectal stool burden.*

Stool burden is recognizable on radiography as visible gas bubbles within semisolid/soft-tissue density. [1]
MRI Pelvis with and without IV Contrast

Aortic bifurcation

S1

Bladder

Restricted diffusion.
Large, heterogeneous pelvic mass with multiple internal septations with hyperintense cystic components and isointense solid components.
Hyperintense fat on T2.

Hypointense, suppressed fat on T2.

Mass components unchanged.
Large pelvic mass with peripheral enhancement and internal septation enhancement.
C/F Appendicitis?

MRI T2 Sagittal View, right/middle abdomen

Appendix Findings: Retrocecal appendix is visualized and is a normal caliber. There is small amount of fluid surrounding the appendix.
Final Impression, MRI Pelvis

1. Large solid cystic pelvic mass measuring up to 9.7 cm in the central pelvis abutting the sacrum at S1. Differential considerations would include sacrococcygeal teratoma although there are no definitive calcifications, rhabdomyosarcoma, or extra-adrenal pheochromocytoma.

2. Mild RIGHT hydronephrosis, likely external compression on the RIGHT ureter from the pelvic mass.

Patient treatment or outcome

• Emergent Exploratory Laparotomy and Tumor Resection
  • Large tumor originating from retroperitoneum and sacrum resected.
  • Tumor biopsied and sent to pathology for pediatric tumor work-up.
  • Evidence of pre-operative rupture with blood in abdomen and pelvis.

• Pathology:
  • *Triphasic Wilms tumor with additional heterologous elements (skeletal muscle and smooth muscle.) The amount of heterologous elements is not sufficient for use of the descriptive term "teratoid Wilms."

• Referred to UNC Heme/Onc and tumor board review
  • CT Chest is recommended to evaluate for metastases.
Pediatric EM Physician concerned for appendicitis opted for MRI over ultrasound due to patient’s body habitus. BMI = 97th percentile for category.
ACR Appropriateness Criteria – MRI Abdominal and Pelvis with and without IV Contrast

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*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies”.

Radiation Dose = 0 mSv

Estimated Costs = $106.93-369.59

- w/ IV Contrast: 94% sensitivity & 94% specificity
- w/o IV Contrast: 86% sensitivity & 94% specificity
**Classic Image Findings: Acute Appendicitis**

- **RUQ U/S:**
  - Findings
    - Dilated appendix w/ diameter >6mm, noncompressible
    - Prominent appendiceal wall layers/wall thickening
    - Possible appendicolith with posterior acoustic shadowing
    - Periappendiceal fluid collection/fat
  - Cons
    - Operator dependent
    - Can be limited by obese body habitus, which is why managing physician ordered MRI for better visualization
    - Difficult to visualize through air or bowel

- **MRI Pelvis & Abdomen w/ & w/o IV Contrast**
  - Findings
    - Appendix lumen distension
    - Wall thickening
    - Periappendiceal free fluid
  - Cons
    - Time
    - Cost
Actual Dx: Extrarenal Wilms Tumor / Nephroblastoma

• Extrarenal manifestation is extremely rare and location can delay diagnosis due to lack of traditional presentation of palpable abdominal mass seen in toddler-aged children. (6)

• Renal Wilms Tumor Imaging:
  • 1ST Line: U/S = vascular tumor with hypoechoic areas of necrosis
  • Abdominal CT/MR = surgical excision mapping
  • CT Chest/Thorax = staging and metastases (7)
    • This is the next step in this patient’s treatment plan to assess for chest/lung metastases and determine next therapeutic steps.
Wrap Up

1. Imaging indications for appendicitis in pediatric patients aim to avoid excessive radiation exposure associated with CT.
   a) Abdominal ultrasound
   b) MRI abdomen and pelvis

2. Respect the limitations of ultrasound before ordering. Additionally, an equivocal or negative ultrasound study does not rule out clinically suspected appendicitis: further imaging is indicated.

3. It’s not always appendicitis, but common things are common. Utilize ACR appropriateness criteria for clinical decision-making.
References


