SOLID TUMOURS IN CHILDHOOD

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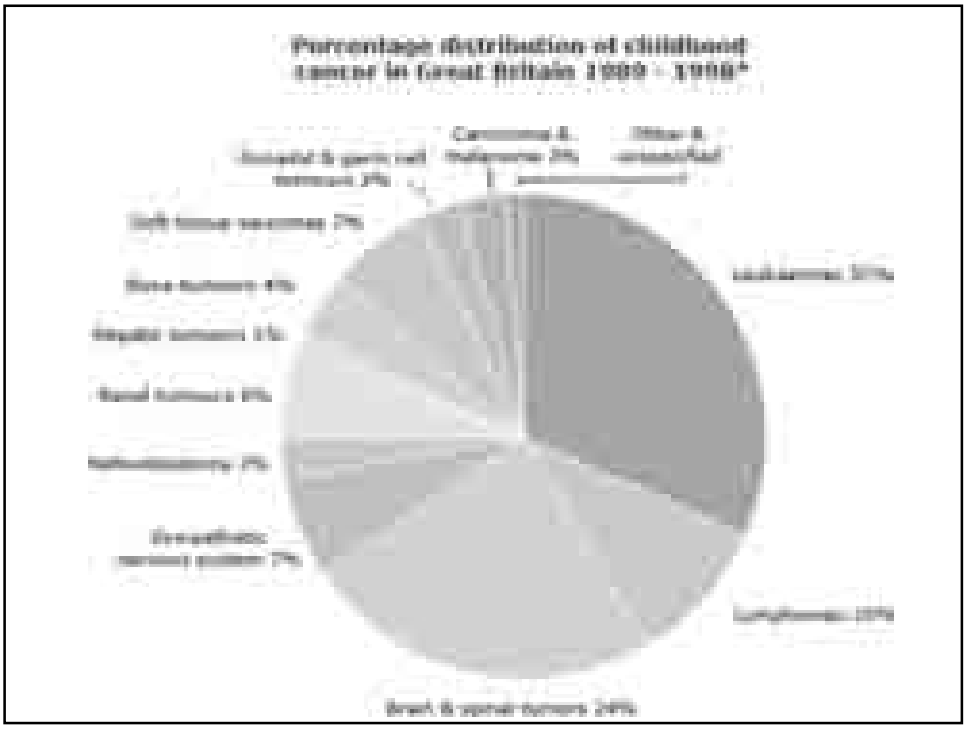
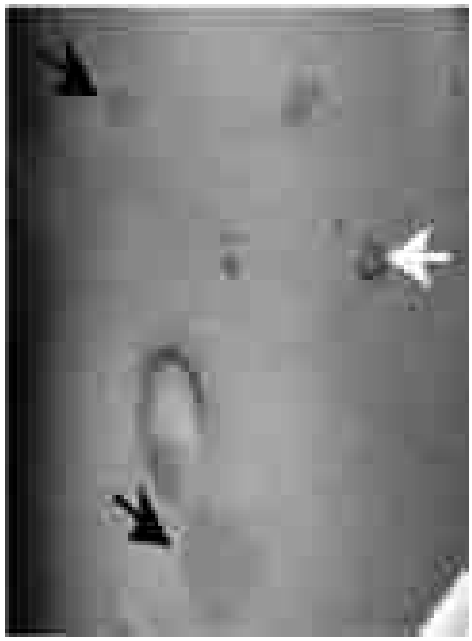


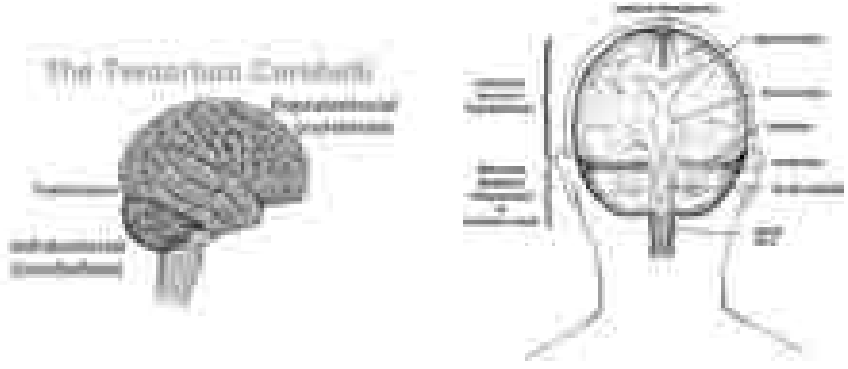
Introduction

- Lymphomas and Leukemias make up about 40% of all childhood Cancers (Systemic cancers)
- 60% of childhood cancers are solid tumors
- About 40% of all solid tumors are CNS tumors

