BOTRYOID RHABDOMYOSARCOMA OF THE BLADDER

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Agenda

- Definition / Incidence
- Types of Rhabdomyosarcoma
- Our Patient Presentation
- Differential Diagnosis based of Patient Presentation
- Radiographic Modalities Pre-op / Post-op
- Other locations of the tumor and subsequent radiographic films
- Prognosis
- Summary

Definition / Incidence

Definition:

- Rhabdomyosarcoma is a type of malignant tumor that usually arises from primitive muscle cells.
- Idiopathic in nature
- Can also arise from other areas that lack skeletal muscles such as genitourinary tract, head and neck.

Incidence:

- Very rare - 6 cases per 1 000 000 population.
- 250 cases diagnosed per year in the United States.

Source: http://emedicine.medscape.com/article/988803-overview

Types of Rhabdomyosarcoma

3 Types of Rhabdomyosarcoma:

- **Embryonal rhabdomyosarcoma – occurs in children**
  - Botryoid rhabdomyosarcoma – Our Patient
  - Spindle cell rhabdomyosarcoma
  - Anaplastic rhabdomyosarcoma
- **Alveolar rhabdomyosarcoma – occurs in adolescents**
- **Pleomorphic rhabdomyosarcoma**

Source: http://radiopaedia.org/articles/rhabdomyosarcoma

Our Patient: Presentation

• Ms. S is a 2yr old girl with history of bleeding and mass coming out of the introitus.

• The mass was thought to be urethral prolapse and she was prescribed Premarin Cream (conjugated estrogens).

• After 3 weeks, she presents with increased bleeding and increase in the size of the mass.

• On physical exam, the mass was found to be sub-urethral.

Therefore, an ULTRASOUND was done followed by other modalities to confirm the diagnosis.

Differential Diagnosis

Based on the Patient Presentation:

- Rhabdomyosarcoma
- Fibroepithelial Polyp
- Metastasis
- Mesenchymal neoplasm

Source: http://gamuts.isradiology.org/data/H-103.htm

Radiographic Modalities

- Ultrasound
- MRI
- Voiding Cysto-Urethrogram (VCUG)
- CT Scan
- Bone Scan
- After Treatment - Loopogram

Ultrasound

One of the **first modalities** used to diagnose pathologies in the bladder.

Benefits:

1. Easy to do and painless
2. Zero radiation
3. Perfect for identifying tumors in the pelvis compared to chest as the ribs obstruct the sound waves.

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Normal Ultrasound of Bladder

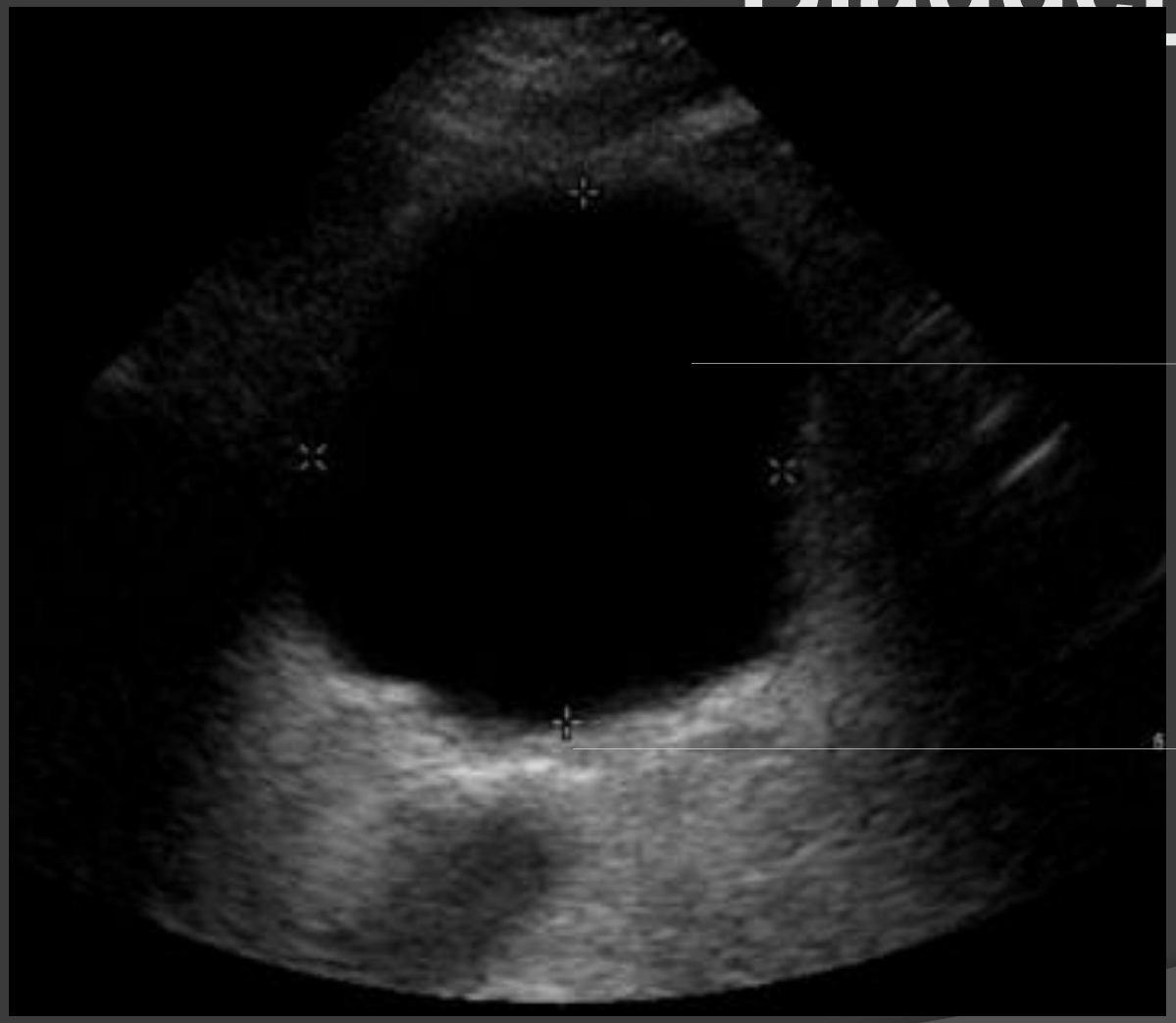

Image Source : http://www.med-ed.virginia.edu/courses/rad/gu/anatomy/bladder.html

Normal Ultrasound of the Bladder

- **Normal fluid-filled bladder**
  - **Anechoic structure**
- **Posterior Bladder Wall**
  - **Hyperechoic in nature**
  - **Normal wall thickness**

