AMSER Case of the Month September 2019

14 year-old male with inability to void, difficulty passing stool, and bilateral abdominal pain

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Patient Presentation

- **HPI**: 14 y.o. male who presented to OSH with bilateral RLQ/ LLQ abdominal pain, vomiting, inability to urinate for 2-3 days, and inability to have a normal bowel movement for the last 1-2 weeks.
  PMH: None
- **Meds**: None
- **Allergies**: NKDA
- **Surg Hx:** None
- **Fam Hx**: Mother passed away from unspecified cancer; father also passed away from a head and neck cancer
- **Social Hx**: Lives with aunt and uncle/ attends public school

Patient Presentation

- **ROS**:
  - +: Night sweats, chills, loss of appetite, low back pain
  - -: Weight loss, headache, chest pain
- **Vitals**:
  - Temp 99.0 F
  - HR 105 bpm
  - BP 146/80 mm Hg
  - RR 22/min
  - SpO2 100%

Hypertension

- **Physical Exam**: Bowel sounds normoactive, soft and minimally distended abdomen, moderate tenderness of RLQ and LLQ, no rebound or guarding, no hepatosplenomegaly

Pertinent Labs

- **Labs**:
  - **Electrolytes normal**
  - **Cr:** 4.34
  - **CBC**:
    - WBC 10.05
    - HgB 10.9
    - Hct 31.5

Acute kidney injury

Anemia

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

American College of Radiology
ACR Appropriateness Criteria®

Clinical Condition: Renal Failure

Variant 1: Acute kidney injury (AKI), unspecified.

Radiologic Procedure	Rating	Comments	RRL*
US kidneys and bladder	9	Assess renal size and echogenicity. Exclude bilateral obstruction in high-risk groups. Doppler may be used to assess renal perfusion.	O
Percutaneous US-guided renal biopsy	6	Especially useful in acute inflammatory conditions such as nephritis. Perform a follow-up after US examination, if needed.	O
Tc-99m MAG3 scan kidney	4	This procedure may be useful if the creatinine level is high. Perform a follow-up after US examination, if needed.	☢☢☢
MRI abdomen without IV contrast	3	This procedure has a potential role in searching for sonographically unclear causes of ureteral obstruction. A nonenhanced MRI may be helpful in selected cases.	O
MRA abdomen without and with IV contrast	3	Because the eGFR and creatinine values are unreliable in the setting of AKI, caution should be used when administering intravenous gadolinium Gadolinium-enhanced studies are very effective for renal artery evaluation.	O
MRA abdomen without IV contrast	3	This procedure can access renal arterial or venous patency in rare instances when vascular stenosis or thrombosis may account for AKI.	O
Arteriography kidney	3	Potentially helpful in trauma evaluation for renal artery occlusion. Consider using aortography with CO^2 to avoid nephrotoxicity of the iodinated contrast.	☢☢☢
CT abdomen without IV contrast	3	Potentially helpful in trauma evaluation. Noncontrast helical CT is more sensitive than KUB for calculi. Evaluation of ureteral obstruction due to retroperitoneal diseases, masses, and tumors (hydronephrosis on US but an undetectable cause).	☢☢☢
CTA abdomen with IV contrast	3		☢☢☢
CT abdomen with IV contrast	2		☢☢☢
CT abdomen without and with IV contrast	2		☢☢☢☢
X-ray abdomen and pelvis (KUB)	2	To assess for calculi; however, it is insensitive for 30% of calculi.	☢☢
X-ray voiding cystourethrography	2	A VCUG may be indicated if a vesicoureteral reflux is suspected as a contributing factor in AKI.	☢☢
MRI abdomen without and with IV contrast	2	Potential role in search of sonographically unclear causes of ureteral obstruction.	O

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level

This imaging modality was ordered by the pediatric attending based on the patient presentation and laboratory results.

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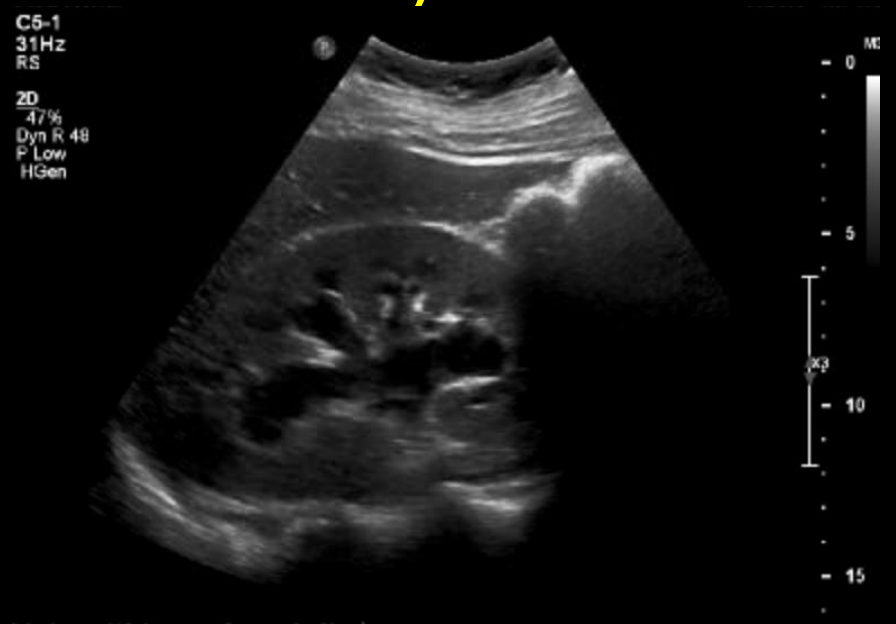
Findings (unlabeled)



