Neuro Oncology- Children and Brain tumors

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Disclosure

- I have no conflict of interest or financial relationship with any topics mentioned in this presentation.

Objectives

- Common types of tumors in children
- Signs and symptoms at presentation
- Diagnostic workup and treatment planning
- Interventions
- Common CNS Complications

Epidemiology

- 30% of all pediatric cancers
  - ~ 3000/year
- Greatest incidence ages 0-4 years
- Greatest incidence in Caucasian children
- Location
  - infratentorial 65%
  - supratentorial 35%

Epidemiology –cont’d

- Most common cause of childhood cancer mortality
  - 25% of deaths
- 60% overall survival
CNS Tumors

**Primary**
- arise from normal cells within the brain
- name includes cell of origin +/- location
- "benign" or malignant

**Metastatic**
- originate outside CNS
- hematogenous spread
- always malignant
- symptoms +/- primary diagnosis or relapse

LOCATION, LOCATION, LOCATION

**Benign**
- typical of cell of origin
- ↓ mitosis/necrosis
- slow growing/low grade
- localized
- may be malignant by location

**Malignant (brain cancer)**
- greater differentiation
- ↑ mitosis/necrosis
- fast growing/high grade
- invade surrounding brain tissues
- metastasize outside CNS

Prognosis

- Depends on several factors including:
  - Age of child at diagnosis
  - Location of tumor
  - Resectability
  - Pathology

Brain tumor Classification

- Astrocytoma/Glial Tumors
- Brain Stem Gliomas
- Medulloblastomas
- Ependymomas
- Midline Tumors/Craniopharyngiomas
- Spinal Cord Tumors

Clinical Presentation

- Generalizing symptoms
  - Caused by increased intracranial pressure
  - Headache, nausea, vomiting, fatigue
  - Decreased up gaze, sixth cranial nerve palsies, papilledema
  - Infants: macrocephaly, failure to thrive, developmental delay
Other presenting symptoms

- Flu-like syndrome
- AM HA/vomiting
- Ataxia
- Cranial nerve deficits; families may report a "crooked smile" or "lazy eye" or "double vision"
- Impaired vision/diplopia
- Mental status changes
- Loss of developmental milestones
- ↑ head circumference
- Seizures 10-15%

Primitive Neuroectodermal Tumor/ Medulloblastoma (PNET/MB) - Epidemiology

- Most common CNS malignancy in childhood
- 20% of primary CNS tumors
- Mean age at presentation: 3-4 years
- More common in males
- Medulloblastoma = posterior fossa PNET
- Presentation
  - 72% are located in posterior fossa
  - 12% are hemisphere
  - 7% are pineal
- Very invasive
  - CNS metastases in 11-43% at presentation
  - Extraneural spread

PNET therapy

- Surgery
- Radiotherapy
  - Local: 5400 – 5940 cGy
  - Craniospinal
    - High Risk: 3600 cGy
    - Standard Risk: 2340 cGy
- Chemotherapy (active agents)
  - Vincristine, lomustine, cisplatin, etoposide, cyclophosphamid

PNET survival rates

- Standard Risk: 86% 3yr PFS
- High Risk: 67% 5yr PFS
- Infants: 30% 5yr PFS
- Pineal: 61% 3yr PFS
- Supratentorial (non-pineal):
  - 33% 3yr PFS

Duffner et al., Neuro-oncol 1999;1:152.
Ependymoma
- 8-10% of childhood brain tumors
- Most occur in kids < 7 yo
- 60% are infratentorial
- 2-5% dissemination at diagnosis

Ependymoma prognostic factors
- Extent of resection
  - Complete: 50-70% 5yr PFS
  - Incomplete: 0-30% 5yr PFS
- Histology (anaplastic)
- Metastases (+)
- Age (< 3 years old)
- Location (infratentorial)

Ependymoma treatment
- Surgery - aggressive resection
- Radiation
  - Focal: 5400 - 5940 cGy
  - Craniospinal: if metastatic disease
- Chemotherapy
  - Several active agents
  - Minimal impact on survival
  - Used in recent trials in attempt to shrink residual tumor
- Observation
  - May be feasible for supratentorial well-differentiated tumors

Glioma - Astrocytoma Grading
- Low-Grade I
- (benign) II
- High-Grade III
- (malignant) IV
  - Pilocytic astrocytoma
  - Fibrillary astrocytoma
  - Anaplastic astrocytoma
  - Glioblastoma Multiforme

LGG
- Most common pediatric brain tumor
- Locations
  - Cerebellum
  - Cerebral hemispheres
  - Deep midline structures
  - Optic pathway/hypothalamus
- MRI
  - Hypointense T1, hyperintense T2
  - Pilocytic: well-circumscribed, cystic component, enhancing mural nodule
  - Fibrillary: little enhancement
Optic Pathway/Hypothalamic Glioma (OP/HG)
- 5% of pediatric tumors
- 2/3 diagnosed before age 5yø
- 70% associated with NF1
- 15% of kids with NF1 have an OP/HG
- Diagnosis often by MRI alone (esp. if NF1)
- Most are fibrillary astrocytomas, although pilocytic astrocytomas are common as well

Supratentorial Malignant Glioma
- 11% of pediatric brain tumors
- MRI
- Staging – consider spinal imaging and cytology

Supratentorial Malignant Glioma – Prognostic Factors
- Extent of surgical resection
  - Median PFS (<90% vs. >90% resection)
    - Grade III: 12 vs. 31 months
    - Grade IV: 8 vs 12 months
Supratentorial Malignant Glioma – Treatment
- Surgery – aggressive resection
- Radiotherapy
  - Focal with 2cm margin
  - Prolongs survival
- Chemotherapy
  - Modest impact on long-term outcome
  - Temozolamide
  - New agents targeting:
    - Tumor neovascularization
    - Specific molecular abnormalities