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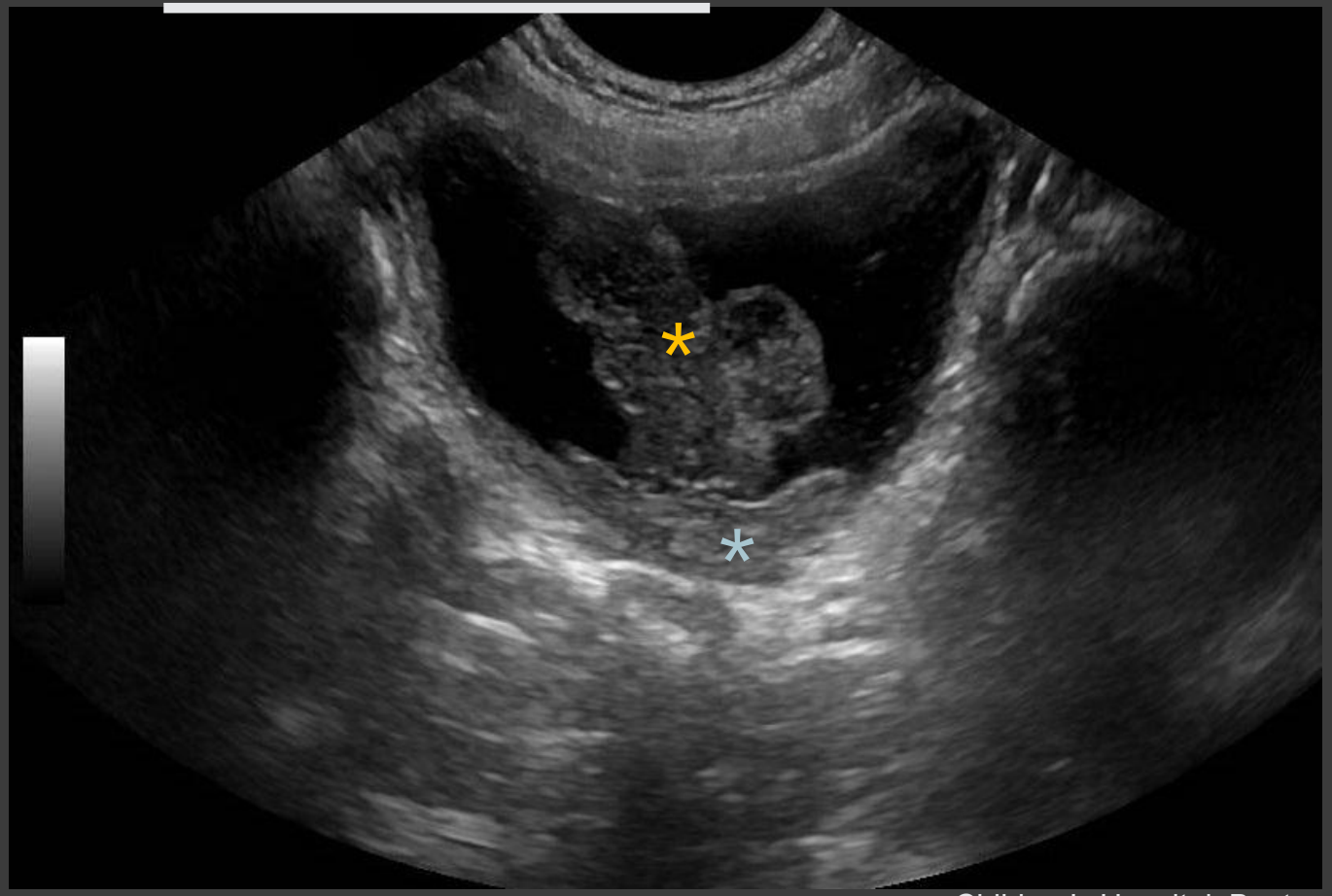
Our Patient – Mass on Ultrasound



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- Mass at the base of the bladder
  - Attached to posterior wall
  - Lobulated Appearance
  - Heterogenous in echogenecity
- Posterior Bladder wall thickness

After the ultrasound, biopsy was done which confirmed a Embryonal Rhabdomyosarcoma.

This was followed by an MRI.

Magnetic Resonance Imaging (MRI)

Advantages:

1. Better suited than the CT scan for imaging tumours
2. Superior to CT scan for imaging soft tissues
3. Good soft tissue differentiation
4. No Radiation

Disadvantages:

1. Takes a long time
2. Less detail of bony structures

Source: http://www.diffen.com/difference/CT_Scan_vs_MRI

Our Patient: Tumor Mass on MRI

Fat-Saturated Post-Contrast T1 Weighted Image

Axial

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- **Multi-lobulated mass in the lumen**
- **Classic Grape-like appearance:**
  - **Boytroid type of Embryonal RMS**

Our Patient: Tumor Mass on MRI

Fat-Saturated Post-Contrast T1 Weighted Image

- **Multi-lobulated mass at the base of bladder**
- **In the Trigonal Area**
- **Grape-like configuration**

Urethra dilated

Sagittal

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Our Patient: Tumor Mass on MRI

Fat-Saturated T2 Weighted Image

- **Multi-cystic mass extending peripherally in the lumen**
- **Grape-like appearance**

Urethra dilated

Sagittal

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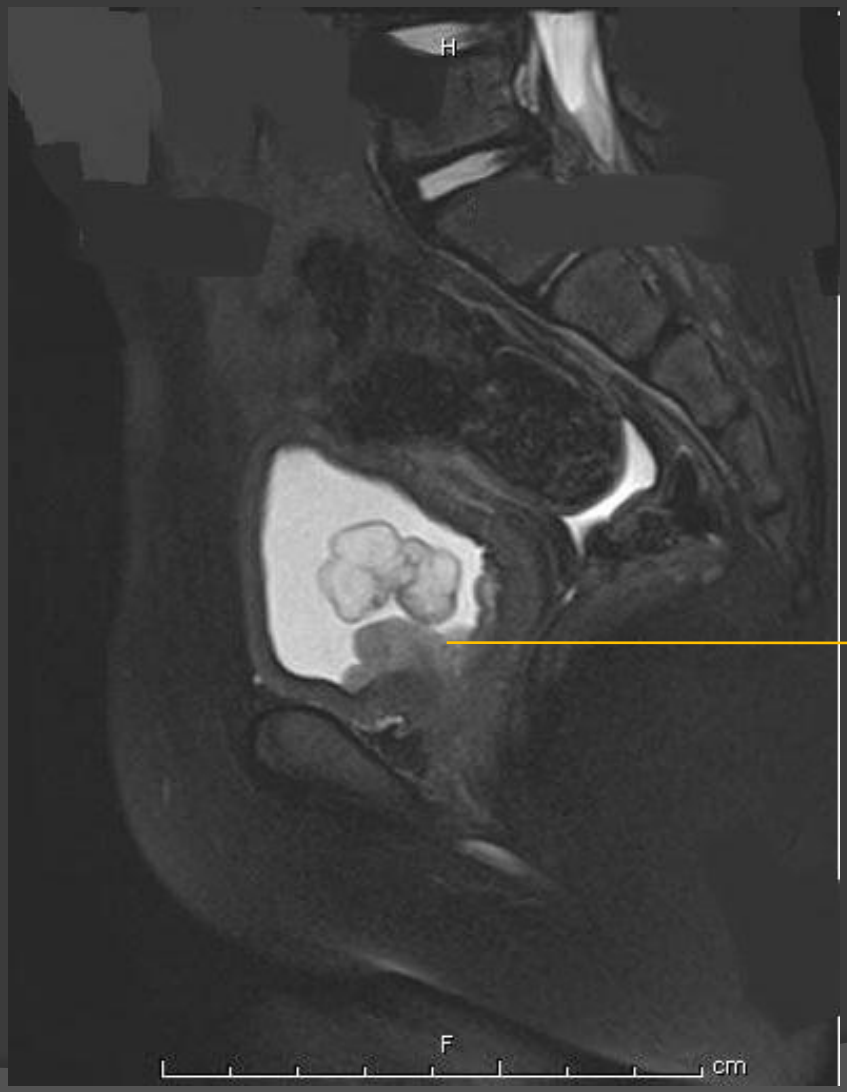
Our Patient: Tumor Mass on MRI

Fat-Saturated T2 Weighted Image



- **Plate-like portion in the bladder base**

  - **Extending in the urethra**

Sagittal

Children's Hospital, Boston

Summary of MRI findings

- Multi-lobulated mass seen in the bladder base
- Present in Trigonal Area.
- Urethra Dilated.
- Mass measured 2 x 3.5 x 3.6 cm **(less than 5 cm)**

Source: Children's Hospital, Boston

This was followed by a Voiding Cystourethrogram (VCUG)

Voiding Cystourethrogram (VCUG)

One of the best modalities for assessing the structure of the genito-urinary system.

