Common Presentations of Childhood Cancer

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Medical Update
UOM
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First Take Home point

- **Childhood Cancer is a rare disease…**

- ✓ 1500 new cases of cancer in Mauritius/ year
- ✓ Only 40 cases (2,5 %) concern children

BUT…

- One in every 330 children develops cancer before age 20.

- 1 in 750 20-year-olds alive today is a survivor of childhood cancer in France.

More Statistics

* **Over 20 children die of cancer yearly in Mauritius**

The overall prognosis is better in child cancers but…

Earlier diagnosis and referral can still improve outcome

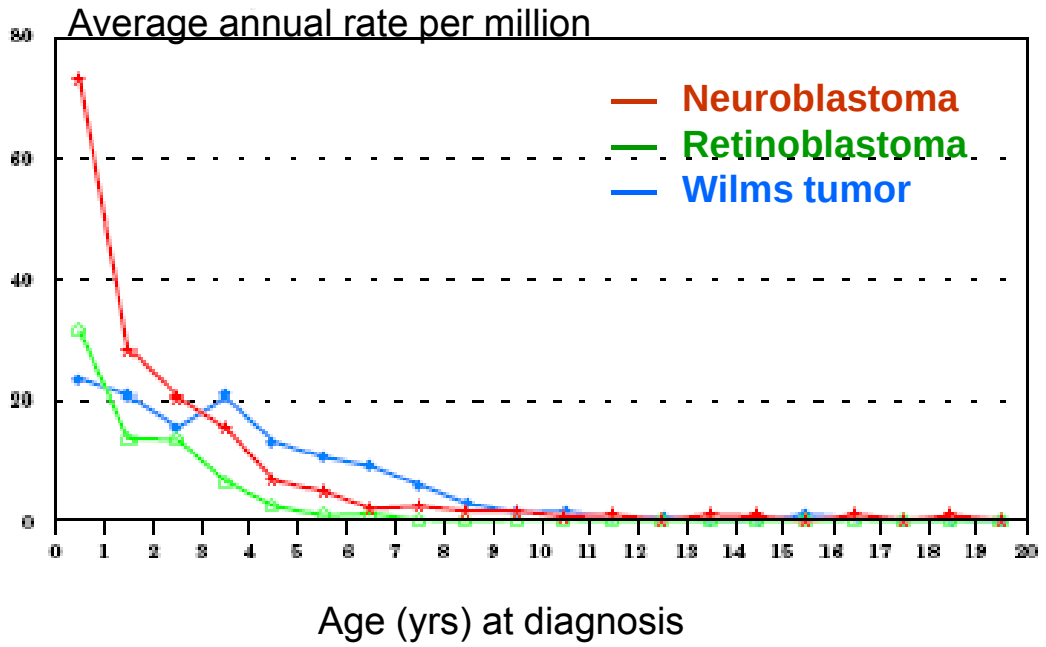
Second Take Home points…

- Leukemia is the most common childhood cancer
- Brain tumors are second most common
- Lymphomas are the third most common
- Then solid tumors outside the CNS
  - Neuroblastoma - neural crest derived
  - Wilms - renal tumors and syndromes
  - Bone tumors
  - Rhabdomyosarcoma - soft tissue sarcomas

Children and adults have very different solid tumors

- ***Children***
  - ***Blastoma*** : neoplasm arising from « blastema », ie embryonic tissue (kidney, neural tissue, liver,eye,…)
  - Mostly before 5 years
- ***Adults***
  - ***Adenocarcinomas*** and ***epitheliomas*** : lung, colorectal, breast, prostate
  - Important role of « external aggression » ex smoking