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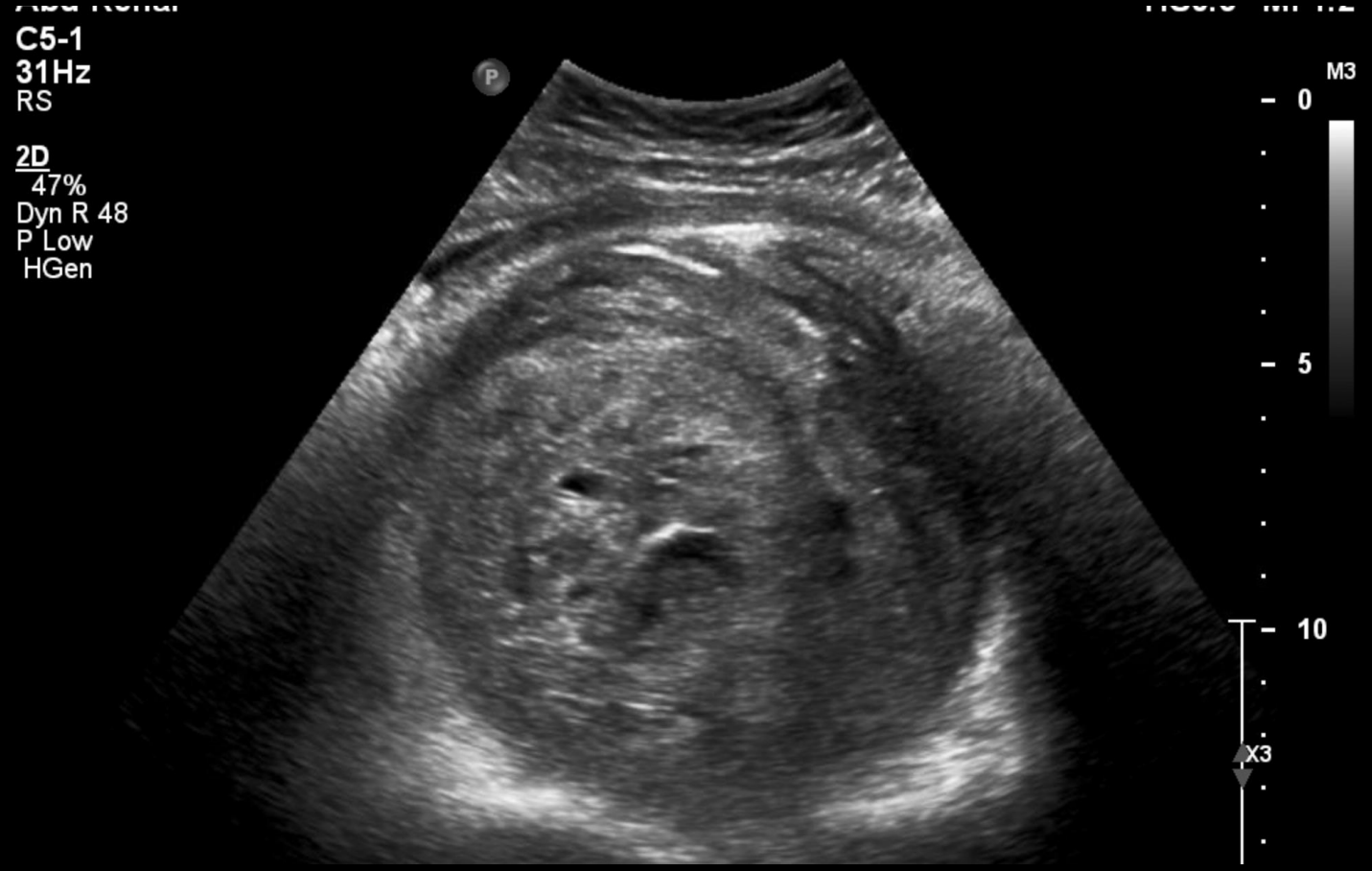

