Common Presentations of Childhood Cancer

First Take Home point

- Childhood Cancer is a rare disease...

New Cases of Cancer in the U.S. in 2003

<table>
<thead>
<tr>
<th>Cancer Type</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breast</td>
<td>25,000</td>
</tr>
<tr>
<td>Lung</td>
<td>12,500</td>
</tr>
<tr>
<td>Prostate</td>
<td>14,000</td>
</tr>
<tr>
<td>Childhood Cancers</td>
<td>12,000</td>
</tr>
</tbody>
</table>

BUT...

- One in every 330 Americans develops cancer before age 20.
- 1 in 750 20-year-olds alive in the U.S. today is a survivor of childhood cancer.

More Statistics

- Over 3,000 children die from cancer yearly in the U.S.
- More than from AIDS, asthma, diabetes, and cystic fibrosis combined

Earlier diagnosis and referral can impact 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
Specific Signs and Symptoms

Depend on

* type of cancer
* site(s) of disease
* age of patient

Cancer in Younger Children

Neuroblastoma
Retinoblastoma
Wilms tumor

Age (yrs) at diagnosis
SEER data, 2002

Cancer in Children and Adolescents

ALL
AML
Lymphoma
Brain Tumors

Age (yrs) at diagnosis
SEER data, 2002

Leukemias

# 1 4 y/o fever x 4 days
ear pain = L otitis, began Amoxicillin
T = 104 + chills next day → urgent care again

PE:
L TM purulent drainage
bilateral cervical, L axillary, L inguinal LNs↑
spleen 5 cm ↓, liver 3 cm ↓
multiple bruises extremities

→ Test
**CBC**
- WBC = 33K
  - 8% Neut, 92% lymphocytes
- Hb = 6.5
- PLT = 40K

**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
  - Orthopnea, cough
  - mediastinal mass
  - tracheal compression
  - Facial nerve palsy
  - Testicular enlargement
  - Skin lesions
  - Gingival hypertrophy
- Fever of malignancy

**CBC and Differential**
- Very helpful in the diagnosis ALL
  - ↑WBC - 50%; nl or ↓WBC – 50%
  - ↑blasts on smear in 80%
  - ≥2 Cytopenias - 95%
    - ↓Hgb - 80% ↓Plts – 90%
    - ↓% Neutrophils – 90%
  - 1 Cytopenia - 4%
  - Normal CBC and diff – 1%
CNS Tumors

Presentation

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

Brain Tumors of Childhood

Heterogeneous
- Cell of origin: glial, neural, other, combination
- 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
  - Mediastinum ± hilum
  - LNs below diaphragm and spleen
  - Liver, lung, bone marrow

95%

Presentation of Hodgkin's Disease

- **Systemic symptoms**
  - Fevers
  - Night sweats
  - Weight loss
  - Pruritus
- "B" symptoms
  - 25%
- Superior Mediastinal Syndrome (SMS)
  - Orthopnea, SOB, stridor, hypoxia
  - Tracheal
    - Bronchial
    - Cardiac

= 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, 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
- > 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

Malignant Abdominal Masses
Most common:
- Burkitt’s lymphoma
- Neuroblastoma
- Wilms Tumor

Other:
- Hepatoblastoma
- Rhabdomyosarcoma
  - pelvic
- Ovarian germ cell tumors
  - pelvic

Other Abdominal Tumors

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
    - Miosis, ptosis, anhydrosis
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

Same patient: 5 years later

- 12 years later

Neuroblastoma: Signs and Symptoms

- Paraneoplastic syndromes
  - Watery diarrhea – Vasoactive Intestinal Peptide
  - Opsoclonus-myoclonus, cerebellar ataxia
  - Cross-reacting antibodies

- ↑ Urinary catecholamines
  - VMA/HVA – 85%

Wilms tumor: Signs and Symptoms

- Abdominal mass
  - Often asymptomatic
  - Healthy appearing

- Mass enlarges toward pelvis

- BP – 25%
Signs and Symptoms of Wilms tumor

- Associated anomalies, syndromes – 15%
  - Hemihypertrophy
  - Aniridia
  - WAGR syndrome
    - Wilms, aniridia, ambiguous genitalia, retardation

- More anomalies, syndromes
  - GU anomalies
  - Denys-Drash syndrome
  - GU anomalies and renal failure
  - Beckwith-Wiedemann syndrome

Presentation

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

Test

CT scan - Stage III Wilms Tumor

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 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

**Presentation of Soft Tissue Sarcomas**
- **Rhabdomyosarcoma** – most common
- **Age**
  - Birth to > 20 y/o
  - 70% < 10 y/o
- **Sites**
  - Head and neck – 40%
  - Genitourinary – 20%
  - Extremities – 20%
  - Trunk – 10%
  - Retroperitoneal – 10%

**Soft tissue sarcomas**
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

Rhabdomyosarcoma – other sites

- Can show up at any site and any age
- 6 week old
- Newborn

Concluding Remarks

Over 70% of children diagnosed with cancer will be cured of their disease.

- 1 in every 1000 young adults alive in the U.S. today is a survivor of childhood cancer.
- Children should be followed throughout adulthood for potential late effects of therapy and second malignancies.