Advantages:

1. Provides detailed information about the conditions of the genito-urinary system like
   - Tumors
   - Bladder Obstruction
   - Vesicoureteral Reflux
   - Stricture of the urethra
   - Stones

Disadvantages:

1. Discomfort after the procedure.

VCUG Procedure

- Urethral catheterization is done
- The bladder is filled with approximately 250cc of contrast material (CystoConray)
- Fluoroscopic images are used to determine any anatomical variants or presence of tumors, stones or a reflux.

Source: Children's Hospital, Boston

Our Patient: Tumor Mass on VCUG

Children's Hospital, Boston

- **Lobulated filling defect seen in the inferior part of bladder**
  - **Present in trigonal area**
- **No Vesicoureteral reflux seen**

Urinary catheter

Our Patient: Tumor Mass on VCUG

- **Lobulated filling defect seen in the inferior part of bladder**
  - **Present in trigonal area**
- **No Vesicoureteral reflux seen**

Urinary catheter

Next, a CT Scan was done

CT Scan

Advantages:

- Provides good detail of all internal structures like soft tissue, bone and the blood vessels
- Takes a short time compared to MRI

Disadvantages:

- Exposure to ionizing radiation

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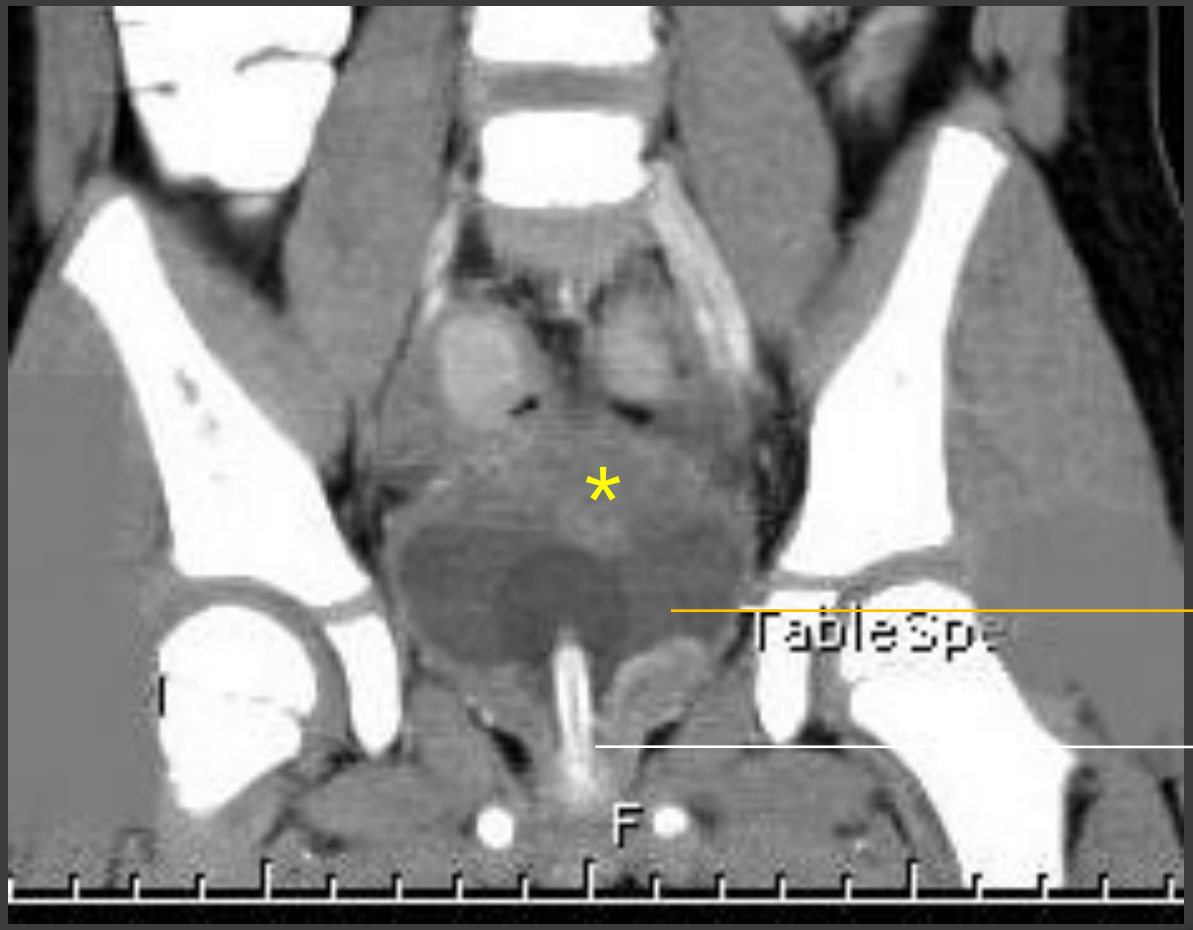
Our Patient: Tumor Mass on CT Scan

CT Abdomen



Coronal

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* **Thickening of the superior bladder wall**