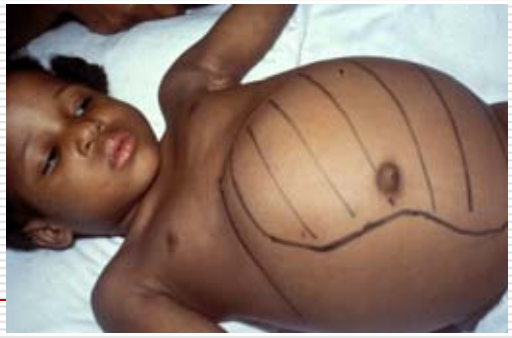
Cancer in Younger Children

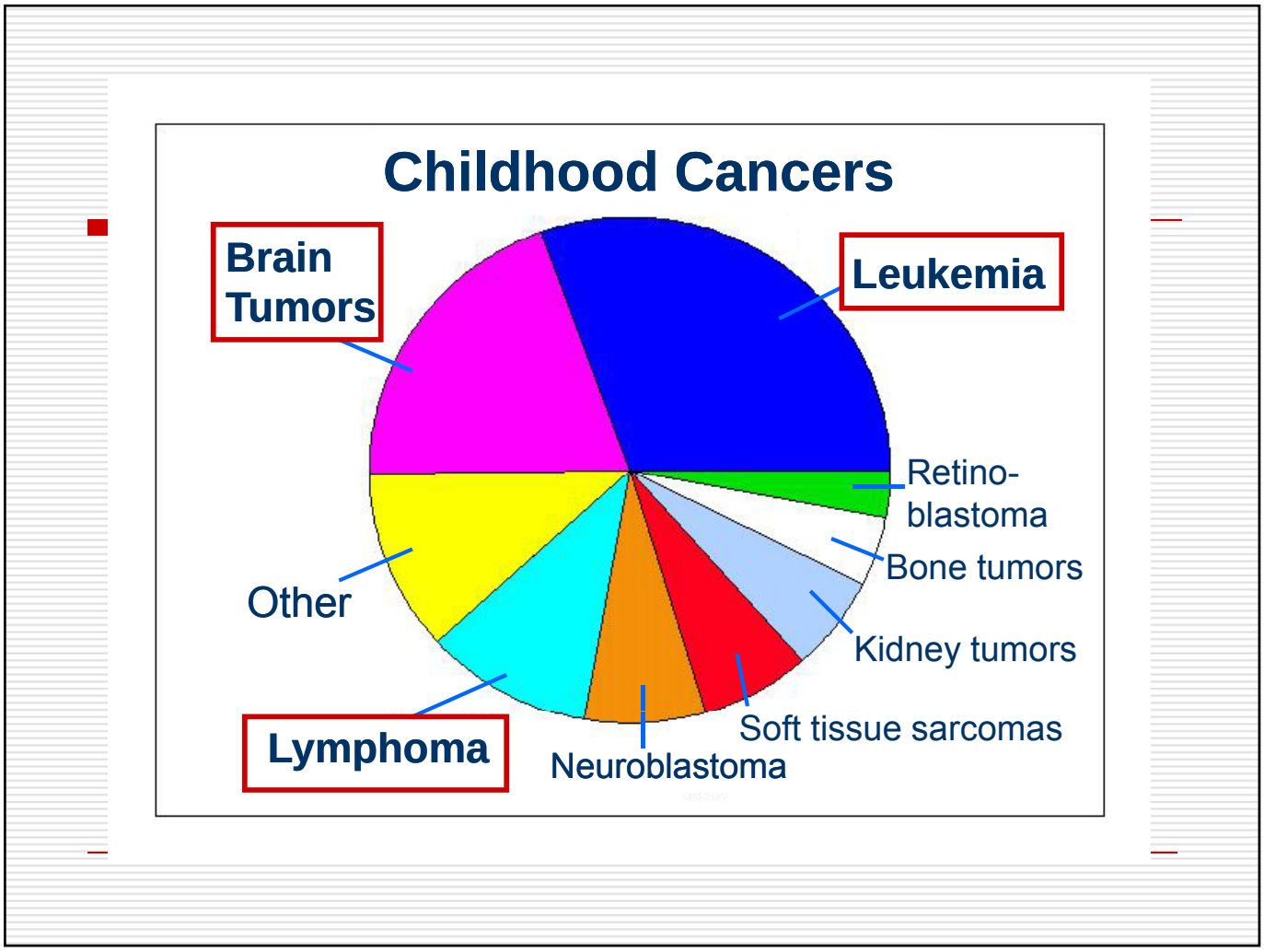


SEER data, 2002

« Blastomas » are different …

- May grow rapidly, and have often huge size at the time of diagnosis
  - prognosis is NOT worse
  - responds well to chemotherapy
- Child remains in a good general condition for a long time




Childhood Cancers
Brain Tumors
Leukemia
Retino-blastoma
Bone tumors
Kidney tumors
Soft tissue sarcomas
Neuroblastoma
Lymphoma
Other

Common risk factors of cancer in children

- Down's syndrome : ALL*20
- Undescended testis : germinal tumour*40, even after surgical correction
- Retinoblastoma : in 40% of cases, there is a mutation in the RB1 gene – autosomic dominant transmission – prenatal counselling if both parents are heterozygotes

- With advances in molecular biology and genetics, better understanding in the role oncogenes and tumor suppressor genes in the future.