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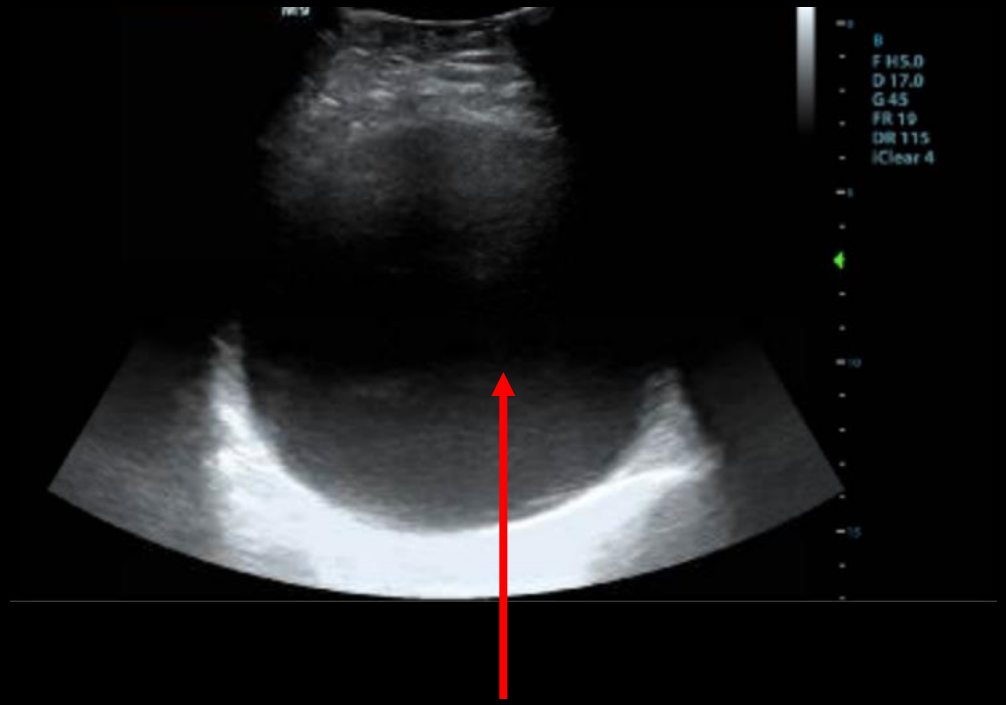
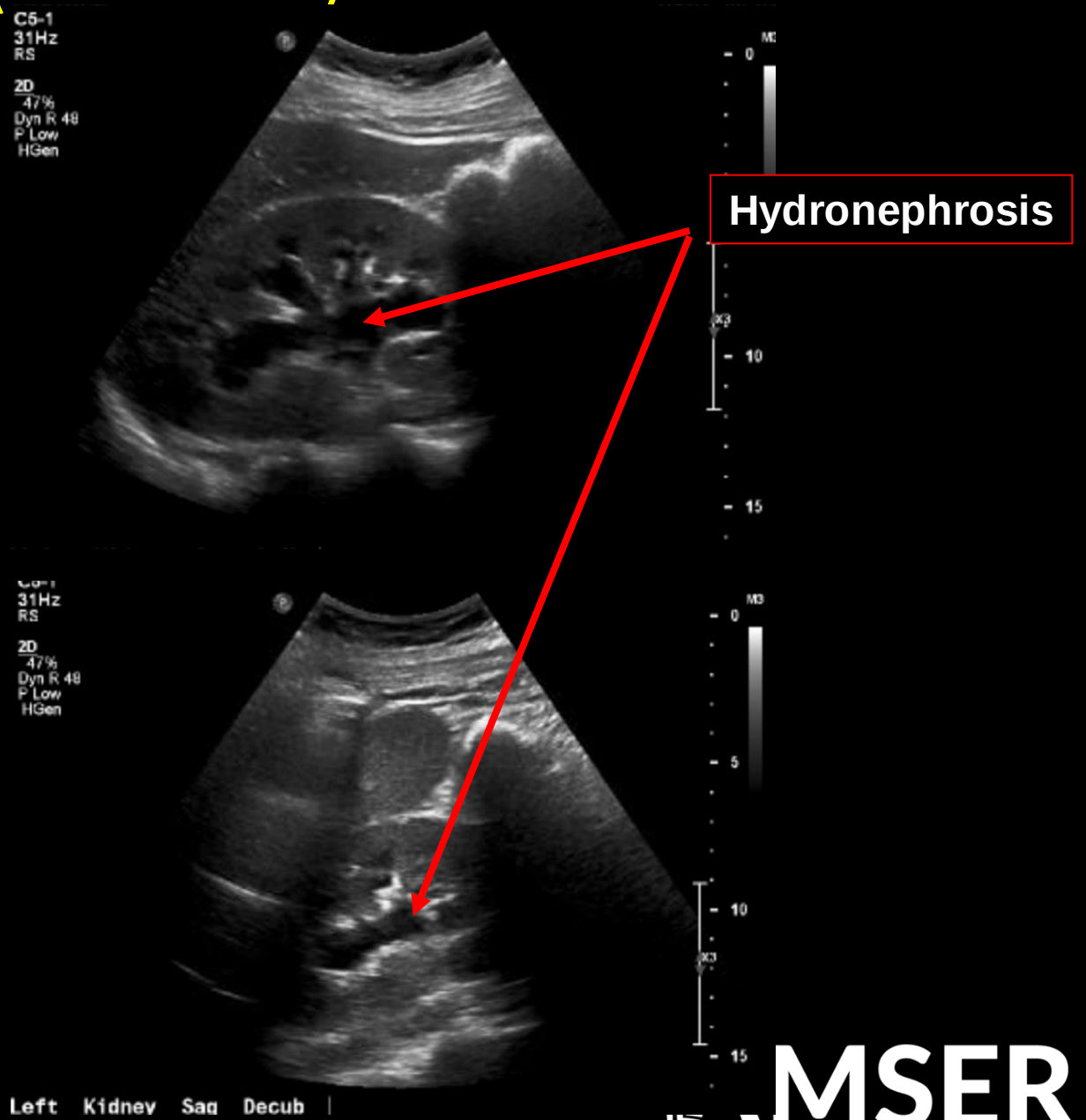


Distended bladder



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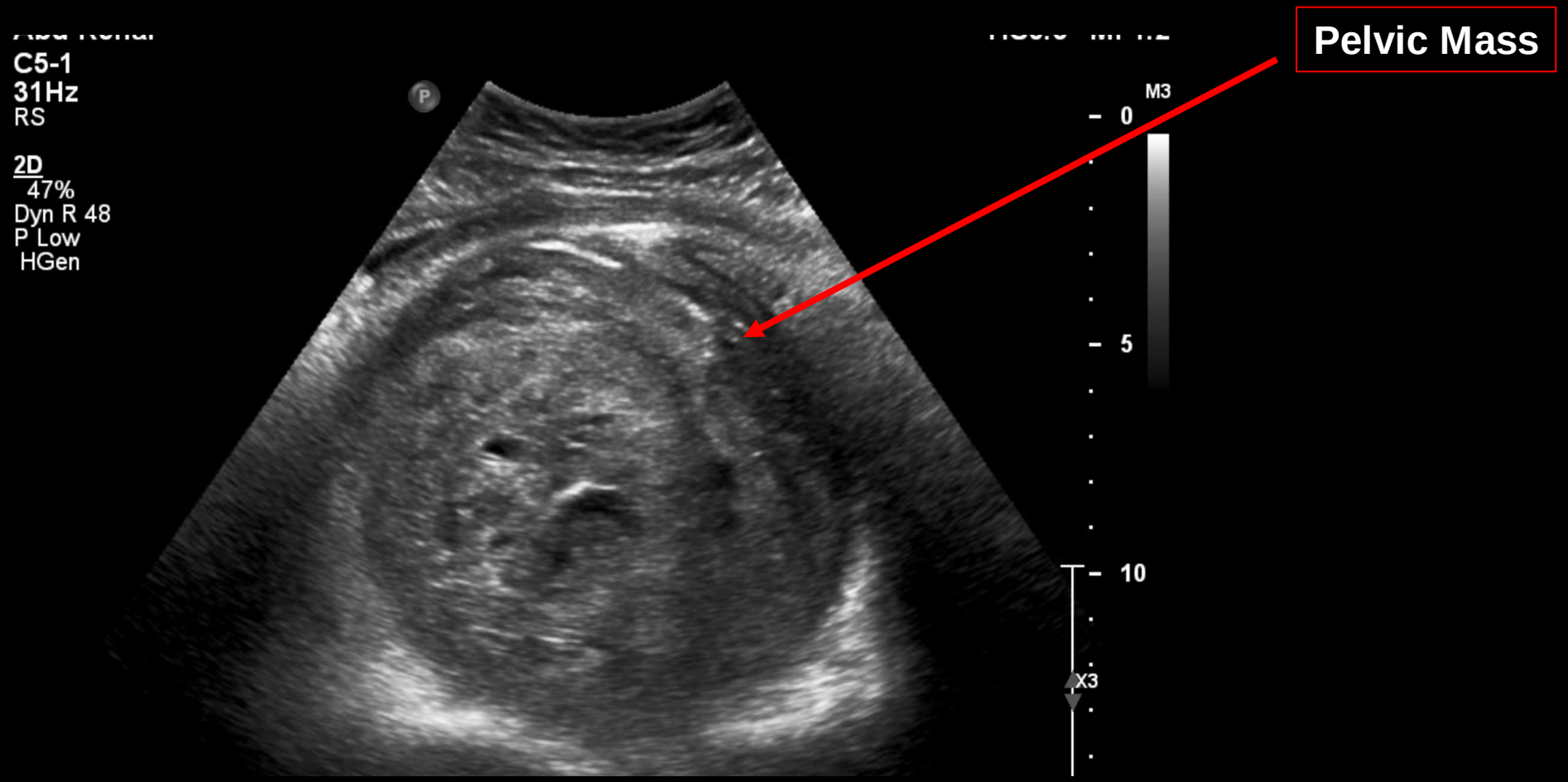