Tumor Mass seen at the bladder base

Foley's Catheter

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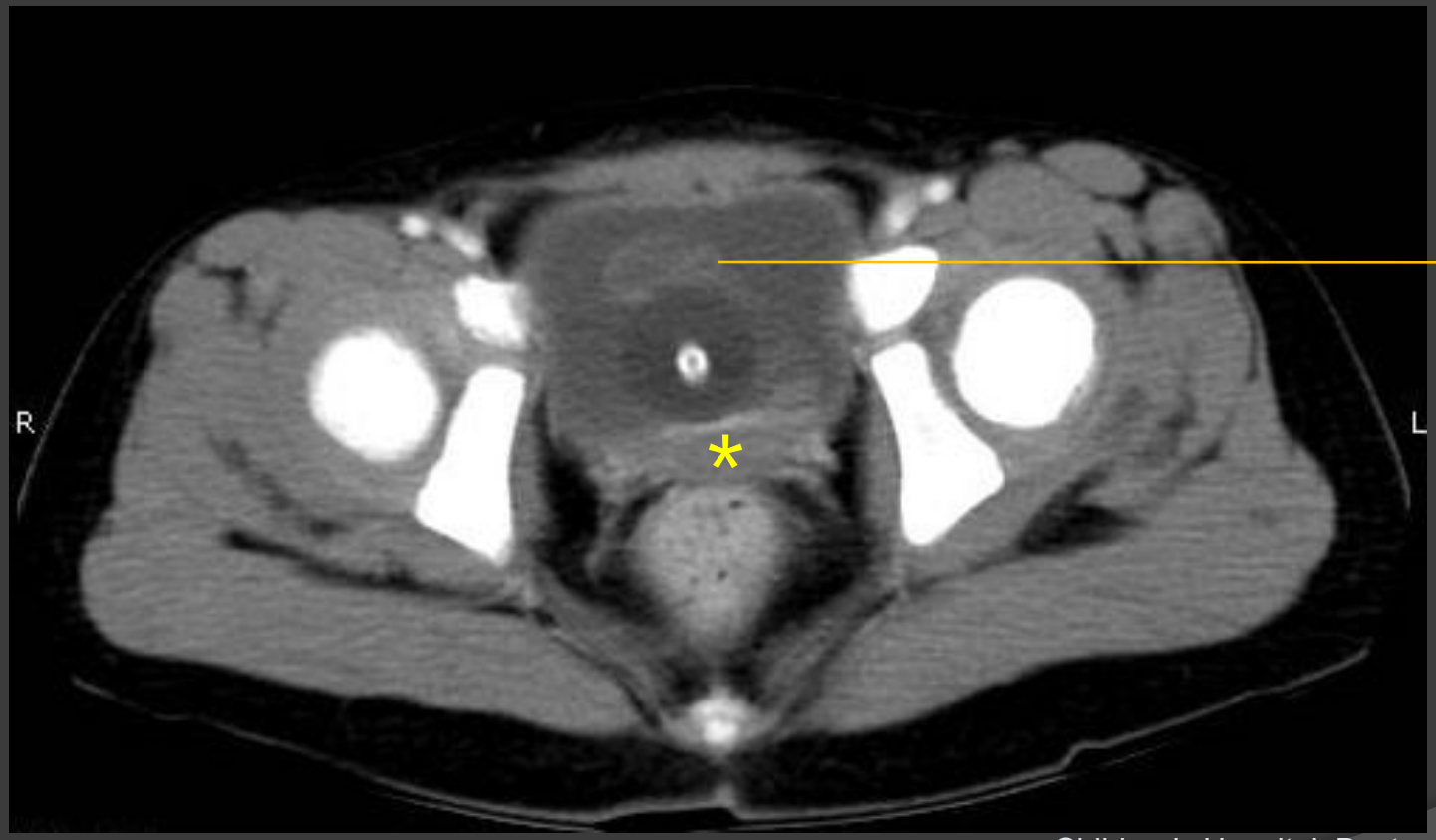
Our Patient: Tumor Mass on CT Scan

CT Abdomen



Axial

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Tumor Mass seen in the anterior aspect

*** Thickening of posterior wall seen**

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Our Patient: Tumor Mass on CT Scan

CT Abdomen

Sagittal

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* **Thickening of the superior and posterior bladder wall**

Tumor Mass seen anteriorly

LETS LOOK AT WHAT WE KNOW SO FAR

4 Stages of Rhabdomyosarcoma

- **Stage I**: N0 , M0
  - Orbit
  - Eyelid
  - Head and neck (excluding parameningeal),
  - Genitourinary (non-bladder, non-prostate)

- **Stage II** : **< 5 cm**, N0, M0
  - **Bladder**
  - Prostate
  - Extremity
  - Parameningeal

- **Stage III** : > 5 cm, N0 or 1, M0
  - Bladder, prostate, extremity, trunk, parameningeal

- **Stage IV** : all others, any N, M1

Source: http://radiopaedia.org/articles/rhabdomyosarcoma_staging

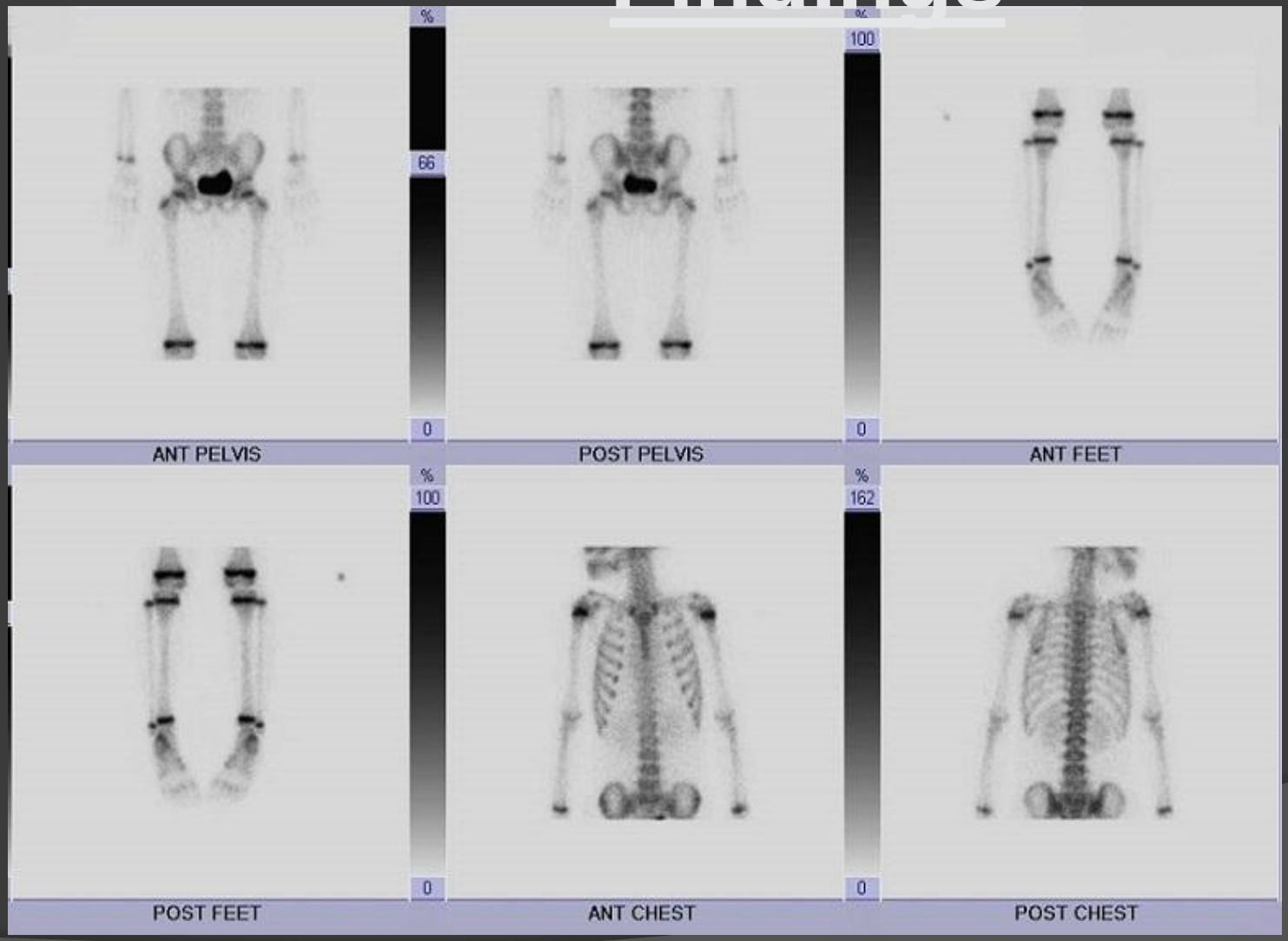
This tumor is Stage II as it's less than 5 cm and is localized to the bladder.