Leukemias

Presentations of Lymphoblastic Leukemia

1 4 y/o fever x 4 days

ear pain = L otitis, began Amoxicillin

T = 39°C + chills next day → urgent care again

PE: L TM purulent drainage

bilat. cervical, L axillary, L inguinal LNs↑

spleen 5 cm ↓, liver 3 cm ↓

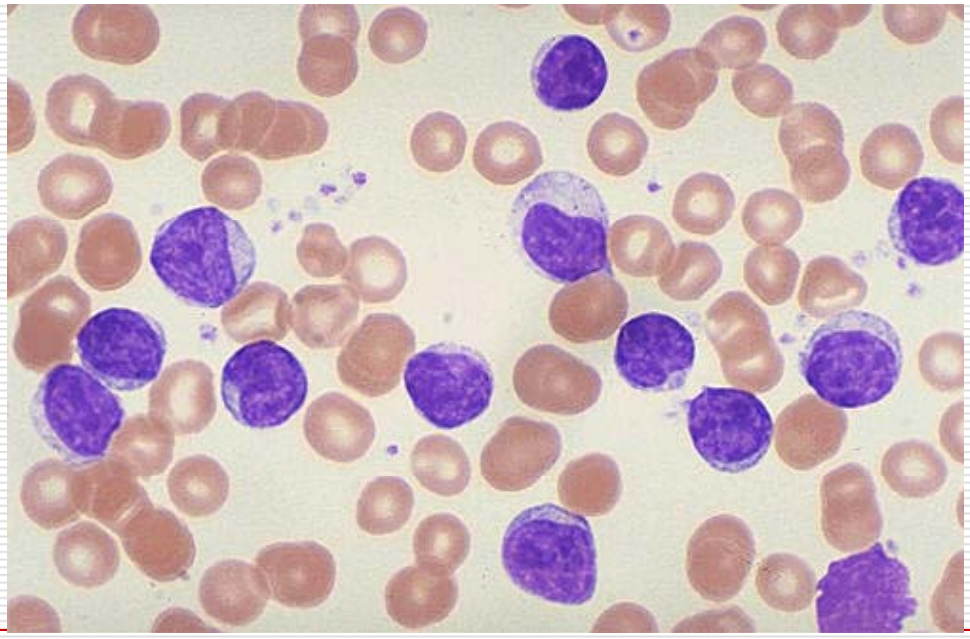
multiple bruises extremities

→ **Test**

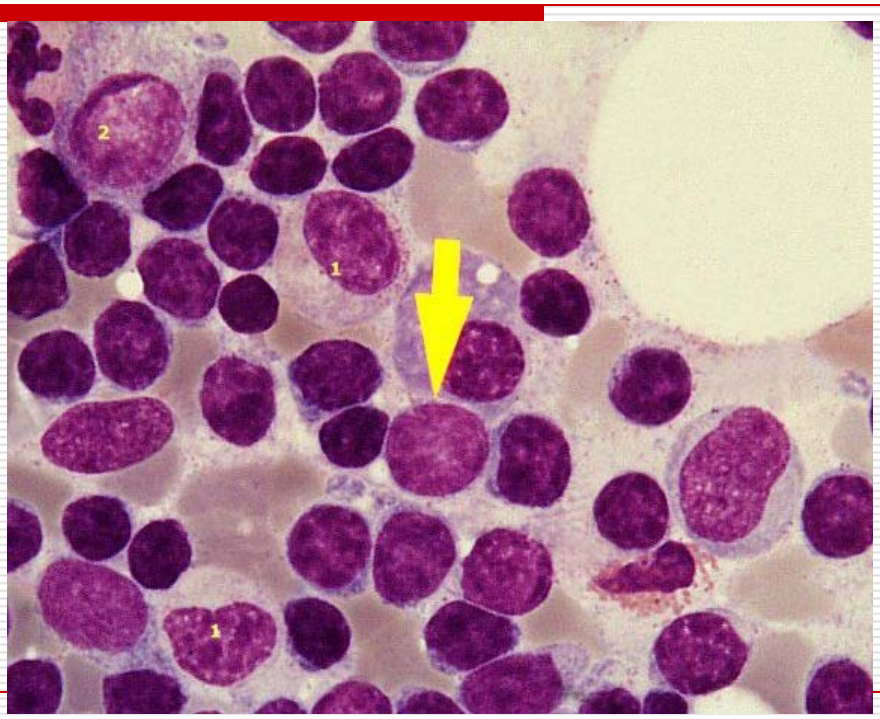
CBC

- WBC = 33 000
  - 8% Neut, 92% lymphocytes
- Hb = 6.5
- PLT = 40 000

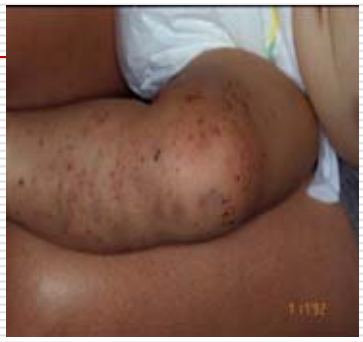
Peripheral smear show lymphoBLASTS



Bone marrow

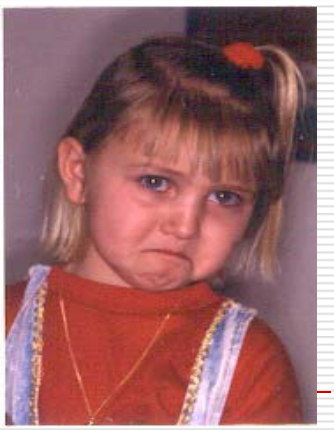


Leukemia: Signs and Symptoms



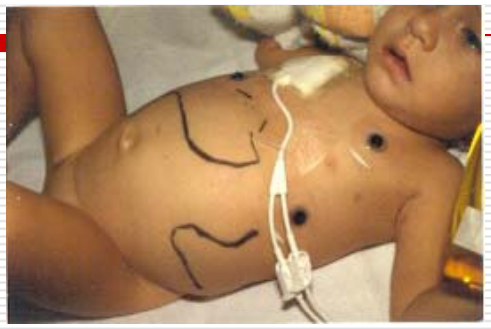
Bone marrow infiltration

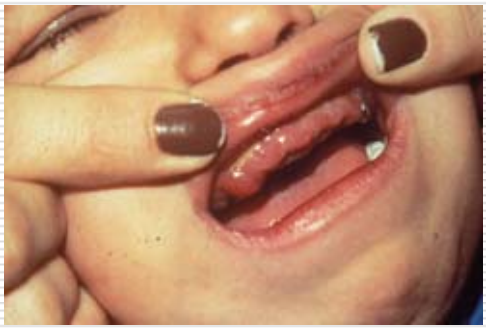
- **Anemia**
  - Pallor, lethargy
  - Dyspnea, ↑murmur
- **↓ Platelets**
  - Bleeding, petechiae, purpura
- **Neutropenia**
  - Fevers and infections
- **Bone pain**
  - Limp, ↓ walking, irritability



Leukemia: Signs and Symptoms

- **Extramedullary spread**
  - Lymphadenopathy
  - Hepatosplenomegaly
  - Dyspnea, cough
    - mediastinal mass
    - tracheal compression
  - Testicular enlargement
  - Skin lesions
  - Gingival hypertrophy
- **Fever of malignancy**





Monoblastic leukemia

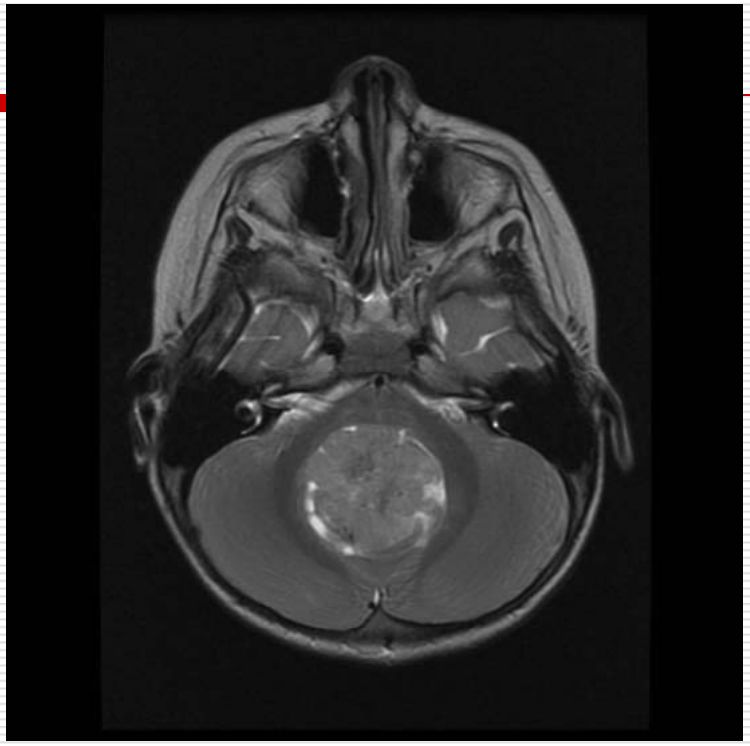
CNS Tumors

Presentation

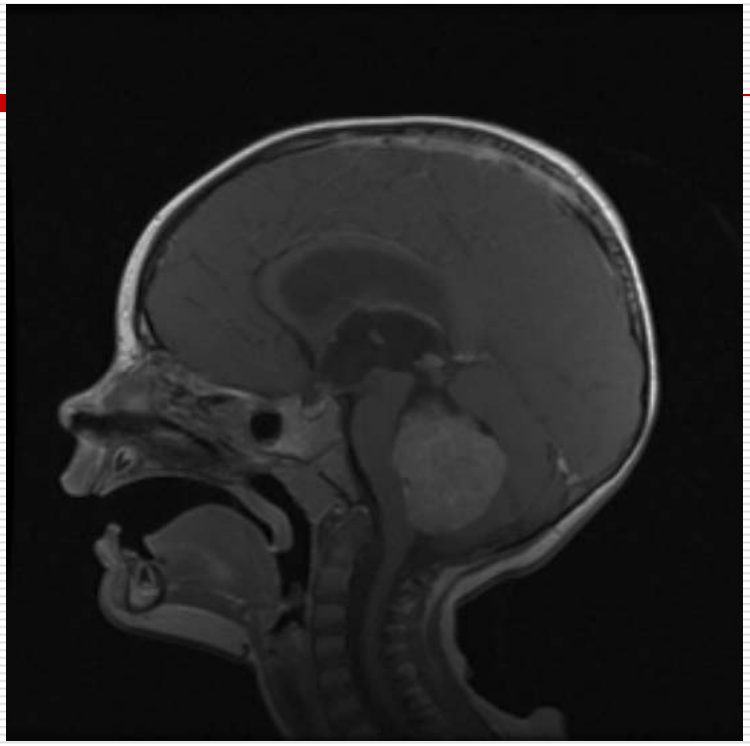
- 23mo female with ataxia X 1month
  - NB NB Emesis, in am X 2weeks
  - Vomiting am
  - Seen by PCP for GERD without improvement
  - No Fevers
  - Increasing vomiting

  - **Test**

MRI



MRI



Brain Tumors of Childhood

Heterogeneous

* Cell of origin:

glial+++, neural, other, combination

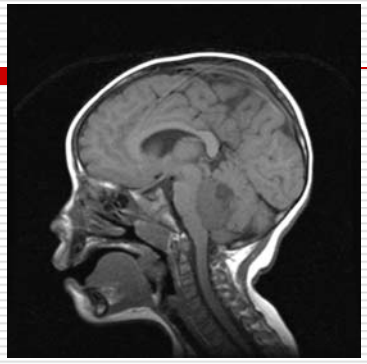
(medullobastoma, astrocytoma, glioblastoma, craniopharyngioma,…)

* Location:
  - posterior fossa: 50%
  - supratentorial: 50%

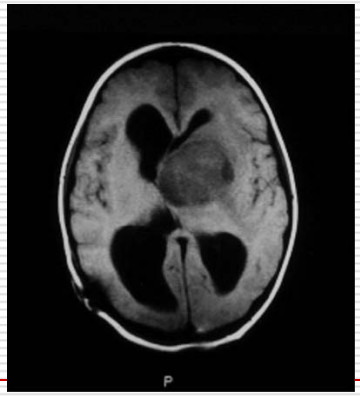
* Clinical presentation:
  - ~~**location**~~
  - **age**