CNS tumours

- 25% of all childhood cancers (2nd most common after leukemia)
- Association with:
  - Neurofibromatosis type I
  - Tuberous Sclerosis
  - Von Hippel-Lindau syndrome
- Supratentorial (40%) vs Infratentorial (60%)



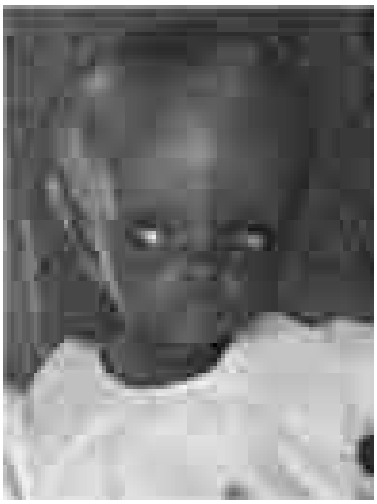


CNS tumours

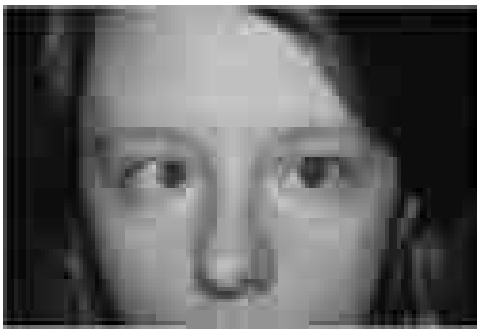
- **Supratentorial**
  - Glioma
    - High grade
    - Low grade
  - Ependymoma
  - PNET
  - Craniopharyngioma
  - Optic nerve glioma
- **Infratentorial**
  - Cerebellum
    - Medulloblastoma
    - Astrocytoma
    - Ependymoma
  - Brain stem
    - Glioma
    - Other

Clinical

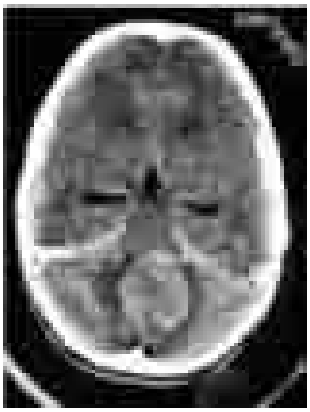
- Headache/irritability, vomiting
- Disturbance of gait and balance
- Hemiparesis/hemiplegia
- Cranial nerve abn: diplopia
- Mental disturbances
- Seizures
- Endocrine abnormalities
- Macrocephaly

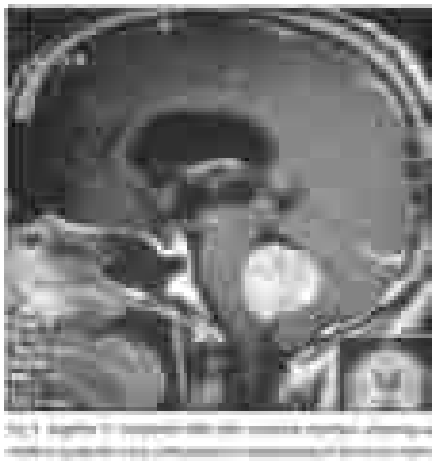


Hydrocephalus



Right cranial nerve VI palsy





Medulloblastoma (Infratentorial)

Management

- Complete surgical resection if possible
- Multimodality treatment approach:
  - Surgery
  - Chemotherapy
  - Radiotherapy