To rule out metastasis, Bone Scan was the next investigation done.

Bone Scan

- Patient is injected a small amount of **radiographic material (tracer)**.
- About half of the tracer localizes in the bones
- The rest is excreted via the kidneys and bladder.
- Useful for identifying **bone lesions / metastasis** in the bones.

Our Patient: Bone Scan Findings



No metastasis seen

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Treatment

- Complete resection of the bladder and removal of tumor
- Sigmoid conduit made as a urinary diversion.
- Ureters attached to the conduit in a uretero-sigmoid anastomosis.
- Stoma present in the patient's left lower quadrant attached to a urostomy bag
- Patient stays on urostomy bag and oral antibiotics for life.
- Regular follow-ups.

After treatment, the patient was followed with regular follow-ups including MRI and Loopogram

Our Patient: Post – Op MRI

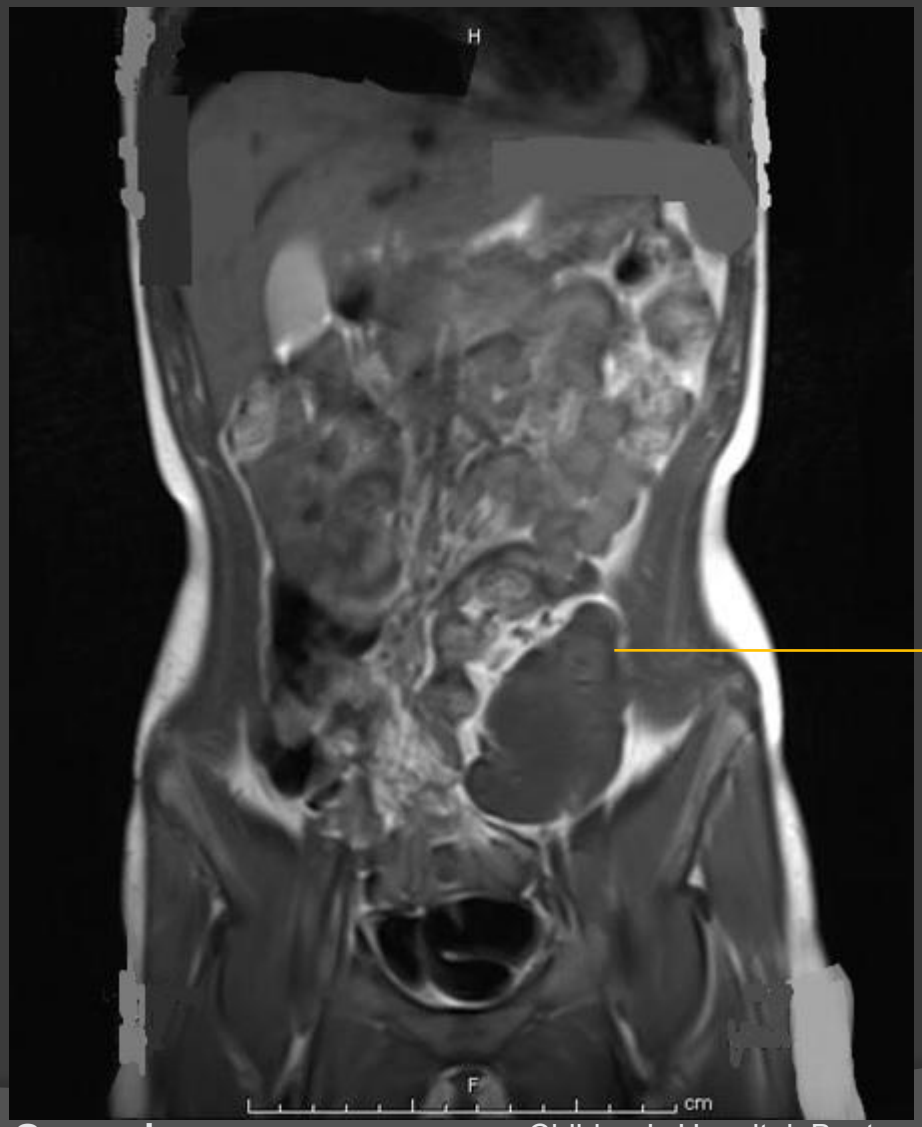
Fat-Saturated Post-Contrast T2 Weighted Image

Coronal

- **Sigmoid conduit present in the left lower quadrant**
  - **Contains urine**
- **No lesions or masses identified in the neobladder**

Our Patient: Post – Op MRI

T1 Weighted Image

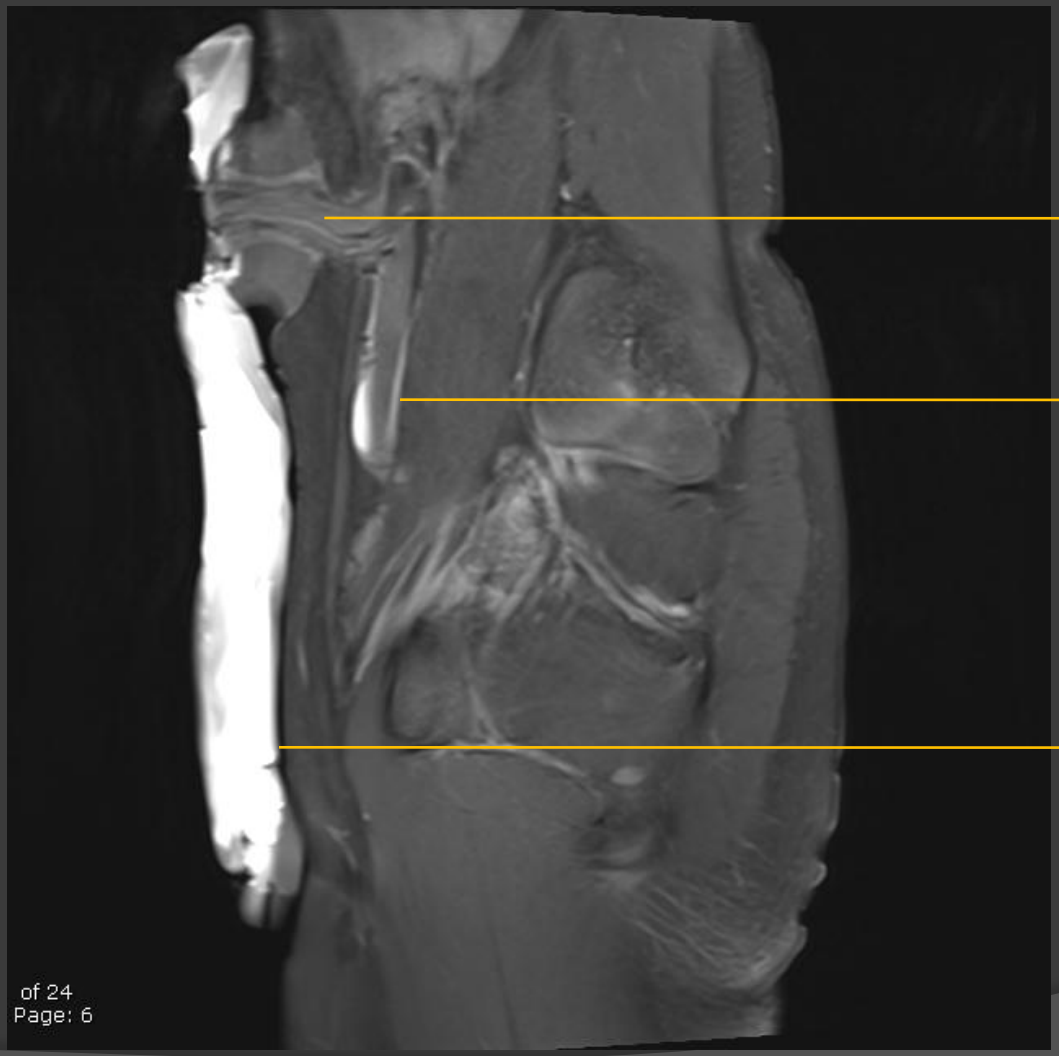


Coronal

- **Sigmoid conduit present in the left lower quadrant**
  - **Contains urine**
- **No lesions or masses identified in the neobladder**

Our Patient: Post – Op MRI

Post contrast T1 Image



Conduit connected to Urostomy bag

Sigmoid conduit

Urostomy bag

Sagittal

Loopogram

Fluoroscopic procedure to determine the anatomy and functionality of the sigmoid conduit.