Brain Tumors of Childhood

- **Infratentorial**
  - 50%
  - esp < 6 y/o



- **Supratentorial**
  - 50%
  - esp > 8 y/o

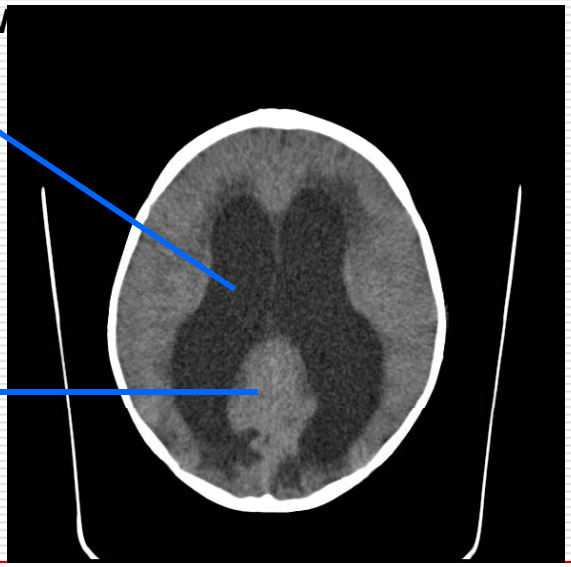


Nonlocalizing Signs of Brain Tumors

Increased intracranial pressure (ICP)

Obstructed CSF flow and hydrocephalus

Child is often asymptomatic until critical threshold reached



Medulloblastoma

Can grow very large before detection

Increased Intracranial Pressure (ICP)

- Headaches, progressively worsening
- Vomiting (morning)
- Irritability
- Papilledema
  - rare < 2 y/o - head can expand
- "Double vision" with 6th nerve palsy
- Head tilt
- Bulging fontanel (infant)

In a young child with ? brain tumor:
Measure head circumference and observe gait

Supratentorial Tumors

Signs depend on location and age

like in adults; in addition:

Younger child:

Developmental delay or loss of milestones

Older child:

Deteriorating school performance

Personality changes

Endocrinopathies:

DI, hypothyroidism, precocious puberty

Lymphomas

Presentation

- 16yo female with cough X 3 weeks
- Fevers for 2 months
- Wt loss X 2 months

- Exam noted to have supraclavicular LAD

- **Test**

Childhood Lymphomas

- **Signs and Symptoms depend on:**
  - Lymphoma subtype
    - Hodgkin's Disease (HD)
    - Nonhodgkin's Lymphoma (NHL)
      - **Burkitt's**
      - **Lymphoblastic**
      - **Anaplastic Large Cell**
  - Location

Presentation of Hodgkin's Disease

- **Age:** adolescents >> young child
- Painless lymphadenopathy
  - Progresses over weeks → months
- **Location**
  - **Cervical/supraclavicular ↑ LNS**
    - unilateral or bilateral
  - **Mediastinum ± hilum**

95%

  - LNs below diaphragm and spleen
  - Liver, lung, bone marrow

Presentation of Hodgkin's Disease

- Systemic symptoms
  - **Fevers**
  - **Night sweats**
  - **Weight loss**

  } **"B" symptoms** 25%
  - Pruritus

- Superior Mediastinal Syndrome (SMS)
  - Orthopnea, SOB, stridor, hypoxia
    - Tracheal
    - Bronchial
    - Cardiac

    } compression

    = Oncologic Emergency

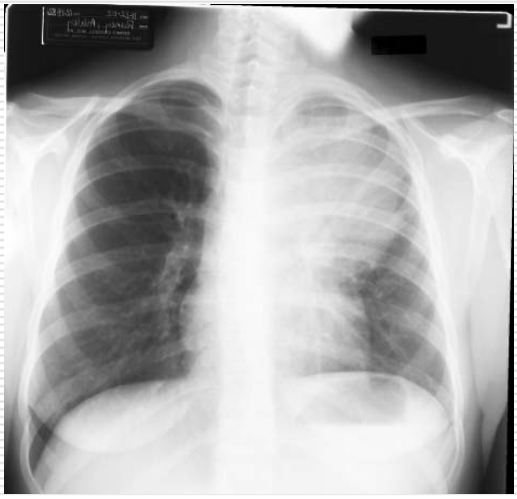
What is the Test to get?

HD in 16 y/o girl

↑ left cervical LNs, 40

wt loss

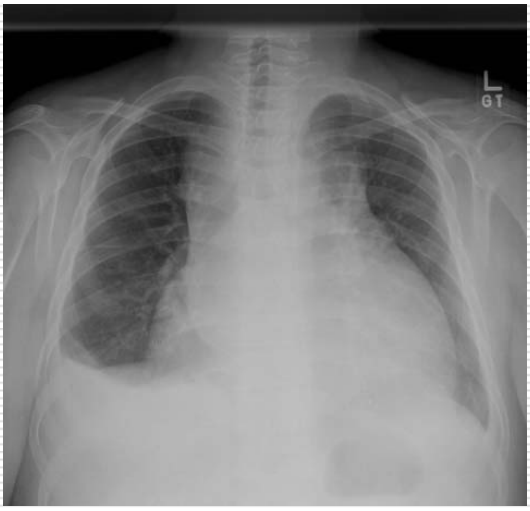
cough, no orthopnea



HD in 9 y/o boy

cough, fever, night sweats

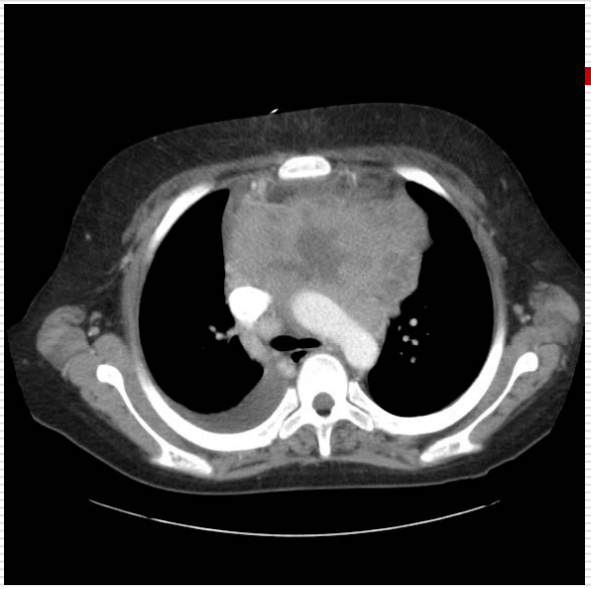
Pruritus shins, + orthopnea



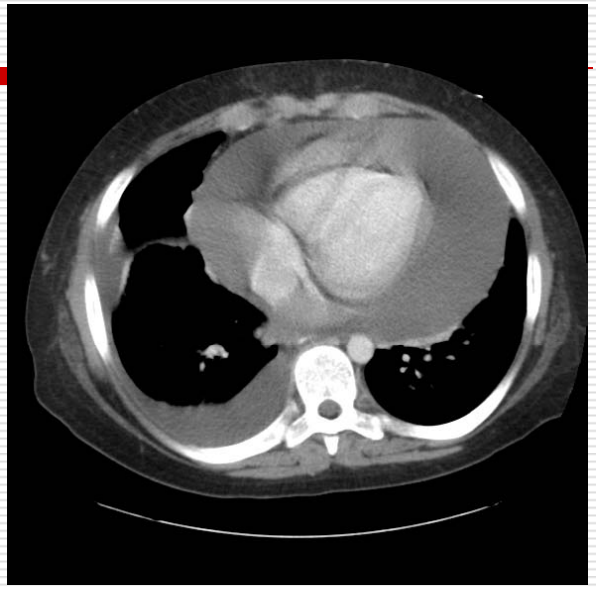
Superior Mediastinal Syndrome (SMS)