Select the applicable ACR Appropriateness Criteria

Variant 1: **Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen with IV contrast	Usually Appropriate	☢☢☢
US abdomen	Usually Appropriate	O
MRI abdomen without and with IV contrast	May Be Appropriate	O
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
MRI abdomen without IV contrast	May Be Appropriate	O
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
Radiography abdomen	Usually Not Appropriate	☢☢
Fluoroscopy contrast enema	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	☢☢☢

This imaging modality was ordered next by the pediatric attending.

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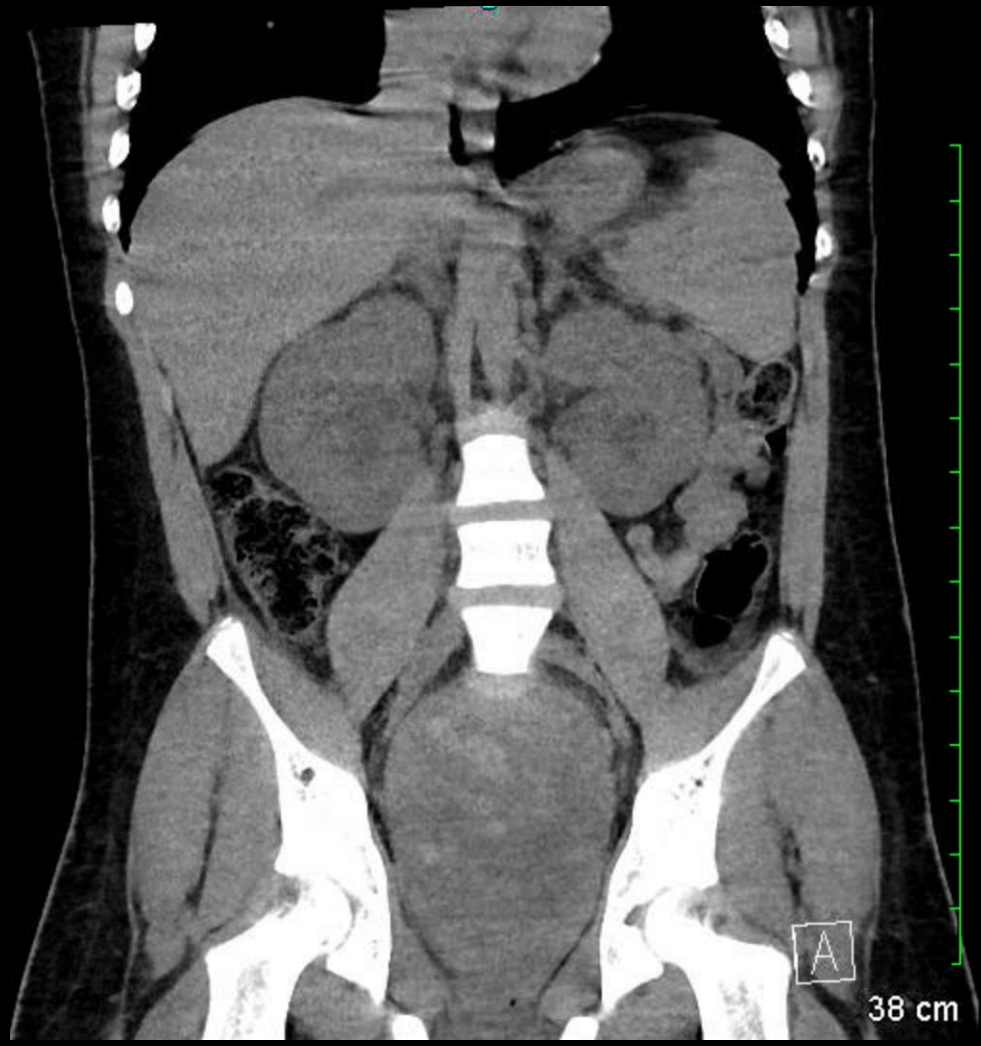

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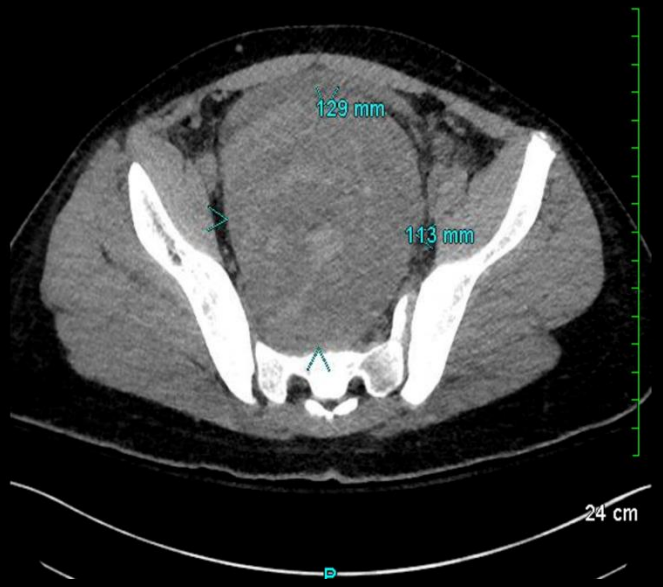