Neuroblastoma

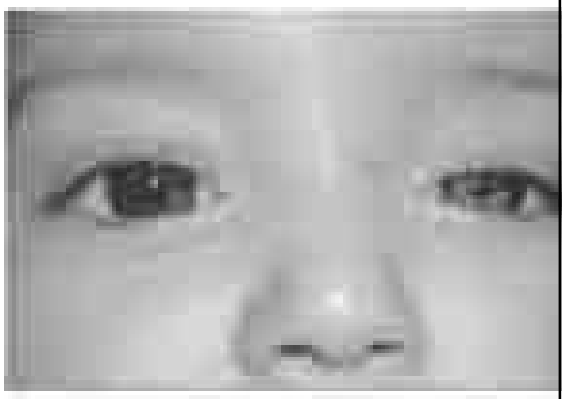
- 7% of all childhood malignancies
- Peak age of incidence: 2 years
- Associated with:
  - NF type I
  - Hirschprung disease
  - Turner syndrome
  - Noonan syndrome

Clinical

- Asymtomatic abdominal mass
- Adrenal mass most common
- Can arise anywhere along the sympathetic neural chain
- 50% - distant metastases at diagnosis

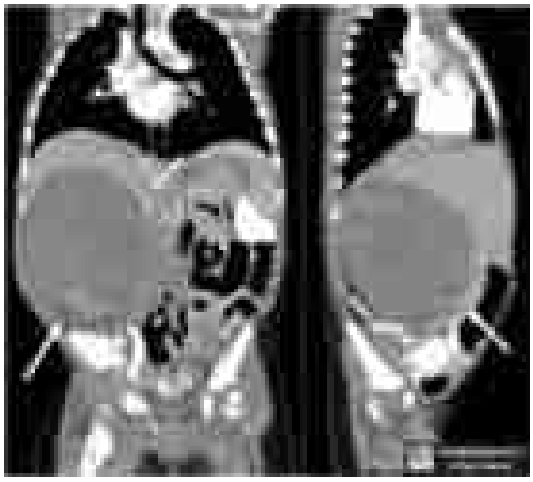
Anatomic site

- Head and neck: neck mass, horner's syndrome
- Orbit and eyes – raccoon eyes
- Chest: dyspnea, pulm infections
- Abdomen
- Pelvis
- Paraspinal: dumbbell tumor
- Bone pain



Other symptoms

- Sweating, flushing, pallor
- Hypertension
- Intractable watery diarrhea (kerner morrison syndr)
- Opsoclonus myoclonus ataxia syndrome (OMAS)



Diagnosis

- CT scan
- Histology
- ↑Urinary levels of catecholamines: homovanillic acid (HVA) and vanillymandelic acid (VMA)
- I^{123}-Meta-iodobenzylguanidine (MIBG) scintigraphy
- Bone scan
- Bone marrow aspirate

Staging and Treatment

- International Neuroblastoma Staging system (INSS): stage 1- 4 and 4s
- Neoadjuvant chemotherapy followed by surgery and adjuvant chemotherapy ± radiotherapy
- Stage 4s: excellent prognosis even with no treatment
- Stage 4: very poor prognosis

Nephroblastoma (Wilms' tumor)

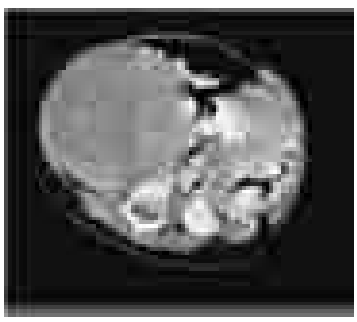
- 6% of all childhood cancers
- Median age: 44 months (unilateral tumors) and 32 months (bilateral)
- Associated with:
  - WAGR syndrome (Wilms', aniridia, genitourinary malf, mental retardation)
  - Denys-Drash Syndrome (Wilms', renal failure and pseudohermaphroditism)
  - Beckwith Wiedemann syndrome

Beckwith Wiedemann syndrome

- Visceromegaly
- Hemihypertrophy
- Macroglossia
- Abdominal wall defect
- Microcephaly
- Mental retardation hypoglycemia

Clinical

- Non-tender abdominal mass
- Microscopic hematuria
- hypertension



Staging

- Stage I: completely resected with intact capsule
- Stage II: completely resected with penetration of capsule
- Stage III: Incompletely resected and or lymphnodes positive
- Stage IV: distant metastases
- Stage V: bilateral

Management and Prognosis

- Neo-adjuvant chemotherapy followed by surgery and adjuvant chemotherapy
- ± Radiotherapy
- Stage I: 98% 4yr OS
- Stage IV: 40%