= Oncologic Emergency

HD – 9 y/o CT scan with SMS



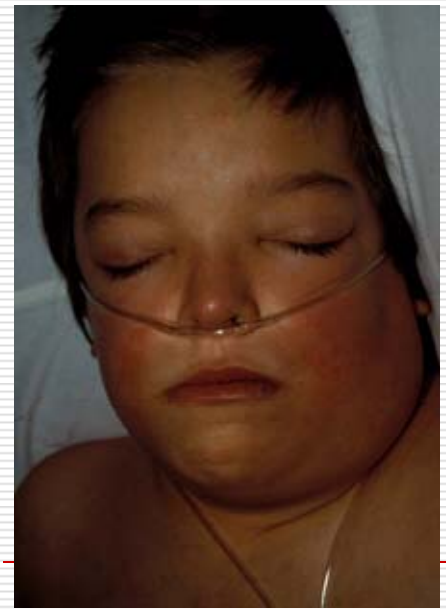
Ant. mediastinal mass compressing trachea; Pleural effusion

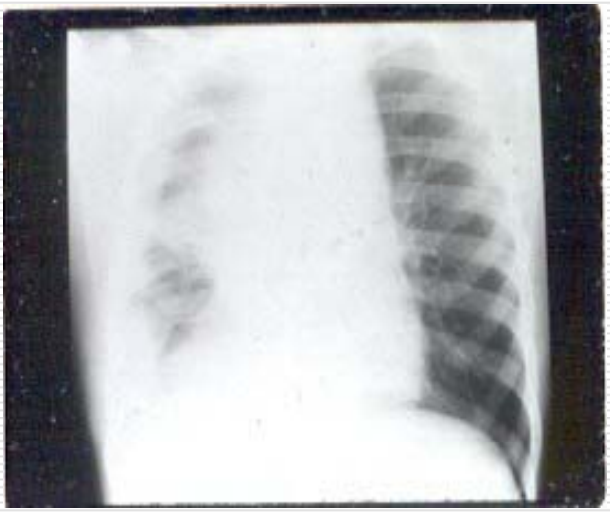


Pericardial effusion with tamponade

Superior Vena Cava (SVC) Syndrome in 10 y/o with **Lymphoblastic Lymphoma**

Facial swelling, plethora, cyanosis, enlarged neck veins



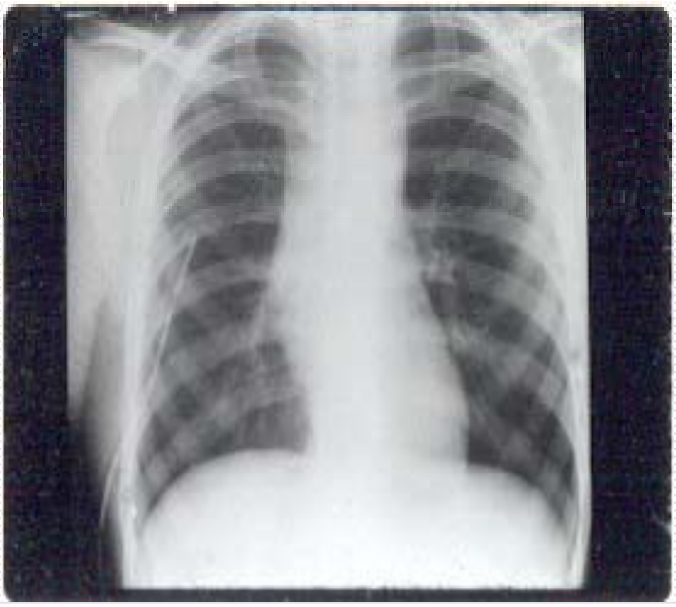


Mediastinal mass: tracheal and SVC compression

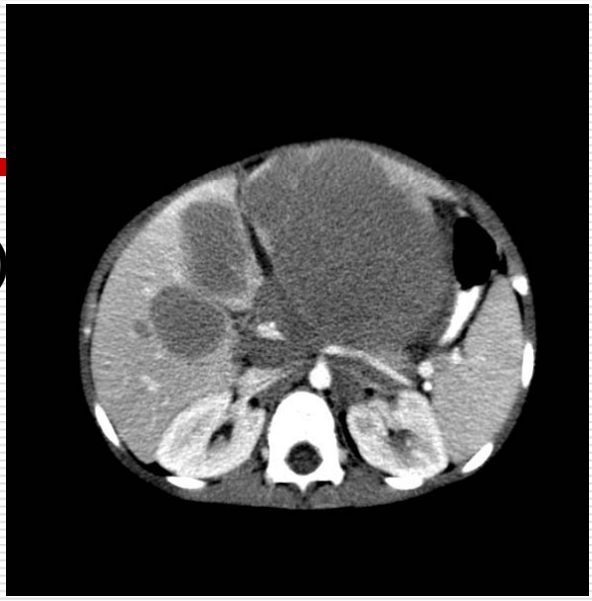
Lymphoblastic Lymphoma (T-cell, thymus)

Same boy 1 week after initial treatment

- **rapid onset**
- **rapid response**



Burkitt's Lymphoma



- B-cell origin
- Epsein-Barr virus (90%)
- > 5 y/o
- Abdominal mass
  - Large mass + □ LNs
  - Cecum or appendix
- Nasopharynx
- Tumor lysis syndrome
  - □ Uric acid, phosphorus, creatinine
  - Treatment can precipitate renal failure

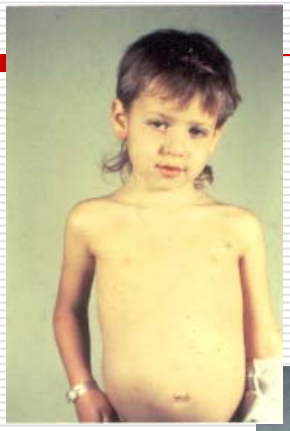
= Oncologic Emergency

Other Abdominal Tumors

Malignant Abdominal Masses

Most common:

- **Burkitt's lymphoma**
- **Neuroblastoma**
- **Wilms Tumor**



Other:

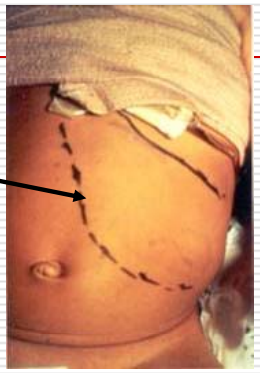
- Hepatoblastoma
- Rhabdomyosarcoma
  - –pelvic
- Ovarian germ cell tumors
  - – pelvic