Indications for Loopogram:

- Follow-up after surgery
- Difficulty emptying the conduit
- Vesicoureteral reflux

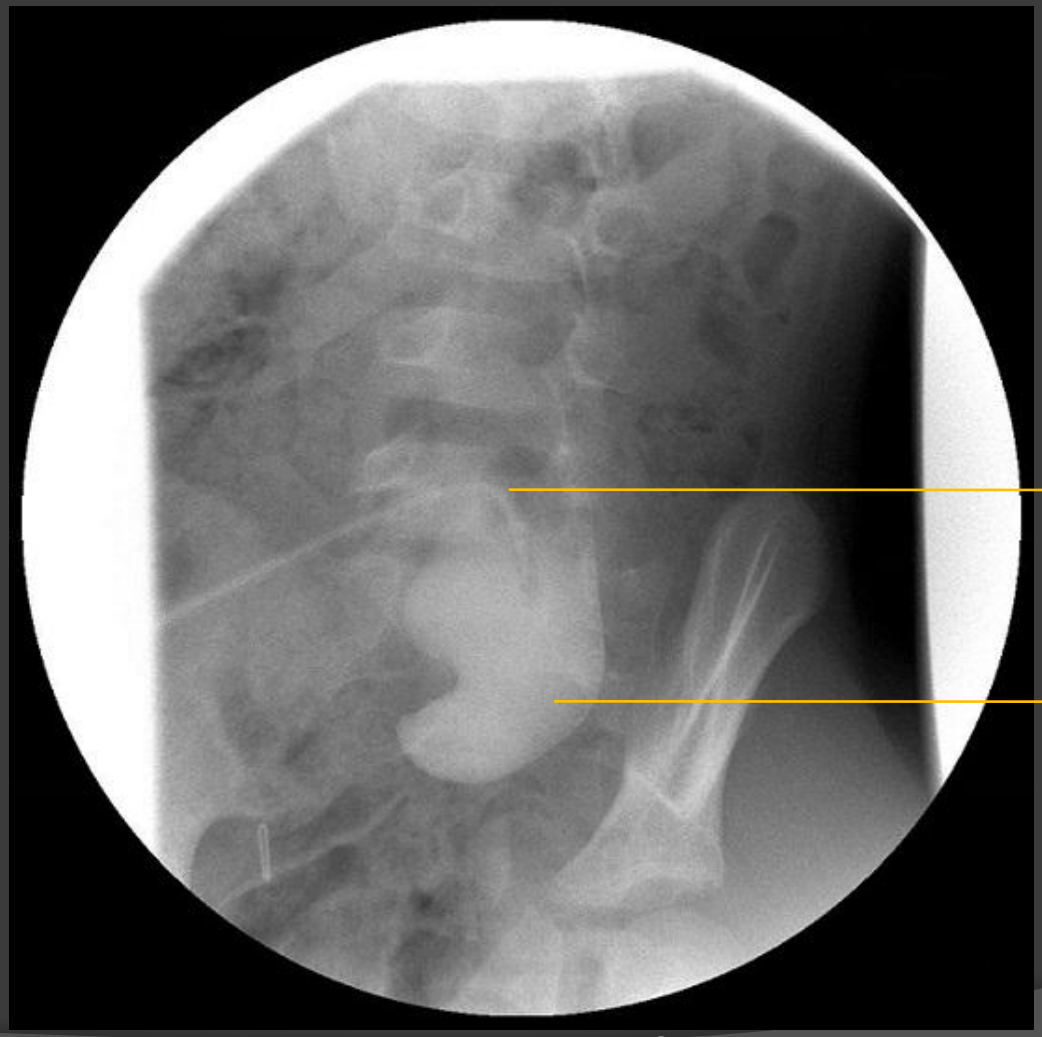
Source: http://www.unc.org/publications/1/UNC_Pipeline_2001FINAL.pdf

Procedure for Loopogram

- 12 French Foley Catheter inserted into the stoma.
- Balloon inflated and mild traction is given.
- Contrast material (CystoConray) instilled into the conduit.
- Fluoroscopic images are taken to visualise the anatomy and functionality of the conduit.

Source: http://www.unc.org/publications/1/UNC_Pipeline_2001FINAL.pdf

Our Patient: Post–Op Loopogram



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Findings :