Brain Stem Glioma (BSG)
- 10-20% of pediatric brain tumors
- Median age at diagnosis is 6.5yrs
- Types
  - Diffuse intrinsic
  - Dorsally exophytic

Diffuse Intrinsic BSG
- 80% of brain stem tumors
- Most arise in pons
- Presentation
  - CN deficits (usually VI/VII)
  - Hemiparesis
  - 10% with hydrocephalus
- Pathology – high-grade glioma

IPG

Prognostic factors (unfavorable)
- Short duration of symptoms at diagnosis
- CN deficits at diagnosis
- Pontine location
- Age <2yo
**IPG**
- Surgery
  - No role
  - Biopsy rarely needed
- XRT
  - 54-55.8 Gy
  - Prolongs survival
  - Neurologic improvement
- Chemotherapy
  - Nothing useful so far
- Survival
  - Median 9-13 months
  - 2 yr survival is <20%

**Craniopharyngioma**
- 6-9% of pediatric brain tumors
- Over 50% of suprasellar tumors
- Peak incidence in kids is 6-10 yrs
- Benign histology
- MRI
  - Heterogeneous with cystic, solid, calcified components
  - Hypointense on T2, enhancing
- Work-up
  - Hormone testing
  - Ophthalmologic evaluation – acuity and fields

**Cranio Treatment**
- Surgery
  - Primary modality
  - Complete resection → improved PFS
- Radiotherapy
  - Subtotal resection + radiation
  - Survival similar to complete resection
- Prognosis
  - 60-80% 5yr PFS
  - 95% 5yr OS
- Multidisciplinary follow-up
  - Neuro-oncology, ophthalmology, endocrinology

**Cranio pre op and post op**

**Tumors that have Leptomeningeal Spread**
- PNET
- Germ cell tumors
- Ependymoma
- Atypical teratoid/rhabdoid tumor
- Glioblastoma multiforme
Diagnostic Testing

- MRI: can reveal hydrocephalus (this is the cause of the headaches and morning vomiting).
- Lumbar puncture: usually completed approximately 2-3 weeks post-operatively to detect the presence of tumor cells in the cerebrospinal fluid
- MRI spine: to rule out metastases
- PET- glucose uptake by tumor cells

Staging Evaluation

- Post-OPERATIVE MRI BRAIN to assess extent of resection
- Bone marrow aspirates and biopsies in children with highly malignant tumors to assess for spread outside the central nervous system
- CT SCAN Chest/Abdomen/Pelvis

Brain Tumor Treatment Modalities

- **SURGICAL RESECTION**
- **RADIATION**
- **CHEMOTHERAPY**

Surgery

- Primary objective: gross total resection
- Secondary objective: correct obstructive hydrocephalus -75% children will require VP shunt post-operatively

Radiation Therapy

- Avoid in children <7yo if possible
- Focal vs. craniospinal XRT
- Administered over a period of six weeks; daily for 5 days/week
- Daily anesthesia possibility
- No risk of Radiation exposure to staff
Chemotherapy

- What is it?
- What is the role in brain tumors?
- How does it cross blood/brain barrier?
- What is the goal of chemotherapy?

CNS Complications

- Diabetes Insipidus
- Cranial Nerve Deficits
- Cognitive Deficits and Behavioral Changes
- Posterior Fossa Syndrome

Supportive Care Issues

- Risk of ICP
- Management of nausea and emesis
  - Many children experience nausea long after completing treatment
  - Many require multiple agents to control nausea
- Management of nutritional deficits
  - Due to continued nausea and early satiety
  - Enteral feedings vs. TPN

Supportive Care

- Management of Endocrinopathies
  - Some children may require thyroid replacement, cortisone replacement, or management of fluids and electrolytes
  - Common cause of morbidity
  - Obesity an issue for many hypothalamic tumors
- Rehabilitation Needs
  - PT/OT needs throughout therapy

Supportive Care Issues

- Management of Radiation Side Effects
  - Children receiving craniospinal have a high incidence of radiation esophagitis and gastritis
  - Radiation Somnolence; very scary for families
- Psychosocial Support
  - Parents often report mourning the loss of their child: “he just isn't the same since the surgery.”
  - Guilt: “he had been vomiting for weeks. I just thought it was a virus.” “It is all my fault.”

Cognitive/Behavioral Changes

- Head trauma
  - Cerebral edema
  - Hemorrhage
- Vascular insults/neoplasms
- Status epilepticus
- CNS infections
- Metabolic
  - Fluid/electrolyte imbalance
  - Medications
Posterior Fossa Syndrome
- CNS deficits after posterior fossa surgery
  - occurs 24-48 hours post-op
  - usually transient lasting 2-6 months
  - may have mutism, weakness, dysphasia, hemiparesis
- Medical/Nursing Management
  - +/- steroids
  - treat hydrocephalus if indicated
  - safety R/T to impaired mobility
  - emotional support
  - OT/PT

CNS deficits after posterior fossa surgery usually transient lasting 2-6 months may have mutism, weakness, dysphasia, hemiparesis.

Medical/Nursing Management
- +/- steroids
- treat hydrocephalus if indicated
- safety R/T to impaired mobility
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Treatment Implications
- Devastating disease
  - high morbidity
  - high mortality
  - Permanent residual deficits
  - Interferes with
    - work/school
    - development/maintenance of family/social relationships
  - Financial burden
  - Loss of previous lifestyle/independence

Role of Nurse
- Assessments
- Interventions
- Expected Patient and Family Outcomes

References
National Brain Tumor Foundation (NBTF). http://www.nbtf.org
American Brain Tumor Association (ABTA) http://www.abta.org