Retinoblastoma

- Mean age at diagnosis: 18 mo
- 40% of cases are hereditary – majority of these presenting with bilateral disease (15% of these have an established family history)
- 60% are non-hereditary mostly presenting with unilateral disease

Genetics

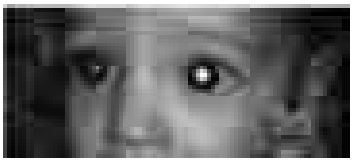
- Knudson's two hit hypothesis – 2 events are required for tumour initiation:
  - 1st –germline or somatic
  - 2nd – somatic in individual retinoblast cells
- RB1 gene (tumour suppressor gene) mutation located on chromosome 13q14
- RB1 gene inherited as autosomal dominant

Genetic counseling

- Unilateral / bilateral disease with + family history of RB:
  - 40% chance for sibling to get RB
  - 40% chance of having offspring with RB
- Bilateral with – family history of RB
  - 6% chance for sibling to get RB
  - 40% chance of having offspring with RB
- Unilateral with – family history of RB
  - 1% chance for sibling to get RB
  - 8% chance of having offspring with RB

Clinical

- Leucocoria
- Strabismus
- Decreased visual acuity
- Inflamatory changes
- Hyphema



Management

- If tumour small: local therapy:
  - Brachytherapy
  - Cryotherapy
  - TTT (Transpupillary thermal therapy)
  - Laser
- Chemotherapy
- Enucleation
- Combination of above esp for bilateral

Prognosis

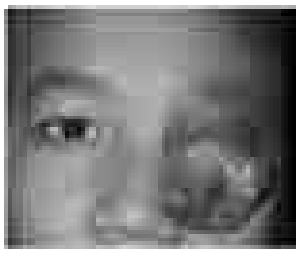
- Stage I-II: Excellent >90% survival (maybe at the expense of losing the eye)
- Stage IV (CNS and or distant metastetic spread): <30% survival
- Very important to refer early

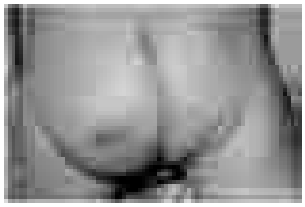
Soft tissue sarcomas

- Rhabdomyosarcoma (RMS) vs non-rhabdomyosarcoma soft tissue sarcomas (NRSTS)
- 7% of all childhood malignancies
- Peak ages: 2-6 years
- Most sporadic
- RMS: alveolar vs embryonal subtypes

Rhabdomyosarcoma Clinical

- Can occur anywhere in the body – muscle
- <10yrs: head and neck or genitourinary areas
- >10yrs: extremity, trunk or para-testicular
- Presents as a painless mass/swelling, or due to compression of mass eg. urinary outflow obstruction





Management

- Need to stage
- Neoadjuvant chemotherapy
- Complete surgical resection
- Adjuvant chemotherapy
- ±Radiotherapy
- Prognosis overall: >70% 5 yr survival (depending on stage)

Bone tumours

- Osteosarcoma (56%) vs Ewing sarcoma (34%) (more prevalent in caucasians)
- 6% of all childhood cancers
- Peak incidence: 15 years (growth spurt)
- Very aggressive tumours
- Majority at presentation have metastases

Clinical

Usually incidental finding after a history of minor trauma.

- Local pain/swelling
- Decreased range of motion
- Pathologic fracture

Management

- Need to stage
- Usually metastases to skeleton, lungs
- Neo-adjuvant chemotherapy followed by surgery
- Limb-sparing surgery if possible
- Adjuvant chemotherapy
- Radiotherapy for Ewing sarcoma
- If metastetic at presentation: very poor prognosis

Other Solid tumours

- Germ cell tumours
- Hepatic tumours: hepatoblastoma and hepatocellular carcinoma
- Carcinomas: usually seen in adults and sometimes in adolescents

Treatment of solid tumours

- Neo-adjuvant chemotherapy
- Surgery
- Adjuvant chemotherapy
- ±Radiotherapy
- Autologous stem cell transplant

Conclusion

- Early stage disease have excellent outcomes
- Therefore extremely important to refer early
- Stage IV disease: dismal outcome
- If in doubt: rather refer early and let us exclude cancer

- Questions