- Smooth shape of sigmoid conduit
- No filling defect seen

Catheter

Sigmoid conduit filling with contrast agent

Next, we will look at a major complication that can occur with this treatment

Complications

Vesicoureteral Reflux

- **Grades:**

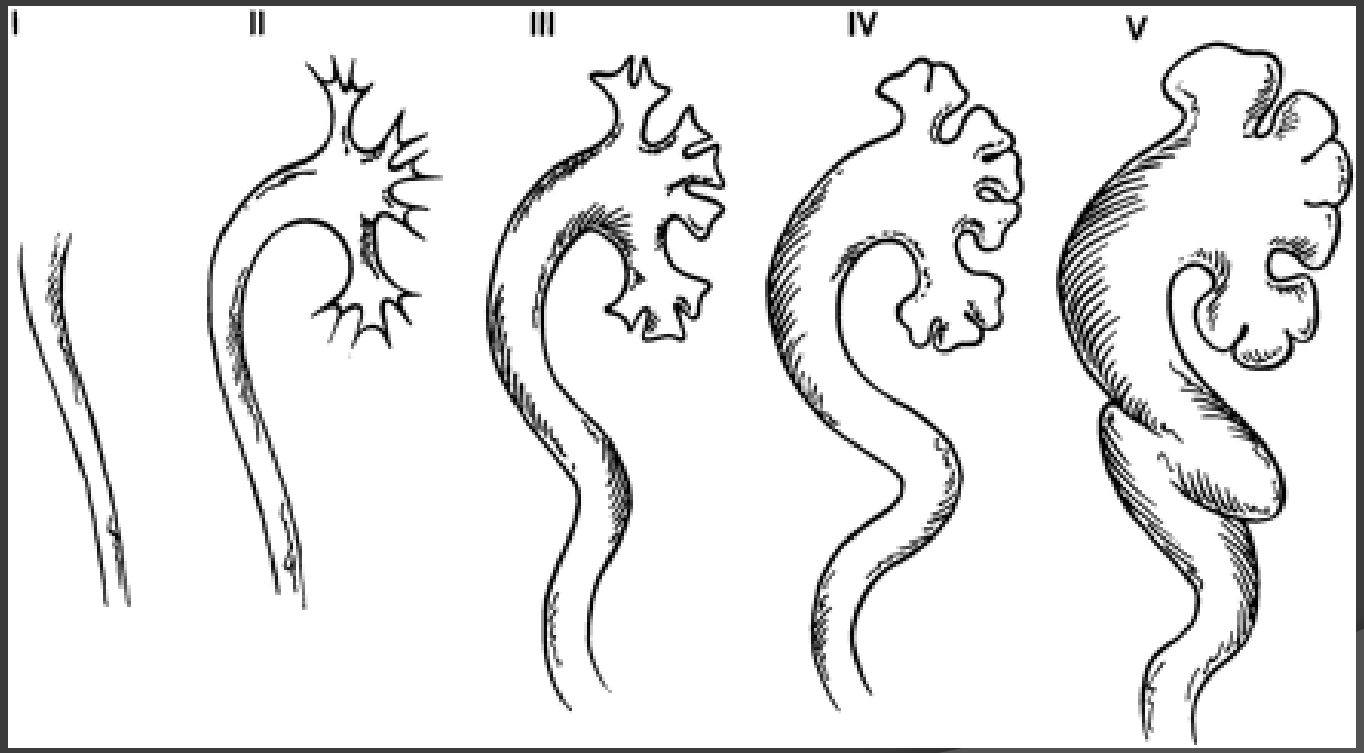

Image Source: http://radiographics.rsna.org/content/20/1/155/F10.expansion.html

Now, we come to the <u>other locations</u> where this tumor can occur

Other Locations

Can occur almost anywhere in the body

- **Usual**
  - **Testicular**
  - **Orbital**
  - **Parameningeal**
  - **Extremities**
- **Unusual**
  - **Pancreas**

Source: http://www.ajronline.org/content/176/6/1563.full

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Companion Patient #1

Testicular Ultrasound

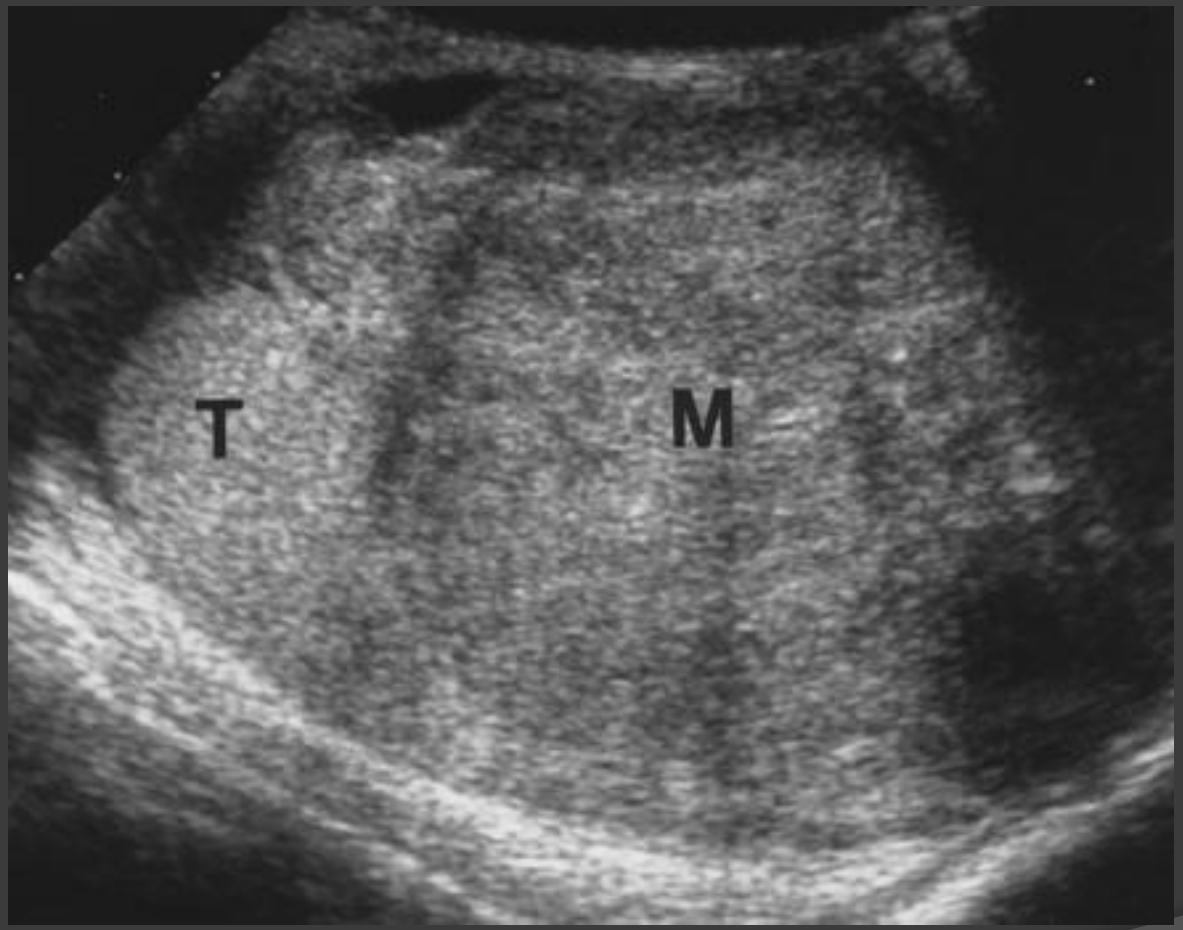

Image Source: http://www.ajronline.org/content/176/6/1563.full

In this patient, the mass was seen in the testicle as seen in the ultrasound.

Ultrasound

T – Testicle M – Mass

Findings:

- **Hypoechoic paratesticular mass**
- **Heterogenous in nature**

Source: http://www.ajronline.org/content/176/6/1563.full

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Companion Patient #2

Orbital Contrast-enhanced CT Scan

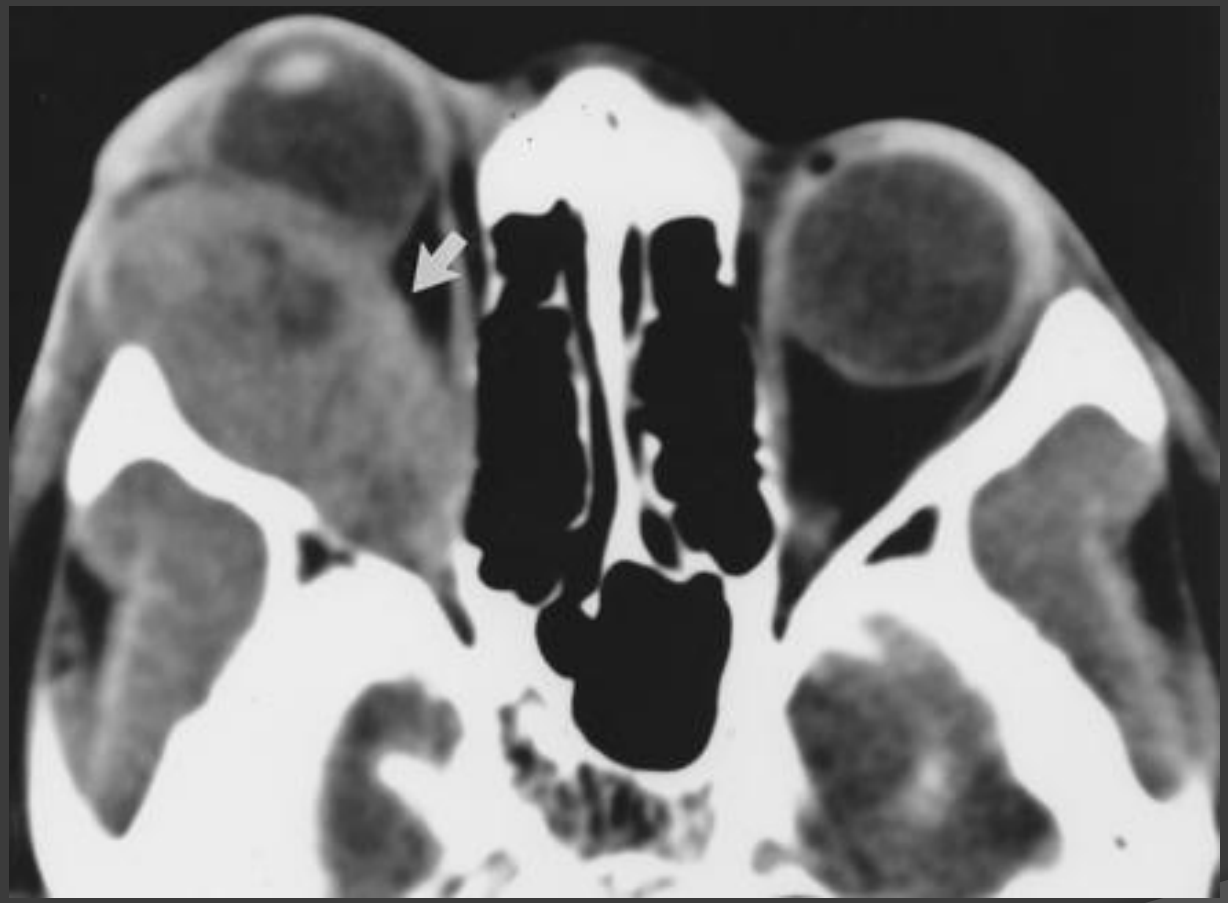

Image Source: http://www.ajronline.org/content/176/6/1563.full

In this case, the patient presented early as the mass caused proptosis and visual changes.

Arrow - Right Orbital Mass

Findings:

- **Heterogenous mass causing proptosis.**

Source: http://www.ajronline.org/content/176/6/1563.full

Companion Patient #3

Pancreas - Contrast-enhanced CT Scan

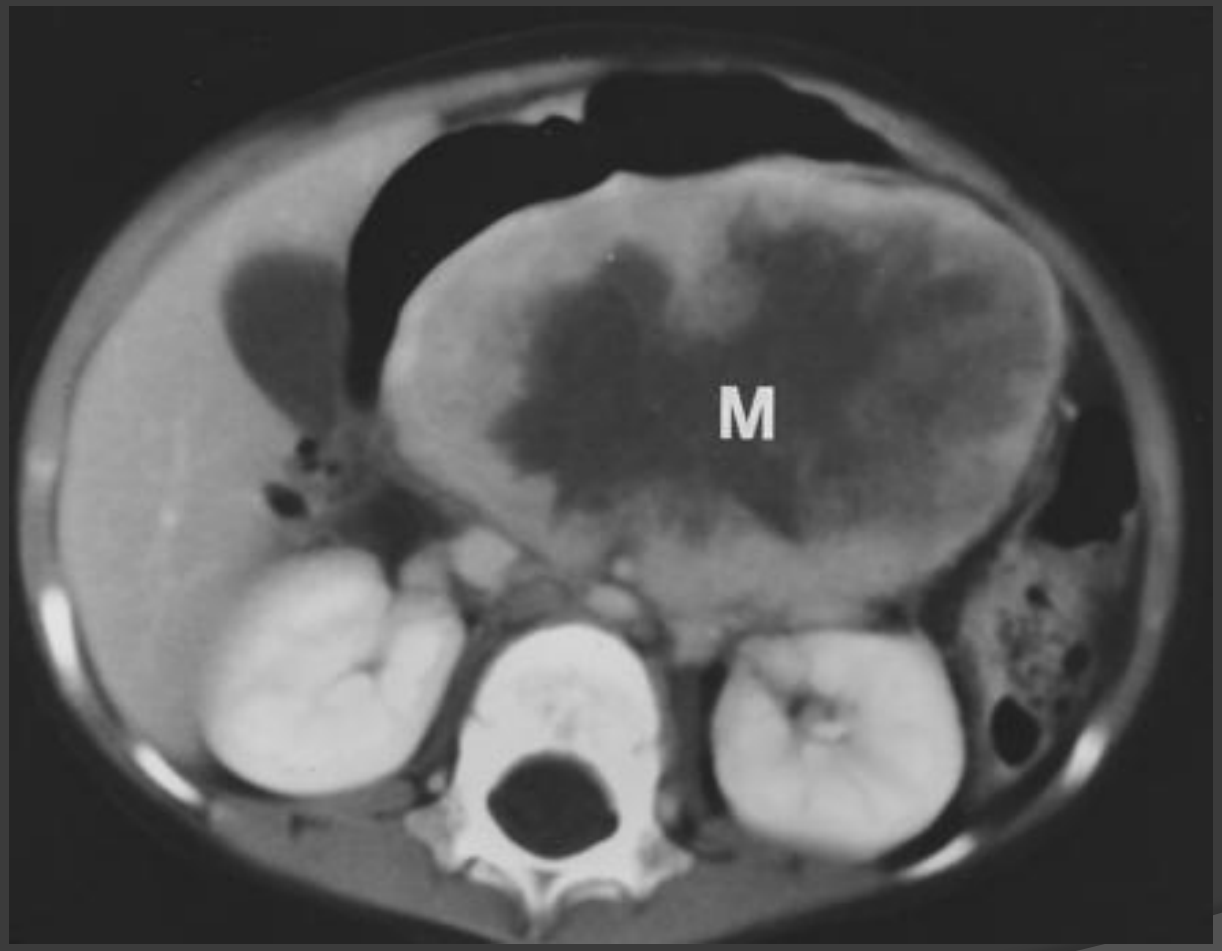

Image Source: http://www.ajronline.org/content/176/6/1563.full

M – Large Mass

Findings:

- **Large centrally necrosed mass**
  - **(biopsy showed pancreatic mass)**

Source: http://www.ajronline.org/content/176/6/1563.full

Prognosis of Rhabdomyosarcoma

Prognosis is different depending on the location and the age of the patient

LOCATION

- In patients with a localized disease – Prognosis good.
  - The 5-year survival rate - 80%
- In patients with metastatic disease – Prognosis poor
  - The 5-year survival rate - less than 30%

AGE

- Highest is children 1-4 years of age – 77%
- Poor in infants and adolescents – 47%

Source: http://emedicine.medscape.com/article/988803-overview

Summary

- Rhabdomyosarcoma is a malignant tumor that can occur almost anywhere in the body.
- Signs / Symptoms depend on the location of the tumor.
- Treatment varies depending on the location and the stage of the tumor.
- Prognosis is better for localized disease compared to metastatic one and better for children 1-4 years of age compared to adolescents.

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