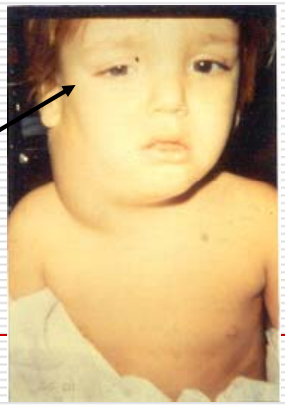
Neuroblastoma

- **Age**
  - 90% < 5 y/o; 50% < 2 y/o
  - Occasional USG detection *in utero*
- **Location:** any neural crest tissue
  - Adrenal
  - Paraspinal sympathetic tissue
    - Cervical, Thoracic, Pelvic
- **Often metastatic at diagnosis**
  - Bone and/or bone marrow
    - > 1 y/o: 70%

Neuroblastoma: Signs and Symptoms

- **Abdominal mass**
  - Often crosses midline
- **Lower extremity weakness**
  - Spinal cord compression
    - Thoracic
    - abdominal
- **Cervical, high thoracic mass**
  - Horner's syndrome
    - Myosis, ptosis



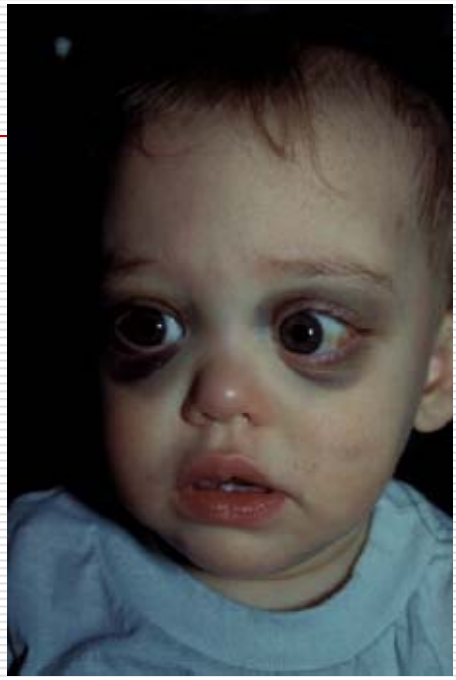


Neuroblastoma: Signs and Symptoms

- Signs of metastatic disease
  - Irritability
  - Weight loss
  - Bone pain
  - Fever
  - Proptosis
  - Bone lesions
  - Periorbital ecchymoses

More Periorbital Ecchymoses of Neuroblastoma

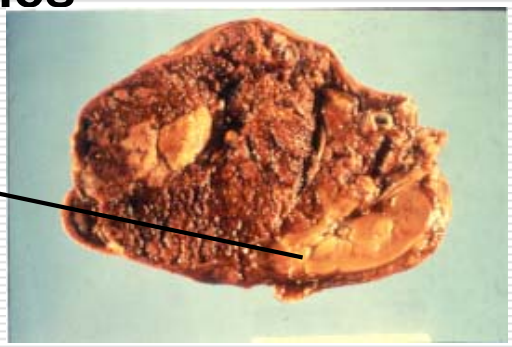
13 months old at diagnosis



1 month into therapy

Neuroblastoma: Signs and Symptoms

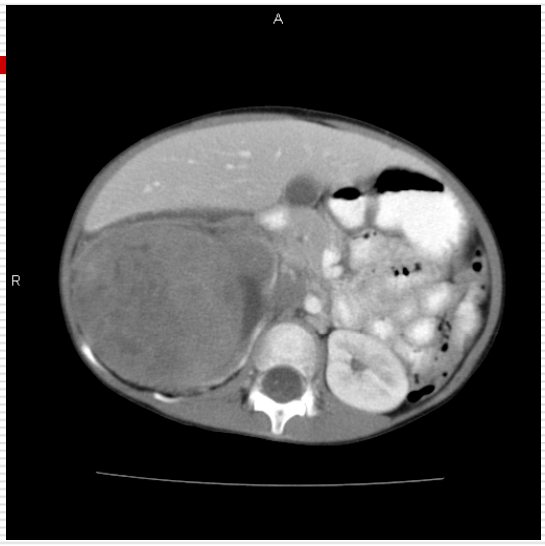
- **Paraneoplastic syndromes**
  - Watery diarrhea – Vasoactive Intestinal Peptide
  - Opsoclonus-myoclonus, cerebellar ataxia
    - Cross-reacting antibodies
- **↑ Urinary catecholamines**
  - VMA/HVA – 85%
- **↑ BP** – 25%
  - Renal compression
  - Catecholamine secretion