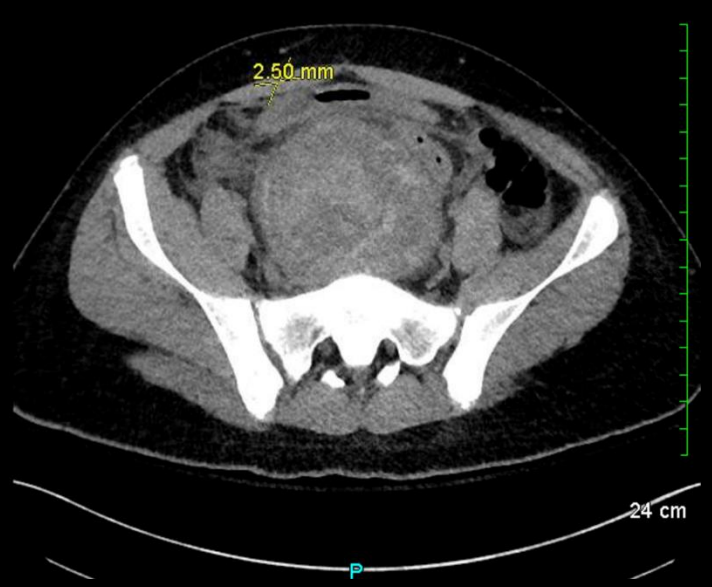

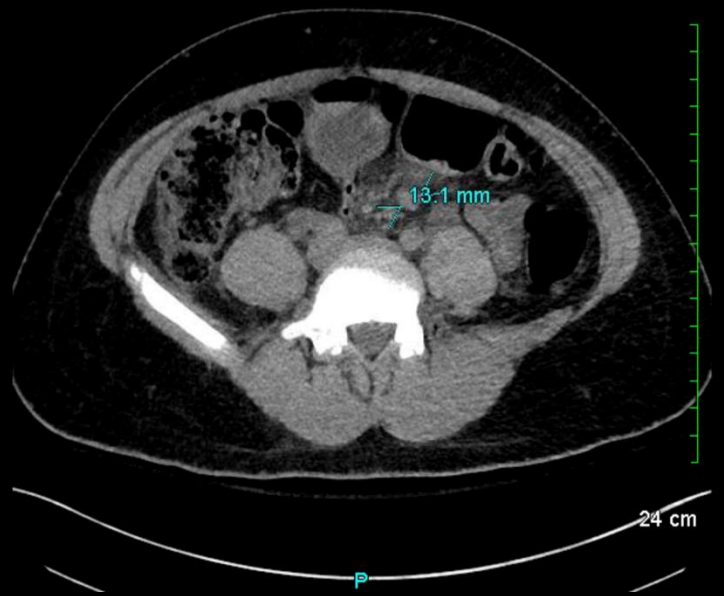

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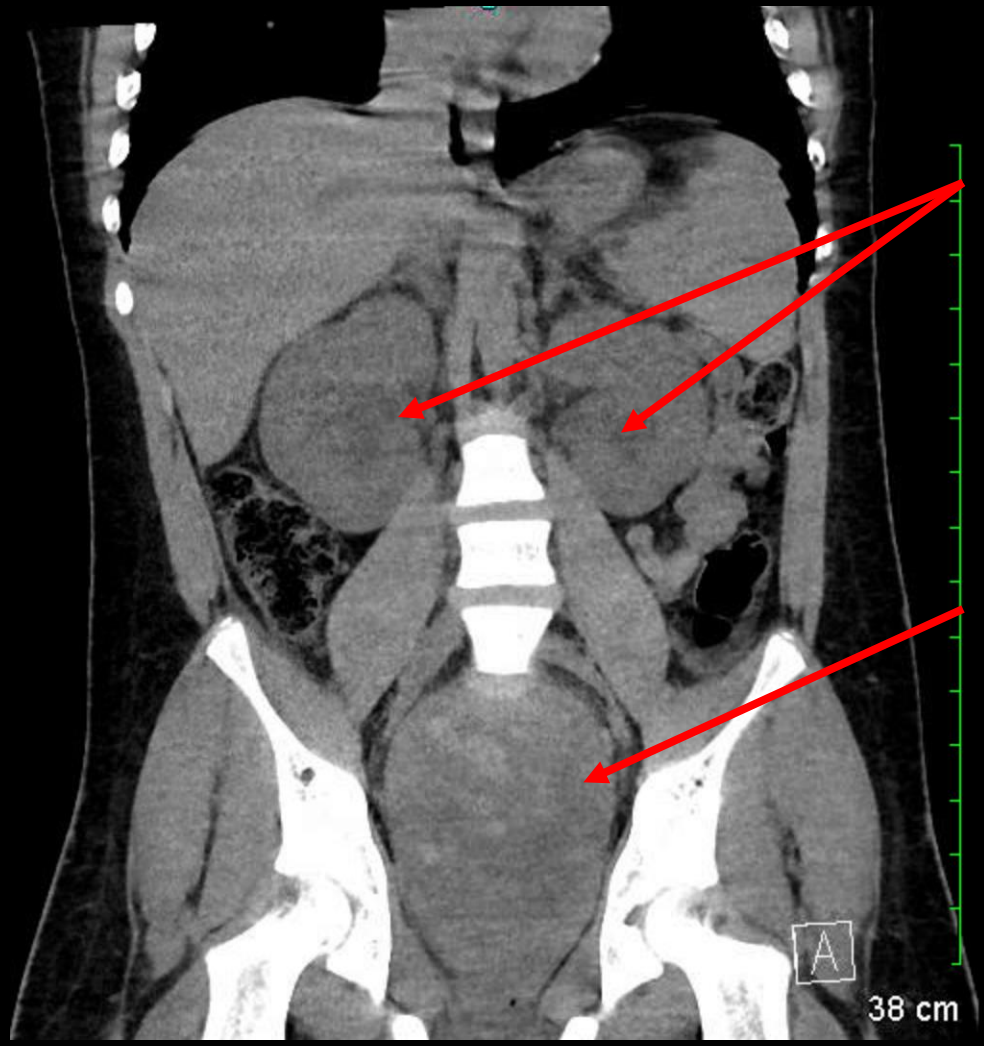

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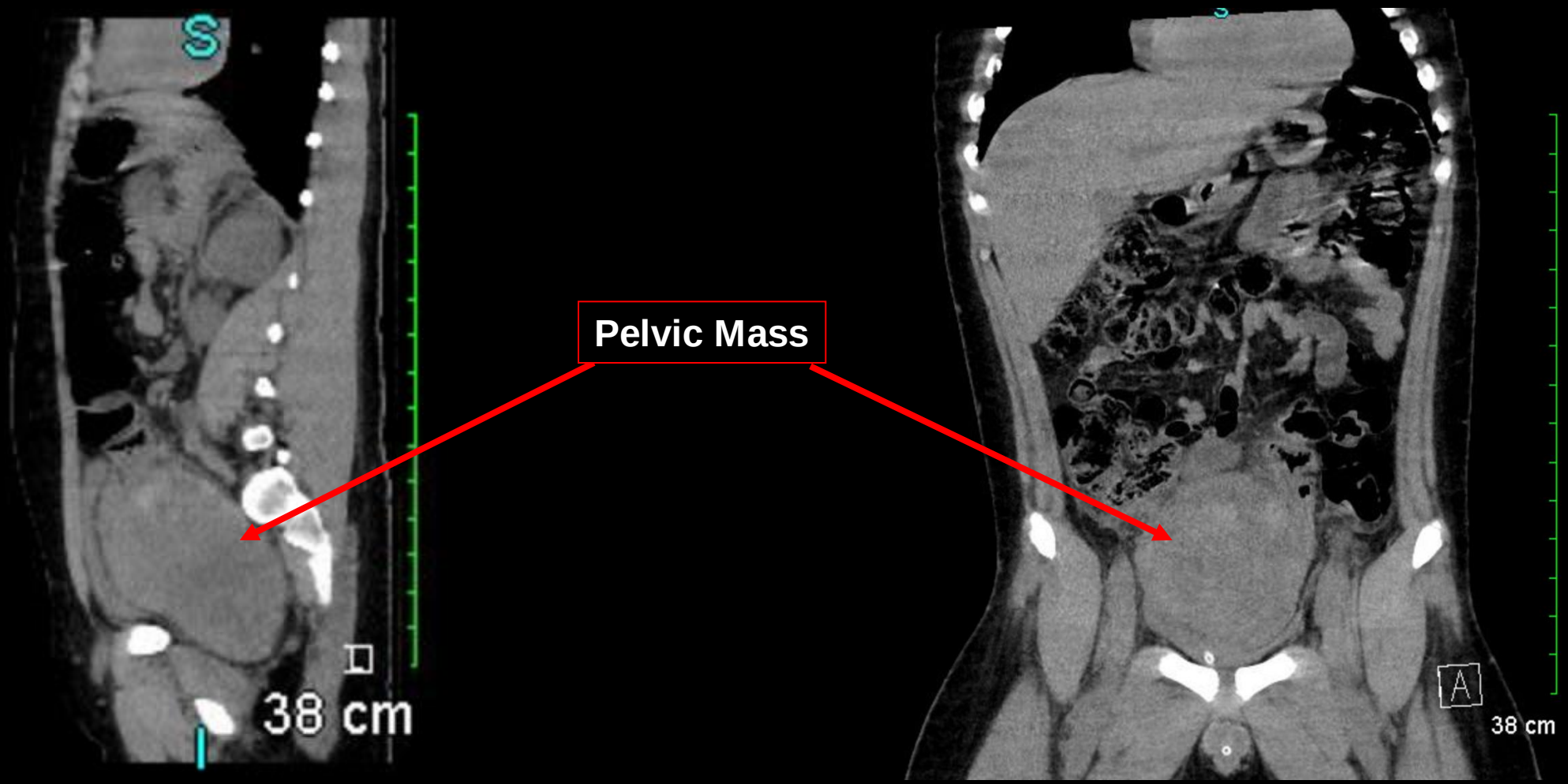

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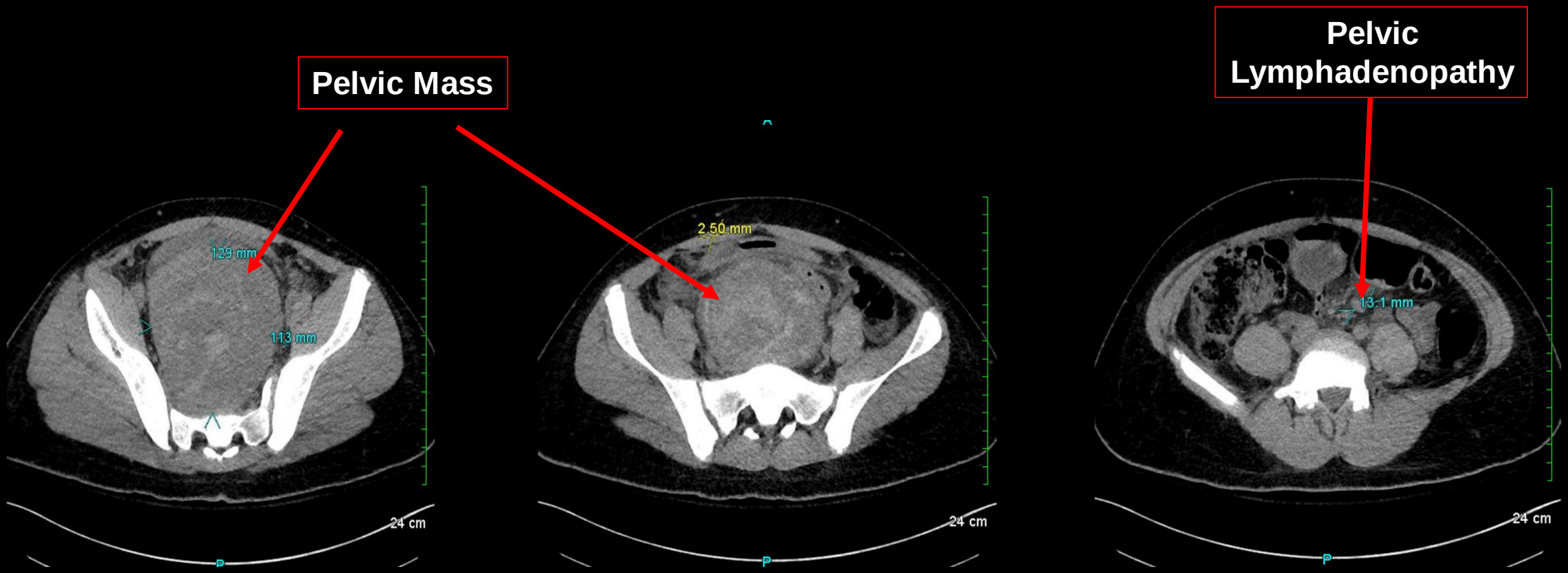