Wilms tumor: Signs and Symptoms

- **Abdominal mass**
  - Often asymptomatic
    - Healthy appearing

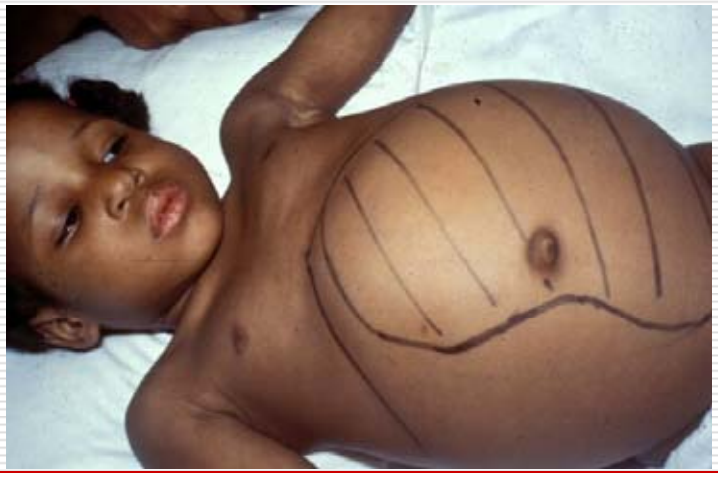
2 days before dx



Encapsulated mass

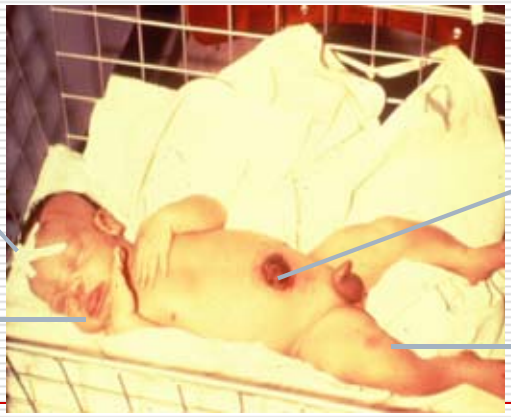
Wilms tumor: Signs and Symptoms

Mass enlarges toward pelvis



WB Syndrome and Wilms tumor

- Beckwith-Wiedemann syndrome - regular screening for nephroblastoma

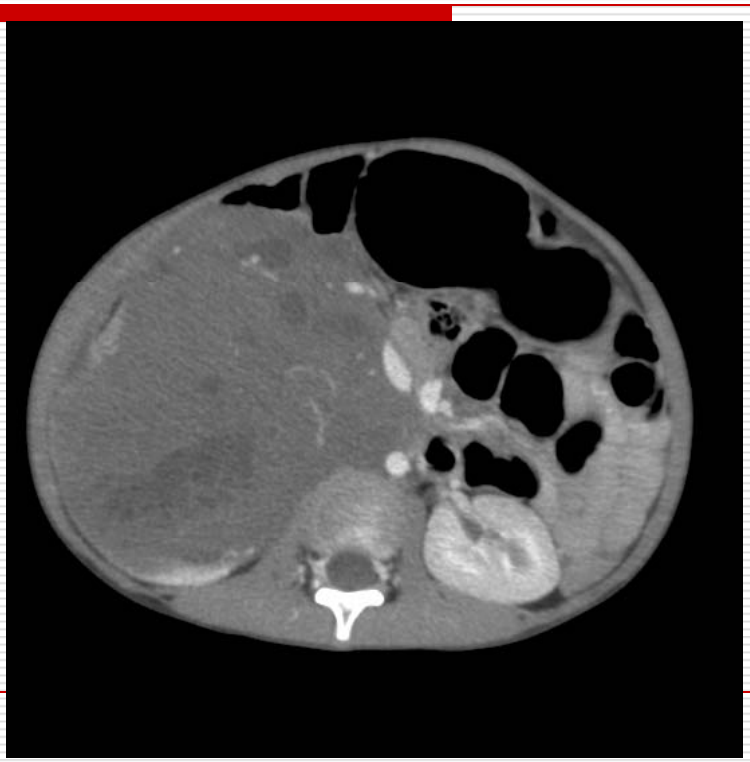


Presentation

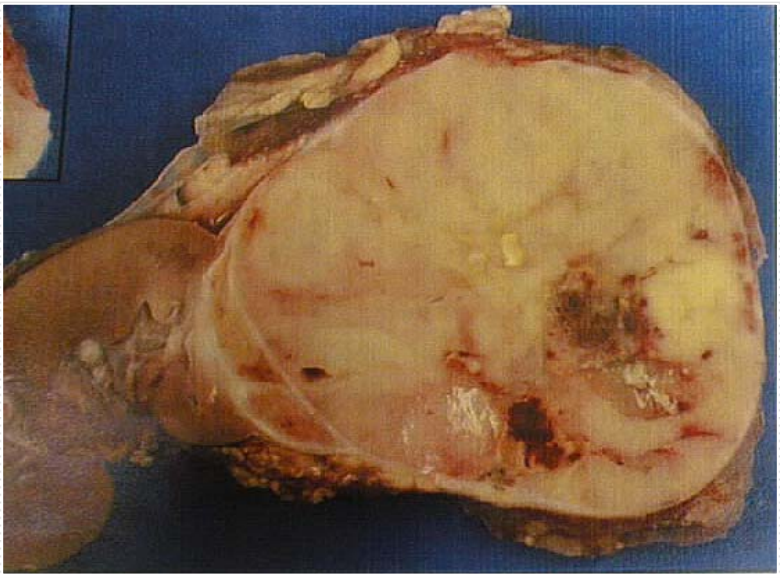
- 6 yo male with abd mass
- 2week hx of abdominal distension
- Otherwise, healthy

- **Test**

CT scan - Stage III Wilms Tumor



Wilms tumor after total nephrectomy

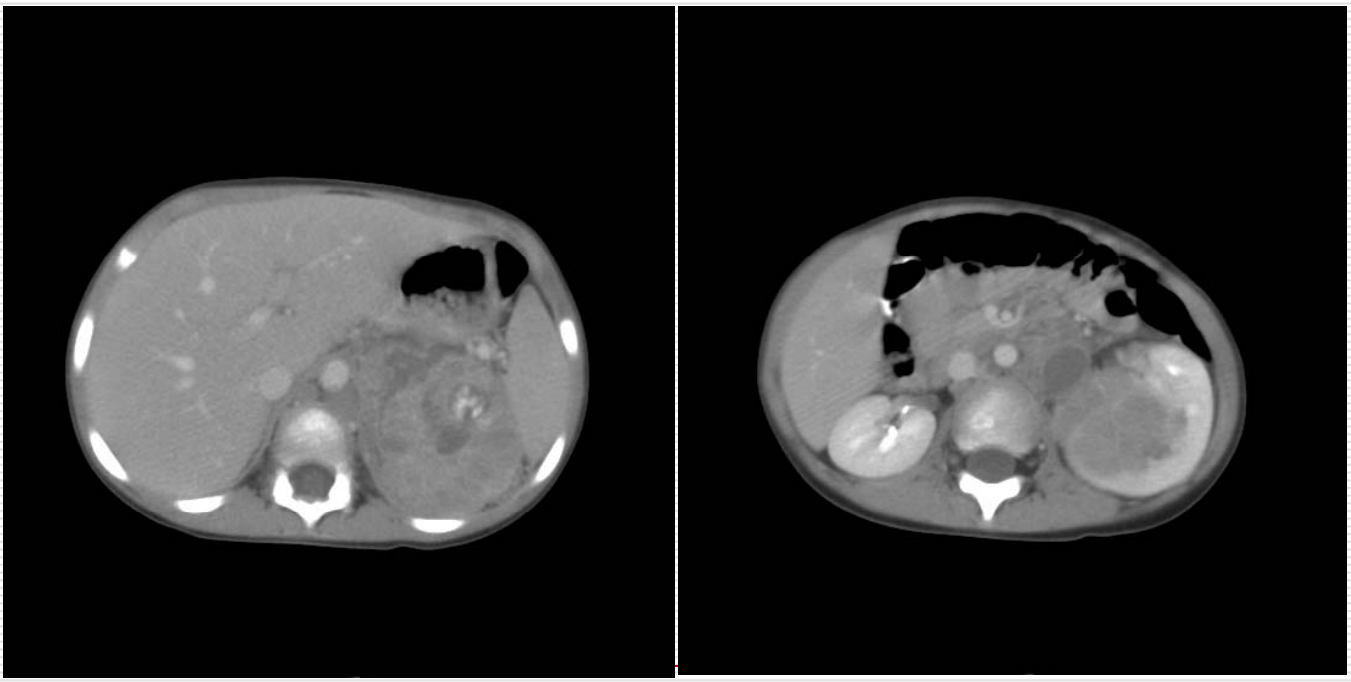


VS. another presentation

- 3yo male with 1 month of fevers
- Irritable, not walking X 4days with bone pain and fevers
- Pale

Test

CT scan - Stage IV Neuroblastoma



Bone tumors

Bone Tumors in Childhood

- **Age** – Adolescents > younger children
- Signs and symptoms
  - **Bone pain**, □ palpable mass, □ □ motion
    - Often hx of sports injury (coincidental)

Osteogenic Sarcoma

Metaphyses of long bones:

Distal femur

Proximal tibia

Proximal humerus

Pelvis

Ewing Sarcoma

All bones:

Long: diaphyses

Flat

Pelvis

Skull

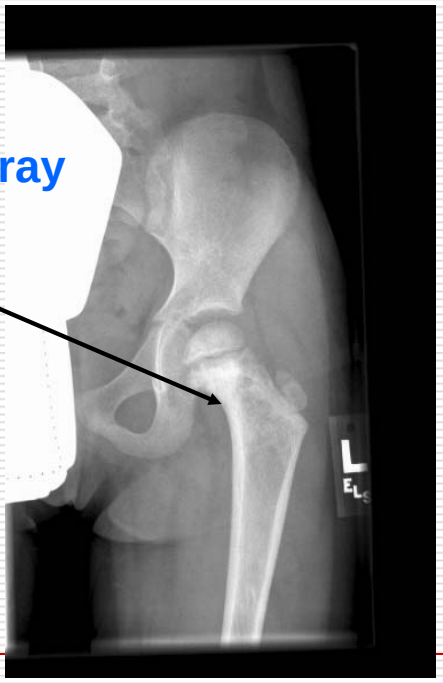
Ribs

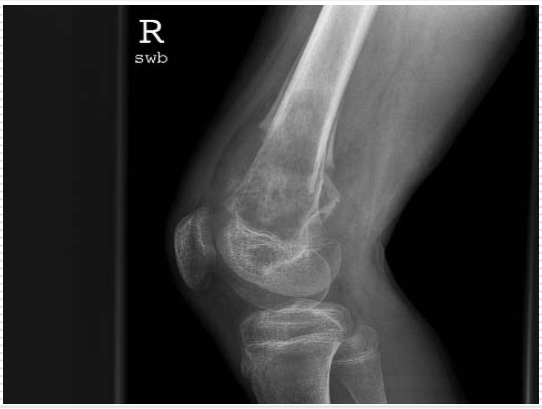
Presentation of Bone Tumors

Plain X-Rays are usually abnormal

Classic X-ray of Ewing:

Moth-eaten lytic lesion





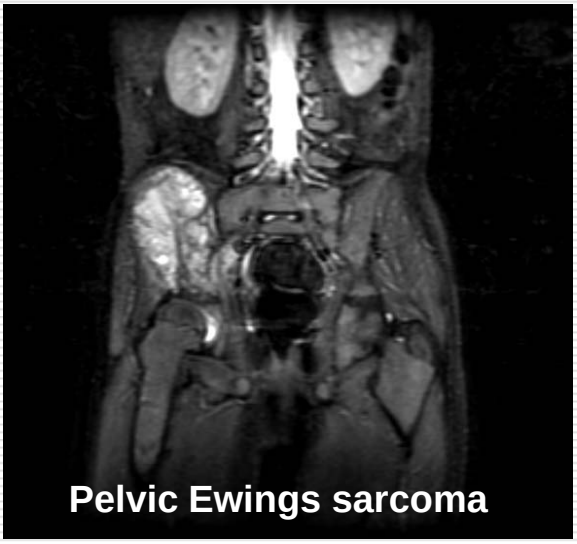
Classic X-ray of O.S.:

"Sunburst pattern"
Periosteal reaction
Soft tissue mass + calcium

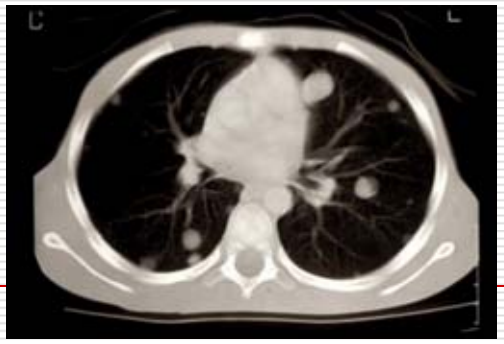
Presentation of Bone Tumors

Further radiographic evaluation may help with differential diagnosis of bone pain

- Bone scan
- MRI
- Chest CT scan
  - Metastases 20%



Pelvic Ewings sarcoma



Soft tissue sarcomas

Presentation of Soft Tissue Sarcomas

- **Rhabdomyosarcoma – most common** (origin striated muscle)
- **Age**
  - Birth to > 20 y/o
  - 70% < 10 y/o
- **Sites**
  - Head and neck – 40%
  - Genitourinary – 20%
  - Extremities – 20%
  - Trunk – 10%
  - Retroperitoneal – 10%

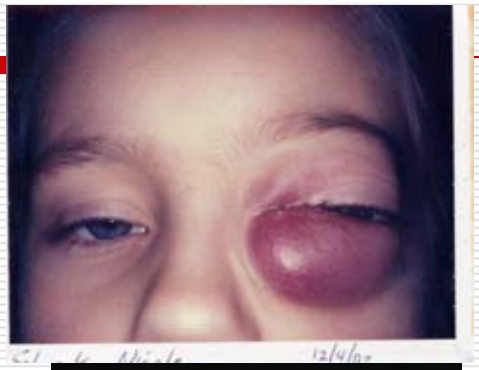
Signs and symptoms depend on age and site

Rhabdomyosarcomas: Signs and Symptoms

Head and neck

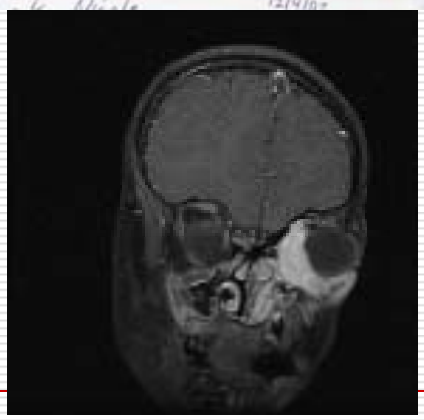
Orbit

- Proptosis
- Periorbital swelling



Parameningeal

- Cranial nerve palsies
- Hearing loss
- Chronic aural or sinus drainage



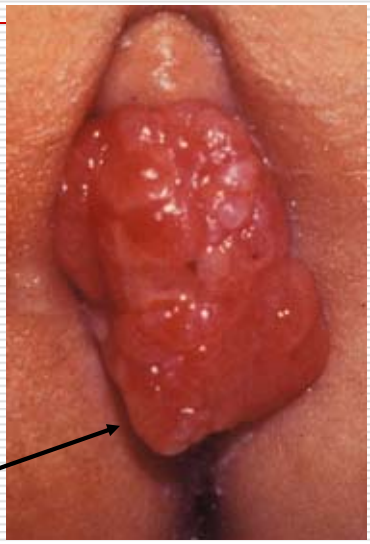
Same patient:

S/P radiation
and chemo

3 months off Rx:
eye lashes regrown

Rhabdomyosarcomas: Signs and Symptoms

- Genitourinary
  - **Bladder and prostate**
    - Hematuria
    - Urinary obstruction
  - **Paratesticular**
    - Painless mass - ↑ testicle
  - **Vagina and uterus**
    - Abdominal mass
    - Vaginal mass
    - Vaginal bleeding or discharge



Botryoid: grape-like

Concluding Remarks

In developed countries, over 70% of children diagnosed with cancer will be cured of their disease (better prognosis than cancer in adults)

- **Children should be followed throughout adulthood for potential late effects of therapy and second malignancies.**