Differential Diagnosis

- Bladder neoplasm
- Bladder clot
- Teratoma
- Desmoplastic Small Cell Round Tumor
- Leiomyoma
- Lymphoma
- Rhabdomyosarcoma
- If female, would consider ovarian germ cell neoplasms

Final Dx:

Undifferentiated Round Cell Carcinoma

Round Cell Tumors

- Round cells with increased nuclear-cytoplasmic ratio.
- This group of tumor includes entities such as peripheral neuroectodermal tumor, rhabdomyosarcoma, synovial sarcoma, non-Hodgkin's lymphoma, neuroblastoma, hepatoblastoma, Wilms' tumor, and desmoplastic small round cell tumor.
- They are **highly aggressive malignant tumors.**
- Often difficult to diagnose when the tumor is poorly differentiated.

The following slides will discuss rhabdomyosarcoma, a more common pediatric pelvic tumor.

Rhabdomyosarcomas

- Rhabdomyosarcomas are thought to originate from immature cells that are destined to form striated muscles.
- Common tumors that can occur in multiple locations including the head and neck, genitourinary system (prostate, bladder, vagina), and extremities.
- Average age of diagnosis is 5 years old.
- Clinical presentation depends on location, but pelvic rhabdomyosarcomas are accompanied by **urinary obstruction, constipation**, and hematuria. **Abdominal masses may cause hypertension.**
- Depending on the subtype, may be aggressive.
- The metastatic workup should be tailored to the clinical and histological characteristics.

Rhabdomyosarcoma - Workup

- **Diagnostic Workup:**
  - Depends on location of the neoplasm. For example, MRI of head and neck or extremities.
  - CT and MRI of abdomen and pelvis to evaluate surrounding structures and evaluate involvement of retroperitoneal nodes.
  - Chest CT with or without contrast
  - Bilateral bone marrow aspiration and biopsy
  - Radionuclide bone scan
  - PET CT

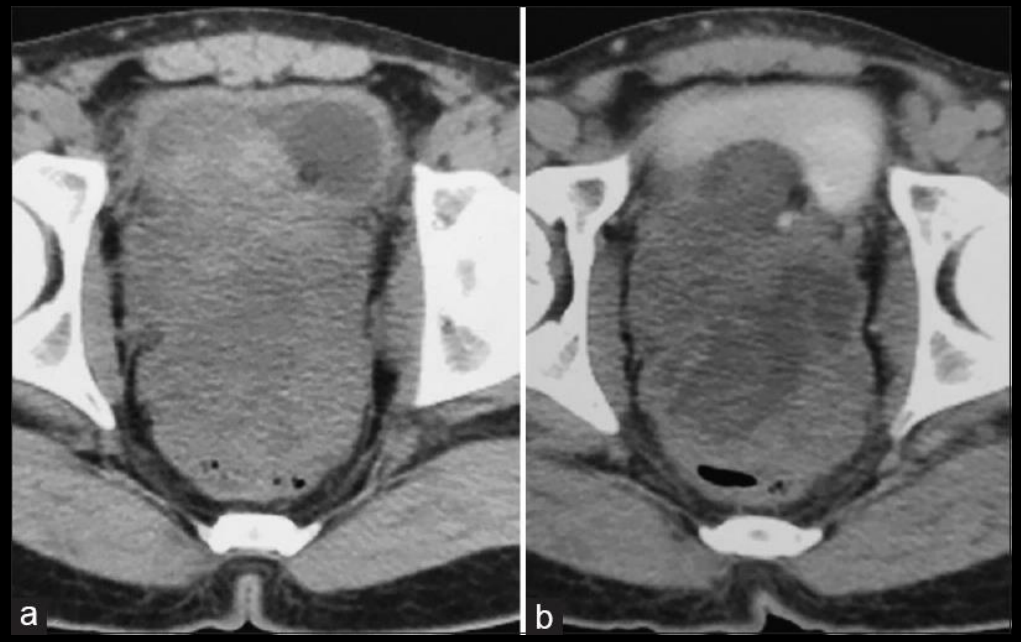
- **Treatment:**
  - Depends on pathology, however the patient usually needs both chemotherapy and surgery.

Genitourinary Rhabdomyosarcoma

Reference: Nerli, et al.

- Pelvic masses with **multilobulated, heterogeneous enhancement**

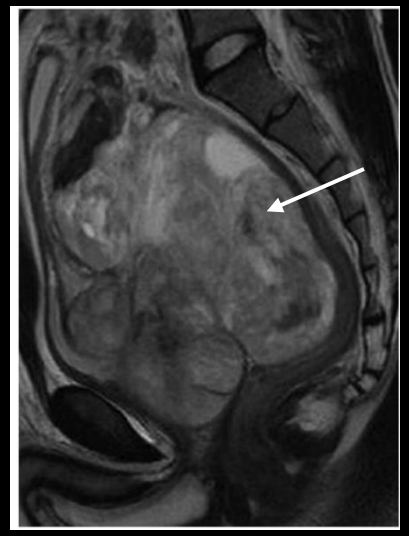
- **CT:**
  - Soft tissue attenuation
  - Contrast enhancement
  - Adjacent bone destruction in >20% of cases



CT of pelvis show a large, heterogeneously enhancing mass with a cystic/necrotic area at the center

Rhabdomyosarcoma

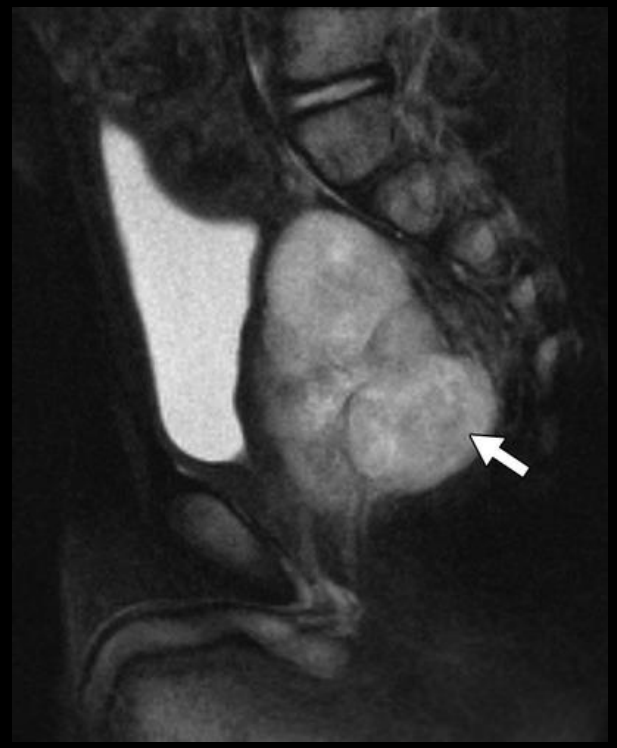
- **T1 MRI:** low to intermediate intensity, isointense to muscle
- **T2 MRI:** hyperintense to muscle
- **Embryonal subtypes** are more homogenous.
- **Alveolar subtypes** may have areas of necrosis, hemorrhage, and cystic changes.



Reference: Kieran, et al.

Reference: Chu, et al.

Sagittal T1-weighted MR image



Sagittal T2-weighted MR image

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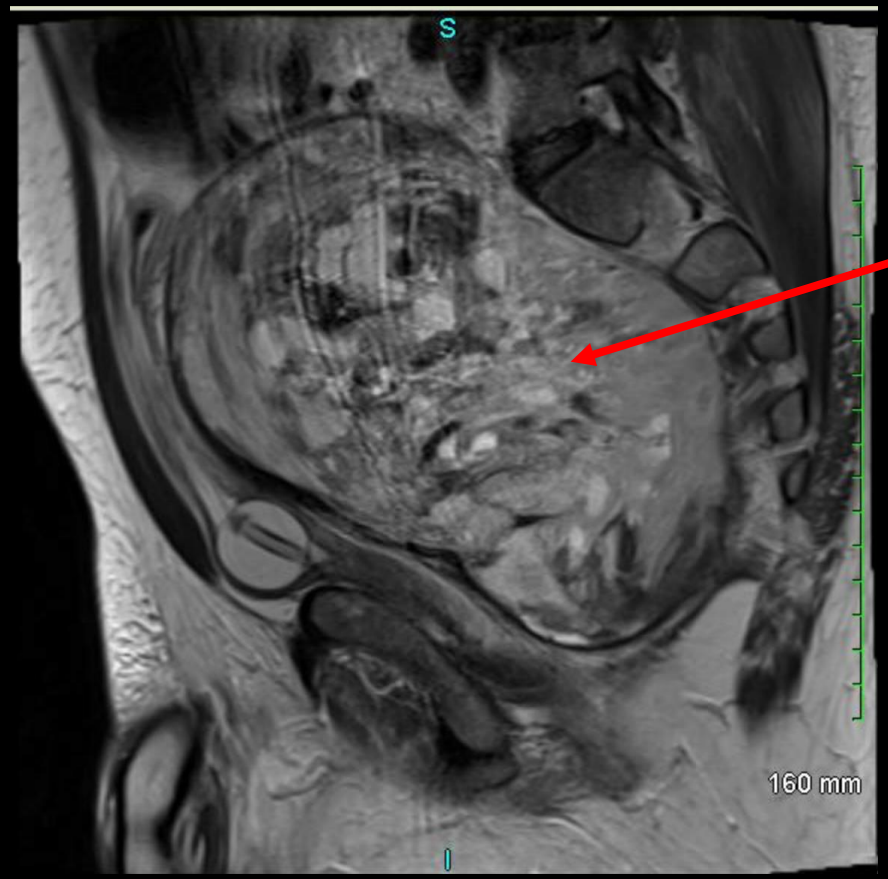
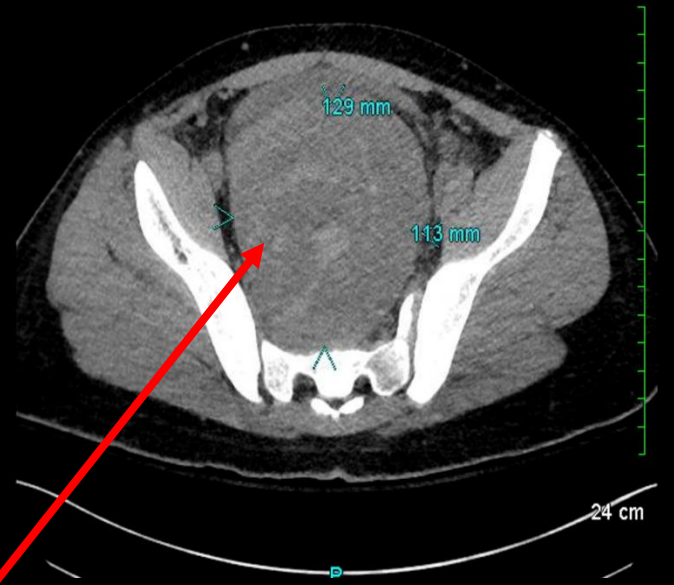
Our Patient – CT and MRI



Bulky Pelvic Mass

18.7 cm x 12.6cm x 16.3 cm AP
Heterogeneous enhancement
Cystic/necrotic areas within the mass



Pelvic Mass

Soft tissue attenuation with mild heterogeneous enhancement

References:

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4. Sharma S, et al. Round Cell Tumors: Classification and Immunohistochemistry. Indian J Med Paediatric Oncol 2017 38(3): 349-353.
5. Nerli R B, Sanikop AC, Kadeli V, Ghagane SC, Dixit NS, Hiremath MB. Alveolar Rhabdomyosarcoma of the Prostate in a Young Adult Presenting with Acute Retention of Urine. Clin Cancer Investig J 2018;7:77-80
6. Kieran K, Shnorhavorian M. Current Standards of Care in Bladder and Prostate Rhabdomyosarcoma. Urologic Oncology 2016; 34: 93-102.
7. UpToDate: Rhabdomyosarcoma in children and adolescence: Clinical presentation, diagnostic evaluation, and staging.